



ABOUT THIS GUIDE

All educational information is vetted by our Medical Advisory Board. We use current guidelines and relevant research. For more detailed information, visit pheopara.org.

SPECIAL THANKS TO

The Medical Advisory Board and the Patient Engagement Committee.

Get support

Order a new patient Pheo Para CareKit, register for Peer Navigation or monthly support groups for patients and caregivers, or find a multidisciplinary care center with expertise in pheochromocytoma and paraganglioma at pheopara.org.

pheopara.org

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MEDICALLY REVIEWED
BY THE PPA MEDICAL
ADVISORY BOARD ·
2026



SCAN TO VISIT

BY THE NUMBERS

Most pheo para tumors are successfully treated with surgery. Risk varies by subtype, location and genetics. Here's what the numbers show:

2–8

per 1 million people
diagnosed each year

up to 35%

of cases are hereditary

10–15%

of pheochromocytomas
become metastatic

40–70%

of extra-adrenal paras
become metastatic[†]

50%

chance of passing an
identified mutation to a child

[†] Head & neck paras are the exception,
rarely metastasizing (2–10%).

QUESTIONS TO ASK YOUR CARE TEAM

- Should I have genetic testing, and should my relatives?
- Is there a multidisciplinary center near me?
- Do I need to be "blocked*" before any procedure?
- What is our surveillance plan?

*blocked = receive drugs to block the effects
of catecholamines secreted by pheo para.

PATIENT GUIDE

Pheochromocytoma & Paraganglioma

Also known as pheo, para, or PPGL



You're not alone. The Pheo Para Alliance's global community is here with you, every step of the way.

If detected early, pheo para can be successfully treated in the majority of cases.

Pheo Para Alliance · pheopara.org

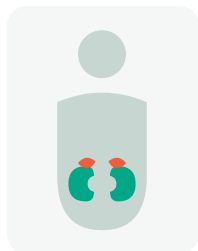
UNDERSTANDING THE DISEASE

What is pheo?

A pheochromocytoma is a rare neuroendocrine tumor in the adrenal medulla, the center of each adrenal gland, above the kidneys. Its behavior varies from person to person. Pheos usually produce excess catecholamines that drive the "fight or flight" response.

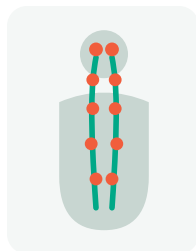
What is para?

Paraganglioma is a closely related tumor, forming outside the adrenal glands in the nervous system, at the base of the skull, neck, chest, abdomen, and pelvis. A person may have a pheo, a para, or both.



Pheo

adrenal glands, above the kidneys



Para

nerve pathways, skull base to pelvis

Head & neck paragangliomas

Head-and-neck paragangliomas are tumors that arise from specialized cells along nerves and blood vessels in the head and neck, including the carotid body, jugular, tympanic, and vagal sites. They are usually non-secreting and slow-growing and may be discovered as a painless mass. However, if they grow, they can affect adjacent structures, including nerves, and cause pain and other pressure symptoms. Symptoms can include hearing changes, hoarseness, or difficulty swallowing, depending on the tumor's location and size.

EPIDEMIOLOGY

Pheo para can occur at any age but most often appears between **20 and 50 years of age**. Children account for roughly **10–20%** of cases and are more likely to carry an inherited mutation.

SYMPTOMS

Excess catecholamines can cause persistent or episodic high blood pressure and other symptoms. See a doctor if you notice:

- High blood pressure
- Rapid or forceful heartbeat
- Profound sweating
- Severe headache
- Tremors
- Paleness in the face
- Shortness of breath
- Sensation of panic or doom
- Anxiety
- Abdominal pain / constipation
- Weight loss

This is not an exhaustive list, and symptoms vary from person to person; some people have no symptoms at all.

DIAGNOSIS

The gold standard is care at a **multidisciplinary center** with pheo para expertise, led by an endocrinologist (a hormone specialist). 24-hour urine and blood (metanephrine) tests confirm suspected cases; CT/MRI or functional imaging then locates and sizes the tumor(s). Biopsy is usually not recommended. Pheo is often called the **"great mimic"** because its symptoms overlap many other conditions. If you felt dismissed before your diagnosis, you're not alone.

BEFORE TESTING: THESE LABS DIFFER FROM ROUTINE

- For a blood draw, rest lying down 20–30 minutes first; a seated draw can cause false positives.
- Avoid caffeine, nicotine, and strenuous activity, and ask which medications to pause first — never stop a medication without your doctor's advice. Fasting and no sports overnight (about 12 hours) is recommended.
- For 24-hour urine, use the provided preservative container and keep it refrigerated.

TREATMENT

Surgery to remove the tumor(s) is usually the treatment of choice and typically restores normal blood pressure. Stable, non-secreting tumors may instead be monitored when surgery carries a disproportionately high risk, particularly head & neck paras involving cranial nerves. If surgery isn't possible, radiation, embolization, radionuclide, targeted, or chemotherapy may be used. After treatment, long-term monitoring is recommended; its frequency and duration are set with your care team, based on tumor type, genetic result, and your situation.

Before surgery, most patients must receive medication to block the effects of catecholamines, since anesthesia and tumor manipulation can otherwise trigger a hypertensive crisis.

Seek emergency care for a sudden, severe episode: an extreme blood pressure spike, chest pain, or a crushing headache with a racing heart. Tell responders you have pheo para.

GENETIC TESTING

Up to 35% of pheo and para are hereditary. The rest are **sporadic or have no identified subtype**: no known inherited mutation is found, though one may not have been identified yet. More than a dozen genes have been linked.

MOST COMMON

SDHB	SDHD	SDHC	SDHA	VHL
RET	NF1			

OTHERS LINKED TO PHEO PARA

SDHAF2	TMEM127	MAX	FH
EPAS1 (HIF2A)	EGLN1 (PHD2)	EGLN2	
KIF1B	MDH2	SLC25A11	DLST
GOT2			

WHY IT MATTERS

A genetic result guides how often you're screened and for what, and lets blood relatives who may carry the same mutation get tested early. Ask your care team about testing. Details at pheopara.org.