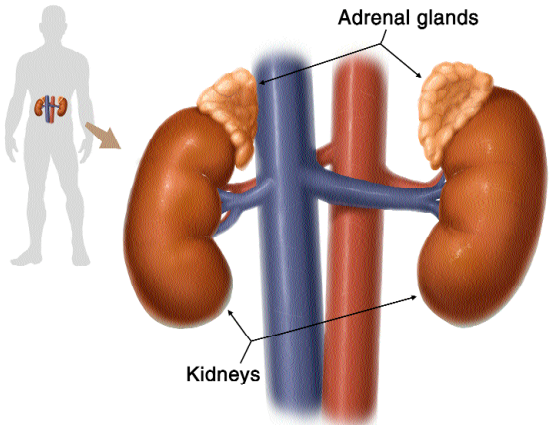
Pheochromocytoma

What is a pheochromocytoma?

A pheochromocytoma (fee ● o ● kromo ● sy ● toma) is a tumor arising most commonly from the adrenal gland. These tumors can cause headaches, sweating, rapid rise and fall in blood pressure, fast heartbeat, and other symptoms.

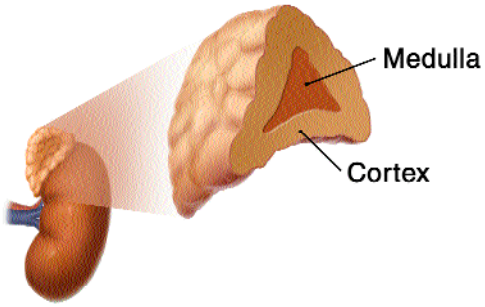
What is the adrenal gland?

An adrenal gland is a small, orange gland. There are two adrenal glands in the human body: one on top of each kidney.



The center of the adrenal gland is called the “medulla.” The outer part of the gland is called the “cortex.”

The medulla makes adrenaline (and adrenaline-like chemicals called catecholamines). The body needs adrenaline to maintain blood pressure and to help cope with stressful situations.



The adrenal cortex makes other hormones (cortisol and aldosterone) that maintain the body's fluid and electrolyte balance.

Eighty percent of the tumors are found on one adrenal gland (unilateral). Ten percent are found on both adrenal glands (bilateral), and 20 percent are found outside the adrenal glands.

This disorder can be inherited, as in von Hippel-Lindau syndrome and neurofibromatomatosis type 1, or it can occur with other endocrine tumors such as multiple endocrine neoplasia type 2 (MEN-2) or familial paraganglioma syndrome caused by succinate dehydrogenase gene mutation (e.g. SDHB). About 80 percent of pheochromocytomas are benign; 20 percent are malignant.

What are the signs and symptoms of a pheochromocytoma?

Symptoms include headache (usually severe), excessive sweating, generalized racing of the heart (tachycardia, palpitations), anxiety/ panic attacks, nervous shaking, tremors, nausea, vomiting, weight loss, pain in the lower chest and upper abdomen, weakness, fever, heat intolerance, sugar intolerance, and sometimes low blood pressure when standing up.

When do they occur?

Signs and symptoms usually follow a pattern of high blood pressure followed by low blood pressure. High blood pressure usually begins with a change in breathing and a pounding or forceful heartbeat. This may occur several times a week and last for up to 15 to 60 minutes. Often, these episodes of high blood pressure are started by activities that press on the tumor, such as changes in position, exercise, lifting, defecation, or emotional distress or anxiety.

How are pheochromocytomas diagnosed?

Reliable levels of adrenal hormones (adrenaline and noradrenaline) and their breakdown products (metanephrines) through blood or urine can usually help your doctor make a diagnosis.

There are three ways your doctor can do this: blood tests, urine tests, and x-ray tests such as computed tomography (CT), magnetic resonance imaging (MRI), and metaiobenzylguanidine (MIBG) scintigraphy.

Blood tests

Special blood tests for pheochromocytomas measure how much adrenaline or noradrenaline and their breakdown products (metanephrines) are in the blood or urine. Further tests include the glucagon stimulation test and clonidine suppression test.

Glucagon stimulation test

Because glucagon causes patients with pheochromocytomas to have symptoms of the disorder, this test is used for patients who have occasional signs and symptoms. Blood samples are drawn at specific times to measure adrenaline and noradrenaline levels. During this test, glucagon is injected into a vein while blood pressure and heart rate are monitored. This test takes about 30 minutes.

Clonidine suppression test

The clonidine suppression test also involves measuring adrenaline and noradrenaline as well as metanephrines in the blood over time. Clonidine normally lowers blood levels of catecholamines, but if a tumor is present, catecholamine levels do not decrease when clonidine is given. For this test, patients are asked to swallow a tablet of clonidine, and over the next 3 hours, blood samples are taken. Blood pressure and heart rate are checked during this time period.

24-hour urine collection

The 24-hour urine collection is also used to measure catecholamines and their breakdown products. High concentrations of these show a positive diagnosis.

Imaging: X-Ray and Nuclear Medicine Scans

Computed tomography

Computed tomography (CT) is a type of x-ray scan that uses a computer to make pictures, like “slices” of the inside of a part of your body.

Magnetic resonance imaging

MRI (magnetic resonance imaging) uses magnetic waves to make these pictures. These scans can help locate the tumor by showing the adrenal glands in great detail.

MIBG scan

MIBG is a type of scan that uses a radioactive compound to find the tumor. For this test, the patient receives a compound (I-123 or I-131 metaiodobenzylguanidine) through a vein. The scanner records the radiation given off by the compound, and when the compound has been taken up by the tumor, the camera will show the tumor's location.

The thyroid gland is sensitive to iodine, and the compounds used for the MIBG scan are iodine-based. To protect your thyroid during this scan, you will be given 100 milligrams of potassium iodine (SSKI). You must take SSKI 24 hours before the MIBG scan and for several days after it. Even if you take SSKI, your thyroid gland could be affected for a short period of time (hypothyroidism). Report any prolonged feelings of fatigue, temperature irregularities, or changes in your heartbeat to your doctor. These could be signs that your thyroid is not working properly (hypothyroidism).

Selective vena cava sampling

This type of blood drawing can be performed when blood tests and scans cannot find the tumor. For this test, a catheter is inserted into a major blood vessel so that blood samples can be taken from veins that supply organs in the neck, chest, abdomen, or pelvis. These samples are tested for levels of adrenaline. High levels of catecholamines pinpoint the tumor's location.

Treatment

Pheochromocytoma can be treated by medications that lower blood pressure and/or surgery to remove the tumor.

Medications

Alpha-adrenergic blockers are medications commonly used for lowering blood pressure (examples: phenoxybenzamine or prazosin). Propanolol/Inderol, a beta-blocker, may be used for controlling a fast and irregular pulse. Beta-blockers are used once the blood pressure comes back to normal with alpha-blockers. If you are being treated with medications, frequent followup with your doctor is crucial for making sure your blood pressure and heartbeat stay normal.

Surgery

Ninety percent of patients are cured by surgery. Surgery for suspected tumors is usually done by laparoscopy (a small incision or cut into the abdomen). This incision allows the doctor to examine other organs

while removing the tumor. Some surgeons prefer to make this incision from the back (posterior); others make the incision from the side (flank) when taking out larger tumors.

Once the tumor is removed, blood pressure usually falls to normal or low normal. Patients who have blood pressure that stays too low, or who have poor circulation in the arms and feet, may need transfusions of blood, plasma, or other fluids.

After surgery, some patients have a fall in blood pressure followed by a rise in blood pressure. Usually blood pressure returns to normal over the next few weeks. Patients with chronic high blood pressure will need treatment with alpha- and/or beta-blockers.

Sometimes surgery is not an option because of the type of tumor growth or because the tumor spreads (metastasizes) to other parts of the patient's body. Current treatments for malignant tumors include chemotherapy, or radioactive MIBG.

These treatments show variable success rates. If the tumor has spread, the usual sites of metastases are the bones and the lymph nodes. Tumors in bone tend to respond well to radiation therapy. A combination of chemotherapy drugs (cyclophosphamide, vincristine, and dacarbazine-CVD), has worked to control tumors in soft tissues (lymph nodes). For patients in whom surgery is not successful, or for those who cannot undergo surgery, symptoms are controlled with medications.

If you have other questions about pheochromocytoma, please feel free to ask your nurse or doctor.

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Questions about the Clinical Center?
http://www.cc.nih.gov/comments.shtml

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