CONDITION

Pheochromocytoma

Pheochromocytoma is a tumor of the adrenal gland that can release hormones causing episodes of high blood pressure.

WHAT IT IS
Hormone-producing tumor arising in an adrenal gland

BODY SYSTEM
Hormone system, adrenal glands, heart and circulation

MAIN SYMPTOMS
Headache, sweating, pounding heartbeat, high blood pressure

WHAT CAUSES IT
Usually unknown; sometimes inherited gene changes

HOW IT IS DIAGNOSED
Blood or urine metanephrines, followed by imaging

MAIN TREATMENT
Surgical removal after careful medicine preparation

CAN IT BE CURED?
Sometimes - surgery can remove localized tumors

IS IT AN EMERGENCY?
Sometimes - severe symptoms may signal a hormone crisis

LONG-TERM OUTLOOK
Often good after removal; recurrence requires long-term checks

Overview

A pheochromocytoma is a tumor that starts in an adrenal gland. You have two adrenal glands, one above each kidney. They make hormones that help control blood pressure, heart rate, and the body's response to stress.

Most pheochromocytomas release hormones called catecholamines, mainly adrenaline and noradrenaline. These hormones can cause sudden episodes of high blood pressure, a pounding heartbeat, sweating, and headache. Some tumors release hormones continuously, while others release them in bursts.

A closely related tumor outside an adrenal gland is called a paraganglioma. The two conditions are often discussed together because they arise from similar cells and are tested and treated in similar ways.

Pheochromocytomas are important because excess catecholamines can place strain on the heart and blood vessels. The tumor may also grow or, less commonly, spread to other parts of the body. With careful preparation, many tumors can be removed by surgery.

Symptoms

Symptoms may come in sudden spells lasting minutes to hours. In other people, symptoms are more constant. Some pheochromocytomas cause no clear symptoms and are found during imaging for another reason.

Common symptoms include:

The combination of headache, sweating, and a pounding heartbeat is characteristic, but it is not unique to pheochromocytoma. More common conditions, including panic attacks, thyroid problems, medication effects, and ordinary high blood pressure, can cause similar symptoms.

Episodes may happen without an obvious reason. They can sometimes be triggered by physical exertion, emotional stress, surgery, anesthesia, childbirth, pressure on the tumor, or certain medicines. Eating may trigger symptoms in some people with particular tumor types.

- Headache
- Heavy sweating
- A fast, forceful, or irregular heartbeat
- High blood pressure, either persistent or episodic
- Shaking, anxiety, or a sense of panic
- Pale skin
- Chest or abdominal discomfort
- Nausea, weakness, or light-headedness
- Unexplained weight loss

Causes and risk factors

A pheochromocytoma develops when specialized adrenal cells, called chromaffin cells, begin growing into a tumor. It is usually not clear why this happens.

Some pheochromocytomas are linked to an inherited gene change. These changes can be passed through families and may also increase the chance of paragangliomas or other tumors.

Associated conditions include hereditary paraganglioma-pheochromocytoma syndromes, multiple endocrine neoplasia type 2, von Hippel-Lindau disease, and neurofibromatosis type 1.

An inherited cause is more likely when:

However, an inherited change can be present without an obvious family history. Genetic counseling and testing are therefore often discussed after diagnosis. A genetic counselor can explain what a result may mean for you and your relatives.

- The tumor develops at a younger age
- There are tumors in both adrenal glands
- There is more than one tumor
- A paraganglioma is also present
- The tumor has spread
- Close relatives have related tumors or a known gene change

Diagnosis

Diagnosis usually begins with tests that look for the breakdown products of catecholamines. These products, called metanephrines, remain in the body longer than adrenaline and noradrenaline and are easier to measure reliably.

Testing commonly uses either:

Preparation matters because stress, body position, illness, caffeine, nicotine, and some medicines can affect results. Do not stop a prescribed medicine on your own. The team arranging the test can explain whether any preparation or medication review is needed.

A mildly abnormal result does not always mean there is a tumor. Testing may need to be repeated under controlled conditions. Strongly abnormal results usually lead to imaging to locate the tumor.

CT or MRI scans can show the adrenal glands and surrounding areas. Specialized scans may be used when there are multiple tumors, an inherited syndrome, uncertain findings, or concern that the tumor has spread. These scans use substances taken up by tumor cells to show where they are in the body.

A needle biopsy is generally not used to diagnose a suspected pheochromocytoma. Disturbing the tumor can release catecholamines and cause a dangerous rise in blood pressure. Biopsy also cannot reliably determine how the tumor will behave.

- A blood test for plasma free metanephrines
- A timed urine collection for fractionated metanephrines

Treatment and management

Surgery to remove the affected adrenal tumor is the main treatment when the pheochromocytoma is localized and can be removed safely.

Before surgery, hormone effects must be controlled. This usually involves an alpha-blocking medicine, which relaxes blood vessels and reduces the risk of severe blood pressure changes during the operation. The care team may also recommend adjustments to fluid and salt intake.

If a fast heartbeat continues, a beta blocker may sometimes be added after adequate alpha blockade. A beta blocker like metoprolol or propranolol should not be started first in a person with an active pheochromocytoma because it can allow blood pressure to rise dangerously. Preparation is normally managed by clinicians experienced with these tumors.

Many tumors can be removed using keyhole surgery. A larger operation may be needed for a very large tumor, suspected local invasion, or other complex findings. If both adrenal glands are affected, the surgical plan will consider whether some adrenal tissue can safely be preserved.

After surgery, blood pressure and blood glucose are watched closely. Hormone tests are repeated to check whether the source of excess catecholamines has been removed. Some people continue to have high blood pressure because of another cause, even when the operation is successful.

When a tumor cannot be completely removed or has spread, treatment focuses on controlling hormone production and slowing tumor growth. Options may include surgery for selected tumors, radiopharmaceutical treatment that delivers radiation to tumor cells, external radiation, chemotherapy, or targeted medicines. The choice is based on the tumor's location, scan findings, gene changes, symptoms, and rate of growth.

Medicines commonly used for this include propranolol . Which one suits you, if any, is a decision for you and your doctor.

Complications

Large or sudden catecholamine releases can cause a pheochromocytoma crisis. This may involve extreme or rapidly changing blood pressure, abnormal heart rhythms, heart failure,

stroke, organ injury, or shock. It requires emergency treatment.

Long-term hormone exposure can damage the heart muscle. This is called cardiomyopathy and may cause breathlessness, swelling, weakness, or chest discomfort. Heart function can improve after the hormone source is controlled, although recovery is not guaranteed.

Some pheochromocytomas return after surgery. A tumor may recur in the adrenal area, develop elsewhere, or spread to organs such as bone, liver, lungs, or lymph nodes. The appearance of tumor cells under a microscope cannot always predict this behavior. Spread to a site where these cells are not normally found is what confirms metastatic disease.

Surgery may also reduce adrenal hormone production, especially if both adrenal glands are removed. In that situation, lifelong adrenal hormone replacement may be needed.

Prevention

There is no established way to prevent a pheochromocytoma from developing.

Inherited gene changes cannot be prevented. If you carry one of these changes, planned screening may find a tumor before it causes major symptoms. Genetic counseling can also help relatives understand whether testing is relevant to them.

For someone with a known tumor, medical preparation before surgery or anesthesia helps prevent severe hormone-related complications. Make sure healthcare teams know about the diagnosis before any planned procedure. Do not try to prevent episodes by stopping medicines or making major dietary changes without medical advice.

Living with it and what to expect

The months around diagnosis may involve repeat hormone tests, several scans, medicine preparation, and surgery. Blood pressure can change as treatment begins, so regular measurements and medication adjustments may be needed.

After complete removal, hormone-related spells often stop and blood pressure may improve. Some people still need blood pressure treatment because they also have ordinary hypertension. Recovery from surgery depends on the operation, the tumor's size, and your general health.

Follow-up does not end when symptoms improve. Repeat blood or urine testing checks for persistent or returning hormone production. Follow-up is usually long term and may be lifelong for people with an inherited gene change, multiple tumors, a paraganglioma, or other features

linked to recurrence.

A genetic diagnosis may lead to screening for other related tumors. It may also have implications for parents, siblings, and children. Genetic counseling can help you decide how and when to discuss testing with relatives.

If the tumor has spread, the condition may be managed over years with a combination of hormone-blocking medicines, scans, and treatments directed at growing tumors. Some metastatic tumors grow slowly, while others behave more aggressively. Your follow-up plan will be based on what the tumor is doing over time rather than on one scan alone.

When to get medical help

Arrange a routine medical appointment if you have repeated episodes of headache, sweating, and a pounding heartbeat, especially if blood pressure is high during the episodes. These symptoms have many possible causes, so they need assessment rather than self-diagnosis.

If you have been treated for pheochromocytoma, contact your follow-up team about returning spells, new persistent pain, unexplained weight loss, or changes in blood pressure. These symptoms do not necessarily mean the tumor has returned, but they should be reviewed.

Seek urgent medical advice if you have a known or suspected pheochromocytoma and develop repeated severe symptoms, very high blood pressure readings, or a new irregular heartbeat.

Call emergency services for:

Do not drive yourself if you may be having a heart, stroke, or blood pressure emergency.

- Chest pain, severe breathlessness, or fainting
- Sudden weakness, facial drooping, confusion, or trouble speaking
- A severe sudden headache with neurological symptoms
- Seizure or loss of consciousness
- Severe symptoms with extreme or rapidly changing blood pressure

Common questions

Is a pheochromocytoma cancer?

Not every pheochromocytoma spreads. However, it is difficult to label one as harmless based only on how it looks under a microscope. A tumor is considered metastatic when it is found in

places where these adrenal-type cells do not normally occur, such as bone or the liver.

Can pheochromocytoma be cured?

A localized tumor can often be removed completely with surgery. Cure is less certain if the tumor cannot be fully removed or has spread. Even after successful surgery, long-term testing is important because recurrence can happen.

Does pheochromocytoma always cause high blood pressure?

No. Blood pressure may be continuously high, rise only during episodes, or occasionally remain within the usual range. A normal reading between attacks does not rule it out, but symptoms alone cannot confirm it.

Can stress cause pheochromocytoma?

Stress does not appear to create the tumor. Physical or emotional stress can trigger hormone release from an existing tumor, which may bring on symptoms.

Are there foods or medicines I need to avoid?

Some medicines, supplements, stimulants like methylphenidate or lisdexamfetamine , or foods may worsen symptoms or interfere with testing in certain situations. The relevant list differs according to the test and treatment stage. Do not stop prescribed medicines without instructions from the team arranging your care.

Can my children inherit it?

They may be at risk if your tumor is linked to an inherited gene change. The chance depends on the specific gene. If no inherited change is found, the diagnosis does not automatically mean your children are at increased risk.

Sources

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