



GUIDE

**The Ottawa Hospital
Divisions of Endocrinology and
Metabolism and Neurosurgery**

**Pituitary Disease
Handbook for
Patients**



The Ottawa Hospital | L'Hôpital
d'Ottawa



Disclaimer

This is general information developed by The Ottawa Hospital. It is not intended to replace the advice of a qualified health-care provider. Please consult your health-care provider who will be able to determine the appropriateness of the information for your specific situation.

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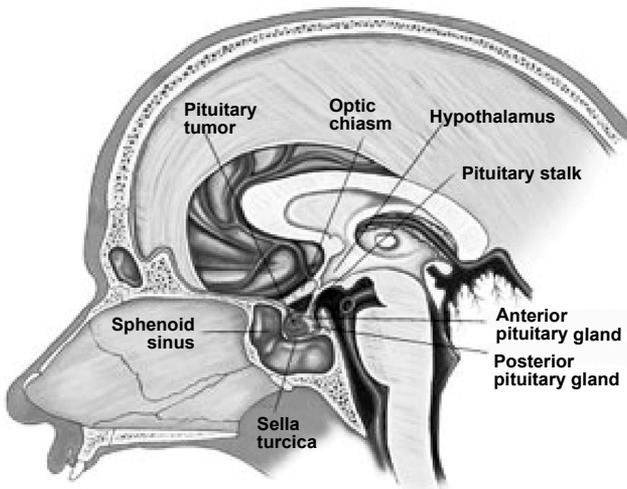
Outline

This handbook is designed to help people who have pituitary tumours better understand their disease. It contains information to help people with pituitary tumours discuss their care with health care providers. This booklet provides:

- 1) An overview of what pituitary tumours are and how they are grouped
- 2) An explanation of how pituitary tumours are investigated
- 3) A description of available treatments for pituitary tumours

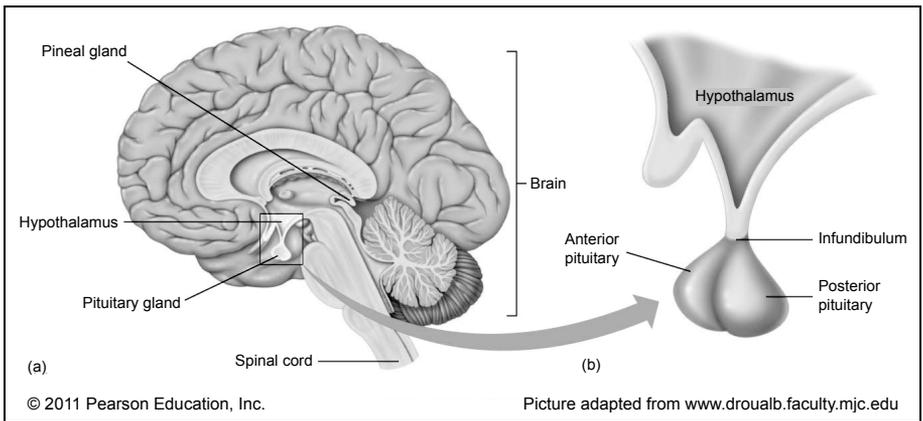
What is the Pituitary Gland?

The pituitary gland is a pea size organ located just behind the bridge of the nose at the base of the brain, in a bony pouch called the “sella turcica.” It sits just below the nerves to the eyes (the optic chiasm). The pituitary gland is divided into two main portions: the larger *anterior pituitary* (at the front) and the smaller *posterior pituitary* (at the back). Each of these portions has different functions, producing different types of hormones.



Picture provided by
pituitary.ucla.edu

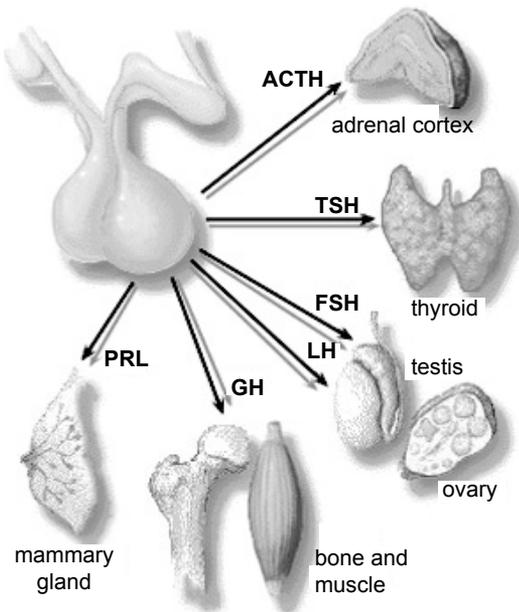
The pituitary gland is known as the “master gland” because it helps to control the secretion of various hormones from a number of other glands including the thyroid gland, adrenal glands, testes and ovaries. The pituitary gland releases various hormones into the blood stream, which then act on other glands and organs. The pituitary gland itself is controlled by the *hypothalamus*, which sits just above the pituitary gland. The hypothalamus acts like a thermostat to signal the pituitary gland to either secrete more or less hormones, depending on the needs of the body.



Hormones made by the pituitary gland	Glands or organs it affects	How it affects the glands or organs
Prolactin	Breasts	Produces milk for breastfeeding
Growth hormone (GH)	Many areas of the body	Controls growth and metabolism (how the body uses energy)
Adrenocorticotrophic hormone (ACTH)	Adrenal glands	Produces cortisol, which is important for dealing with stress

Hormones made by the pituitary gland	Glands or organs it affects	How it affects the glands or organs
Thyroid stimulation hormone (TSH)	Thyroid gland	Regulates metabolism (how the body uses energy)
Lutenizing hormone (LH) and Follicle stimulating hormone (FSH)	Ovaries and Testes	Controls reproduction (woman's ability to become pregnant)
Antidiuretic hormone (ADH)	Kidneys	Controls water balance

Hormones of the Anterior Pituitary



Picture adapted from www.resources.med.fsu.edu

What are Pituitary Tumours?

Pituitary tumours develop from a single abnormal cell that multiplies into many abnormal cells, eventually forming a tumour. What causes this is still not completely understood.

- Pituitary tumours are almost always benign (not cancer) and can be treated with medications and/or surgery, depending on the type of tumour. Although cancer can develop in the pituitary gland, this happens very rarely. Because the pituitary gland is important for other glands of the body to function, many different health care providers and regular follow up appointments are often needed to treat someone with a pituitary tumour.
- Most pituitary tumours are “pituitary adenomas”— the term “adeno” means gland, and “oma” means tumour. Other common types of pituitary tumours include craniopharyngioma and Rathke’s cleft cysts.
- Abnormal cells and small tumours of the pituitary gland are actually quite common. About 20 to 25 percent of the general population may have small, pituitary tumours that do not cause any problems and do not need to be treated.

Types of Pituitary Tumours

Pituitary tumours may be grouped and named by:

- the hormones they secrete, if any
- their size
- what the tumour cells look like under a microscope

Hormones

Almost 70 percent of pituitary tumours may make too much of a particular hormone. Tumours that make extra hormone may be called “hormonally active”, “secreting”, or “functioning”

adenomas. Sometimes these functioning pituitary tumours can produce more than one type of hormone.

Other pituitary tumours do not make hormones at all and may even stop making hormones the body needs (a condition called hypopituitarism). These tumours are called “non-functioning”, “non-secreting”, or “hormonally inactive”.

Size

Pituitary tumours are also classified by their size. Tumours less than 1 cm in diameter are called microadenomas. Those larger than 1 cm are called macroadenomas.

Tumour size is important because bigger tumours are more likely to affect vision by pressing on the nearby nerves of the eye or optic chiasm. When a big tumour is in the pituitary gland, the rest of the gland may not be able to make the normal hormones that the body needs.

Signs and Symptoms

People with pituitary tumours can have many different symptoms for any of the following reasons:

1. The tumour is producing too much hormone (hypersecretion). The extra hormone causes the symptoms. For example, too much of a hormone called prolactin may cause release of milk from the breasts (lactation).
2. Not enough pituitary hormones are being produced (hyposecretion).
3. A big tumour may push on the nearby eye nerves (optic chiasm), which could lead to changes in vision.

Specific Tumours, Hormone Changes and their Symptoms

1. **Prolactinomas:** Also known as prolactin-producing adenomas, these represent around 30 to 40 percent of pituitary adenomas, making them the most common type. Prolactinomas are often found in women of child-bearing age. In men they often appear during their 40s and 50s. Women with prolactinomas may have loss of regular menstrual periods or breast discharge, while men may have decreased sex drive or infertility.
2. **Growth Hormone-Producing Adenomas:** These tumours represent about 20 percent of pituitary adenomas and are more common in men than in women. These tumours often become quite large before they are noticed. Children who have this type of tumour grow more quickly (gigantism). When adults have this type of tumour, they do not grow taller, but they may notice changes in their facial features and in the size of their hands and feet (acromegaly).
3. **ACTH-Producing Adenomas:** These tumours represent about 16 percent of pituitary adenomas. They are much more common in women than in men. ACTH stimulates the adrenal glands to make cortisol. High cortisol in this condition causes Cushing's disease. People who have Cushing's disease may notice weight gain in the abdomen, face and upper back, thin skin, easy bruising and stretch marks. They may also have high blood pressure, and diabetes.
4. **Other Hormone-Secreting Pituitary Adenomas:** This group represents less than 1 percent of pituitary adenomas. Some of these tumours may produce TSH, LH or FSH. Often there are no symptoms caused by tumours making these hormones.

5. **Non-Secreting Adenomas:** Also called “non-functioning tumours”, these represent about 25 percent of the pituitary adenomas. These tumours usually grow slowly. If they grow to become large, they can cause headaches, problems with vision, and stop the pituitary gland from making hormones that the body needs.

Symptoms Caused by Pituitary Insufficiency (hypopituitarism)

When a pituitary tumour is large enough it compresses the normal pituitary gland. This can cause the normal pituitary gland to be unable to make hormones (hypopituitarism). The symptoms depend on the hormones involved.

- Reduction in sex hormones can lead to symptoms like low sex drive and impotence in men and infertility, and loss of normal menstrual cycles in women.
- Reduction in TSH can lead to hypothyroidism which can cause symptoms such as weight gain and fatigue.
- Reduction in ACTH can cause adrenal insufficiency. Symptoms include fatigue, low blood pressure, and electrolyte abnormalities.
- Reduction of growth hormone can cause delayed puberty and stunted growth in children. In adults symptoms are more subtle such as decreased muscle strength and fatigue.

Diagnosis

In order to know what type of tumour is in the pituitary gland, a medical history, physical exam, and many tests must be done. Tests may include:

- Blood tests to measure levels of hormones
- MRI scan
- Visual field testing

Tumour Pathology

Pituitary tumours removed during surgery are examined by a pathologist, a doctor who specializes in looking at tumour cells under a microscope. Pathologists often provide information that helps other doctors know what treatments and tests are best for people with pituitary disease. The pathologist will examine the sample of tumour tissue, and provide a report to your doctor. The “pathology report” describes the hormone content, structure, and the cells that caused your tumour. It usually takes about a week for your doctor to receive this report. All of this information is used to decide your tumour type, help predict the possible future effects of the tumour and decide what treatment will be needed.

Treatment Options

Treatment of pituitary tumours includes short and long-term management. The goals of treatment will change depending on which phase of management you are in. Treatment of a pituitary tumour depends on the hormonal activity of the tumour, the size and location of the tumour, as well as the age and overall health of the person with the tumour. The goals of treatment may be to remove the tumour, reduce or control the tumour size, and/or to re-balance hormone levels.

Surgery

If surgery is recommended for a pituitary tumour, the goal is to remove as much tumour as possible without damaging nearby structures or parts of the brain. A “trans-sphenoidal approach” (literally meaning “across the sphenoid sinus”) is the most common way pituitary surgery is done today because it is associated with fewer complications and faster recovery time. During trans-sphenoidal surgery, a neurosurgeon and an ENT (ear, nose and throat) surgeon work together. Small

instruments, microscopes and cameras are used to remove the tumour through the nose. Less often, a portion of the skull bone is temporarily removed to reach the pituitary gland. This type of surgery is called a craniotomy. Your neurosurgeon will speak with you about the surgery planned for your tumour, the risks and benefits of the procedure, and your follow-up care.

Radiation Therapy

Radiation therapy is sometimes given to people with pituitary tumours. Radiation oncologists are the doctors who are specialized in giving this type of treatment. The goal of radiation therapy is to reduce or control the tumour size. It may take several months or even longer to see the effects of radiation therapy.

There are different types of radiation therapy, including conventional or “standard radiation” and “stereotactic radiation”. At The Ottawa Hospital, CyberKnife radiation therapy is available. The CyberKnife system has a robotic arm, which can move around the patient to deliver radiation to a tumour from any direction. This form of treatment may be recommended for some patients with pituitary tumours that cannot be easily treated by surgery.

Medications Used to Treat Pituitary Tumours

Sometimes medications can be used to treat pituitary disease. Endocrinologists, doctors specialized in hormone disorders, will select drug treatments based on hormone levels. Some pituitary tumours can be treated with medications that reduce extra hormones being made. These include:

- *Prolactinomas*: Dopamine agonists are used to treat prolactinomas. Bromocriptine (Parlodel), cabergoline (Dostinex) and quinoglidle (Norprolac) are the medications in this class. They can lower prolactin levels and shrink prolactinomas. Many people with prolactinomas need to be on this type of medication for a long time.

- *Growth hormone and TSH-secreting adenomas*: Somatostatin analogues, such as octreotide injections (Sandostatin, Sandostatin LAR, Lanreotide or Somatuline Autogel) are used for this type of tumour.
- *Growth hormone-secreting adenomas*: Pegvisomant (Somavert) injections can be used to decrease the effects of high growth hormone levels in people who have this type of tumour.

What Treatments Has My Doctor(s) Recommended?

What to Expect—Next Steps

First step after the diagnosis is deciding if surgery, medication treatment, or both are needed.

- **If surgery is planned**, you will have an appointment with your neurosurgeon to explain the surgical procedure, sign a consent form, plan a surgery date, and organize tests that need to be done before surgery. During this appointment, the surgery and any risks with the surgery will be explained to you. Before having pituitary surgery, you will also have seen an endocrinologist, ENT surgeon and ophthalmologist (eye specialist).
- **If only medication treatment** is needed and no surgery planned, an endocrinologist will start you on a medication.
- **If radiation therapy is planned**, you will see a radiation oncologist.

What Happens After Pituitary Surgery?

After pituitary surgery at The Ottawa Hospital, you will be cared for in the Neurosciences Acute Care Unit (NACU). The function of your pituitary gland will be monitored carefully by doing frequent blood tests and monitoring your fluid intake and urine production after surgery. You will receive intravenous fluids and medications during your hospital stay. You may see doctors from the neurosurgery, endocrinology, and ENT surgery teams regularly in the hospital. On average, people are able to go home 3 to 5 days after pituitary surgery. Blood tests should be done to check hormone levels again after you leave the hospital. Appointments will be made for you to see your doctors again a few weeks after surgery.

What to expect in the Neurosciences Acute Care Unit:

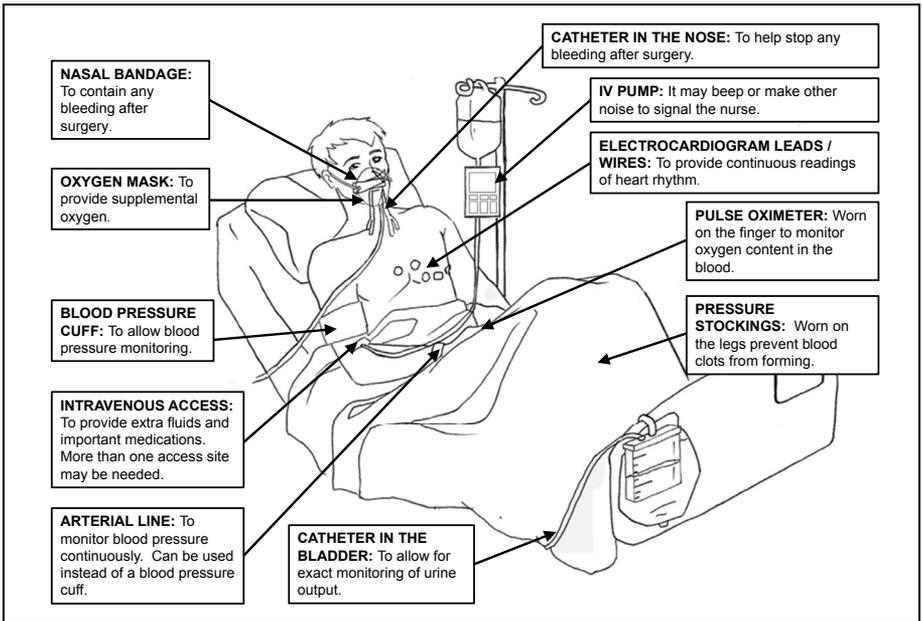


Illustration by Dr. Yingwei Liu

Possible Complications After Surgery

Complications may occur after pituitary surgery. Your surgeon will discuss these with you prior to your surgery. These may include infection, patch leak, spinal fluid leak, bleeding, stroke, risk of damage to the nerves of the eye, and hypopituitarism.

Limitations and expectations post surgery

After surgery you can expect to have a blocked nose and possibly a headache. You will not be able to drink through a straw or lift more than 10 pounds for a few weeks. You will be advised to avoid sneezing if possible. You may feel fatigued and lose some of your sense of taste and smell for a few weeks after surgery.

Long-Term Follow-up – What to Expect Next

There are three important types of tests that patients with pituitary tumours should have regularly:

1. Monitoring the size of the tumour with MRI scan.
2. Checking hormone levels with blood tests.
3. Vision tests.

Some patients with pituitary disease need to take medications for the rest of their lives. Medications may be needed if the pituitary gland does not make enough hormones or if there is some tumour left in the pituitary that is making extra hormones.

People with pituitary tumours usually need to have regular appointments with an endocrinologist, neurosurgeon and ophthalmologist. These specialists will advise you on how often testing is needed, if medications need to be changed, or if surgery or radiation therapy is needed to treat a growing tumour.

Take Charge of your health by being part of the team

You are the most important part of your healthcare team. You can stay healthy and feel some sense of control by making sure you:

- Understand the reason for your treatment
- Ask questions to help you fully understand
- Report new symptoms or concerns to your health-care team
- Take medication as prescribed
- Attend all scheduled appointments and tests

Frequently Asked Questions

We hope that the information in this pamphlet helps you communicate better with the people who are caring for you. We encourage you to ask questions.

What kind of tumour do I have?

- What is the size of my tumour? (Is my tumour a microadenoma or a macroadenoma?)
- Is my tumour secreting hormones or is it non-secreting?
- Which hormones are affected?

What are the recommended treatments?

- Surgery – If so, when?
- Do I need any medications? What are the names and the doses?
- Will I need radiation therapy?

Who is on my medical team?

- Neurosurgeon
- Endocrinologist
- Ear Nose and Throat (ENT) surgeon
- Ophthalmologist

Who do I call if?

- I need to reschedule an appointment:
- I am experiencing problems with the medication that was prescribed:
- I have experienced complications from my surgery after I have been discharged home:

Are there other resources that I can access?

Notes

Glossary

ACTH (Adrenocorticotrophic hormone): A hormone produced by the pituitary gland that stimulates the adrenal glands to make cortisol.

Acromegaly: A rare disease in adults caused by too much growth hormone. It is caused by a tumour of the pituitary gland.

Adrenal Glands: Glands situated just above each of the kidneys. They produce essential hormones such as cortisol and aldosterone.

Anterior pituitary (adenohypophysis): The front part of the pituitary that makes hormones that control important functions in the body, such as growth, reproduction, stress, metabolism, and lactation.

Cortisol: A hormone produced by the adrenal glands in response to stress.

Craniopharyngioma: A brain tumour that arises from embryonic cells in the pituitary gland. They are most common in children, but can occur rarely in adults.

Cushing's Disease: A disease caused by overproduction of cortisol caused by a tumour in the pituitary gland.

CyberKnife: A method of giving radiation therapy that targets disease in a precise location more accurately than standard radiation therapy.

Diabetes Insipidus: A disease caused by not enough antidiuretic hormone that may result in a water imbalance. Common symptoms are thirst and frequent urination.

Dopamine Agonists: Medications used to treat certain pituitary tumours, particularly prolactinomas.

Endocrinologist: A doctor who specializes in treating diseases of the glands and changes in hormone levels.

FSH (Follicle Stimulating Hormone): A hormone produced by the pituitary gland that stimulates ovaries in women and testes in men.

Glucocorticoid: A medication that has effects similar to cortisol (for example, prednisone, hydrocortisone, dexamethasone).

Transphenoidal Surgery: A surgery that involves approaching the pituitary gland through the nose or upper lip and the sphenoid sinus. It avoids the need to go through the upper skull.

TSH (Thyroid stimulating hormone): A hormone produced by the pituitary gland that stimulates the thyroid to produce thyroid hormone.

Hydrocortisone: The drug name of cortisol when it is made into a tablet or injection.

Hypothalamus: The part of the brain that controls the pituitary gland.

Hypopituitarism: The pituitary is no longer able to make the hormones required for function.

LH (Luteinizing Hormone): A hormone produced by the pituitary gland that stimulates the ovaries in women and the testes in men.

MRI (Magnetic Resonance Imaging): A medical imaging technique that uses magnetic fields and radio waves to make images of the body.

Optic Chiasm: The part of the brain where the optic nerves (nerves to the eyes) cross. It is located at the bottom of the brain, just above the pituitary gland.

Pathologist: A doctor who specializes in looking at tumour cells under a microscope.

Pathology Report: A report prepared by the Pathologist that describes the hormone content, structure and the cells that gave rise to your tumour.

Pituitary Adenoma: A non malignant tumour of the pituitary gland.

Posterior Pituitary: The back portion of the pituitary gland that makes antidiuretic hormone (a hormone that controls water balance) and oxytocin (another hormone signal made in the brain).

Prolactin: A hormone produced by the pituitary gland that causes breast milk production.

Prolactinoma: A pituitary tumour that produces prolactin, a hormone that causes breast milk production.

Rathke's Cleft Cyst: A benign fluid filled growth on the pituitary gland.

Somatostatin Analogues: Medications used to treat certain pituitary tumours, particularly growth hormone or TSH-secreting tumours.

Sphenoid Sinus: An air-filled bone cavity between the nasal cavity (nose) and the pituitary gland.

Visual Field Testing: An eye test that is used to look for problems in central and peripheral vision.

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. Please note that this publication is not a recommendation of any specific product, treatment plan, physician, or hospital.