NIH NATIONAL CANCER INSTITUTE

# Pheochromocytoma and Paraganglioma Treatment (PDQ®)–Patient Version

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## General Information About Pheochromocytoma and Paraganglioma

### KEY POINTS

- Pheochromocytoma and paraganglioma are rare tumors that come from the same type of tissue.
- Pheochromocytoma is a rare tumor that forms in the adrenal medulla (the center of the adrenal gland).
- Paragangliomas form outside the adrenal gland.
- Certain inherited disorders and changes in certain genes increase the risk of pheochromocytoma or paraganglioma.
- Signs and symptoms of pheochromocytoma and paraganglioma include high blood pressure and headache.
- Signs and symptoms of pheochromocytoma and paraganglioma may occur at any time or be brought on by certain events.
- Tests that examine the blood and urine are used to detect (find) and diagnose pheochromocytoma and paraganglioma.
- Genetic counseling is part of the treatment plan for patients with pheochromocytoma or paraganglioma.
- Certain factors affect prognosis (chance of recovery) and treatment options.

### Pheochromocytoma and paraganglioma are rare tumors that come from the same type of tissue.

Paragangliomas form in nerve tissue in the adrenal glands and near certain blood vessels and nerves. Paragangliomas that form in the adrenal glands are called pheochromocytomas. Paragangliomas that form outside the adrenal glands are called extra-adrenal paragangliomas. In this summary, extra-adrenal paragangliomas are called paragangliomas.

Pheochromocytomas and paragangliomas may be benign (not cancer) or malignant (cancer).

### Pheochromocytoma is a rare tumor that forms in the adrenal medulla (the

### center of the adrenal gland).

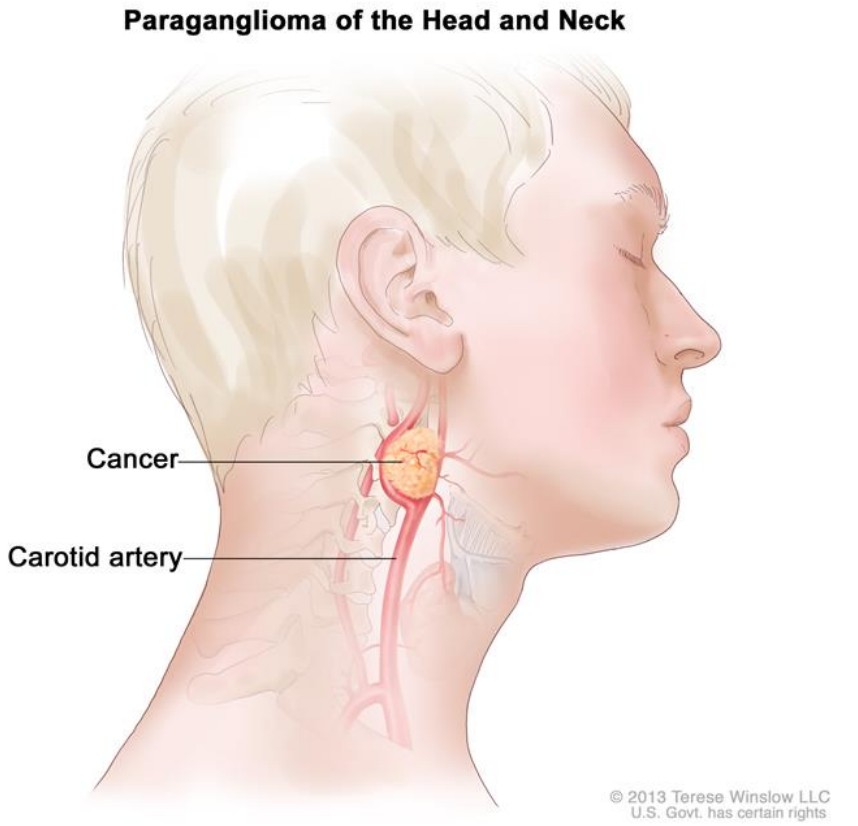
Pheochromocytoma forms in the adrenal glands. There are two adrenal glands, one on top of each kidney in the back of the upper abdomen. Each adrenal gland has two parts. The outer layer of the adrenal gland is the adrenal cortex. The center of the adrenal gland is the adrenal medulla.

Pheochromocytoma is a rare tumor of the adrenal medulla. Usually, pheochromocytoma affects one adrenal gland, but it may affect both adrenal glands. Sometimes there is more than one tumor in one adrenal gland.

The adrenal glands make important hormones called catecholamines. Adrenaline (epinephrine) and noradrenaline (norepinephrine) are two types of catecholamines that help control heart rate, blood pressure, blood sugar, and the way the body reacts to stress. Sometimes a pheochromocytoma will release extra adrenaline and noradrenaline into the blood and cause signs or symptoms of disease.

### Paragangliomas form outside the adrenal gland.

Paragangliomas are rare tumors that form near the carotid artery, along nerve pathways in the head and neck, and in other parts of the body. Some paragangliomas make extra catecholamines called adrenaline and noradrenaline. The release of these extra catecholamines into the blood may cause signs or symptoms of disease.



**Paraganglioma of the head and neck. A rare tumor that often forms near the carotid artery. It may also form along nerve pathways in the head and neck and in other parts of the body.**

## Certain inherited disorders and changes in certain genes increase the risk of pheochromocytoma or paraganglioma.

Anything that increases your chance of getting a disease is called a risk factor. Having a risk factor doesn't mean that you will get cancer; not having risk factors doesn’t mean that you will not get cancer. Talk to your doctor if you think you may be at risk.

The following inherited syndromes or gene changes increase the risk of pheochromocytoma or paraganglioma:

- Multiple endocrine neoplasia 2 syndrome, types A and B (MEN2A and MEN2B).
- von Hippel-Lindau (VHL) syndrome.
- Neurofibromatosis type 1 (NF1).
- Hereditary paraganglioma syndrome.
- Carney-Stratakis dyad (paraganglioma and gastrointestinal stromal tumor [GIST]).
- Carney triad (paraganglioma, GIST, and pulmonary chondroma).

## Signs and symptoms of pheochromocytoma and paraganglioma include high blood pressure and headache.

Some tumors do not make extra adrenaline or noradrenaline and do not cause signs and symptoms. These tumors are sometimes found when a lump forms in the neck or when a test or procedure is done for another reason. Signs and symptoms of pheochromocytoma and paraganglioma occur when too much adrenaline or noradrenaline is released into the blood. These and other signs and symptoms may be caused by pheochromocytoma and paraganglioma or by other conditions. Check with your doctor if you have any of the following:

- High blood pressure.
- Headache.
- Heavy sweating for no known reason.
- A strong, fast, or irregular heartbeat.
- Being shaky.
- Being extremely pale.

The most common sign is high blood pressure. It may be hard to control. Very high blood pressure can cause serious health problems such as irregular heartbeat, heart attack, stroke,

or death.

## Signs and symptoms of pheochromocytoma and paraganglioma may occur at any time or be brought on by certain events.

Signs and symptoms of pheochromocytoma and paraganglioma may occur when one of the following events happens:

- Hard physical activity.
- A physical injury or having a lot of emotional stress.
- Childbirth.
- Going under anesthesia.
- Surgery, including procedures to remove the tumor.
- Eating foods high in tyramine (such as red wine, chocolate, and cheese).

## Tests that examine the blood and urine are used to detect (find) and diagnose pheochromocytoma and paraganglioma.

The following tests and procedures may be used:

- **Physical exam and history** : An exam of the body to check general signs of health, including checking for signs of disease, such as high blood pressure or anything else that seems unusual. A history of the patient's health habits and past illnesses and treatments will also be taken.
- **Twenty-four-hour urine test**: A test in which urine is collected for 24 hours to measure the amounts of catecholamines in the urine. Substances caused by the breakdown of these catecholamines are also measured. An unusual (higher or lower than normal) amount of a substance can be a sign of disease in the organ or tissue that makes it. Higher-than-normal amounts of certain catecholamines may be a sign of pheochromocytoma.
- **Blood catecholamine studies**: A procedure in which a blood sample is checked to measure the amount of certain catecholamines released into the blood. Substances caused by the breakdown of these catecholamines are also measured. An unusual (higher than or lower than normal) amount of a substance can be a sign of disease in the organ or tissue that makes it. Higher-than-normal amounts of certain catecholamines may be a sign of pheochromocytoma.
- **CT scan (CAT scan)**: A procedure that makes a series of detailed pictures of areas inside the body, such as the neck, chest, abdomen, and pelvis, taken from different angles. The pictures are made by a computer linked to an x-ray machine. A dye may be injected into a vein or swallowed to help the organs or tissues show up more clearly. This procedure is also called computed tomography, computerized tomography, or computerized axial tomography.
- **MRI (magnetic resonance imaging)**: A procedure that uses a magnet, radio waves, and

a computer to make a series of detailed pictures of areas inside the body such as the neck, chest, abdomen, and pelvis. This procedure is also called nuclear magnetic resonance imaging (NMRI).

## Genetic counseling is part of the treatment plan for patients with pheochromocytoma or paraganglioma.

All patients who are diagnosed with pheochromocytoma or paraganglioma should have genetic counseling to find out their risk for having an inherited syndrome and other related cancers.

Genetic testing may be recommended by a genetic counselor for patients who:

- Have a personal or family history of traits linked with inherited pheochromocytoma or paraganglioma syndrome.
- Have tumors in both adrenal glands.
- Have more than one tumor in one adrenal gland.
- Have signs or symptoms of extra catecholamines being released into the blood or malignant (cancerous) paraganglioma.
- Are diagnosed before age 40.

Genetic testing is sometimes recommended for patients with pheochromocytoma who:

- Are aged 40 to 50 years.
- Have a tumor in one adrenal gland.
- Do not have a personal or family history of an inherited syndrome.

When certain gene changes are found during genetic testing, the testing is usually offered to family members who are at risk but do not have signs or symptoms.

Genetic testing is not recommended for patients older than 50 years.

## Certain factors affect prognosis (chance of recovery) and treatment options.

The prognosis (chance of recovery) and treatment options depend on the following:

- Whether the tumor is benign or malignant.
- Whether the tumor is in one area only or has spread to other places in the body.
- Whether there are signs or symptoms caused by a higher-than-normal amount of catecholamines.
- Whether the tumor has just been diagnosed or has recurred (come back).

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