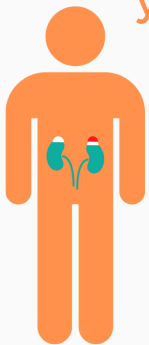


The illness can be referred to as pheochromocytoma (pheo), paraganglioma (para), pheo para or PPGL

what is pheo?

A pheochromocytoma (fee-o- kroe-moe-sy-TOE- muh) is a rare, usually non-cancerous (benign), slow-growing neuroendocrine tumor that develops in cells in the center of an adrenal gland, called the adrenal medulla. These two adrenal glands, one located above each kidney, produce hormones called catecholamines. Pheochromocytoma (pheo) typically produce an excess amount of catecholamine hormones, which include norepinephrine (noradrenaline), epinephrine (adrenaline), & dopamine. These hormones produce the "fight or flight" response.

about 2 to 8 per 1 million people are diagnosed each year with pheo para



Pheos form in the adrenal glands located above kidneys.

30-40% of pheos and paras are hereditary. If you are diagnosed talk to your doctor about genetic testing.

There is no test to determine if a pheo or para is malignant. The disease is cancerous if it has spread to other parts of the body. This happens in 15-25% of cases. It is more common for paras than for pheos to become malignant, except for paras in the head and neck. The likelihood that a pheo or para become malignant also depends on the type of genetic mutation, if one is present.

GENETIC TESTING

30-40% of pheos and paras are hereditary. There are more than a dozen genes that have been linked to pheo para. The most common are SDHA, SDHB, SDHC, SDHD, VHL, NF1 & RET. More information about each of these genes can be found in the genetics brochure and at pheopara.org.

If you have a genetic mutation, your children have a 50% chance of inheriting it. Even if your children inherit the gene, that doesn't mean that they will develop a tumor, but regular monitoring is important. Regular screening can catch tumors before you have symptoms.

All educational information is vetted by our Medical Advisory Board. We also utilize current guidelines and relevant research. For more detailed info go to pheopara.org. Printed 3/2020.

Special Thanks to...



Pheo Para Alliance Medical Advisory Board
pheopara.org / info@pheopara.org / @pheopara



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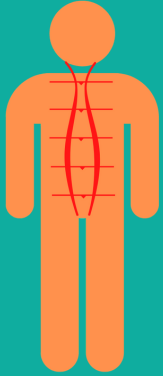


A GUIDE TO PHEOCHROMOCYTOMA & PARAGANGLIOMA

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

what is para?

Paraganglioma (pär'ə-gäng'glē-ō'mə) or para is a rare, usually noncancerous (benign), slow-growing tumor that is closely related to pheo, but even more rare. Paras are extra-adrenal which means they form outside of the adrenal glands, in the sympathetic and parasympathetic nervous system.



Para commonly form at the base of the skull, neck, chest, abdomen, and pelvis.

SYMPTOMS

Pheo para can occur at any age, but most commonly affect people between the ages of 20 and 50. If you experience the symptoms below, you should see a doctor to get an accurate diagnosis, as many of these symptoms can be caused by other conditions.

The release of catecholamines from pheos and paras can cause persistent or episodic high blood pressure, headache, sweating and other symptoms. If left untreated, life-threatening damage, especially to the cardiovascular system, can result.

Although it is rare, some para do not produce catecholamines so symptoms may not occur. These non-secreting paras are often found incidentally from imaging for another reason.

Symptoms of pheo para:

- 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

DIAGNOSIS

If pheo para is suspected, you should see an endocrinologist, (a doctor who specializes in hormonal disorders) who has pheo para experience. The twenty-four hour urine and blood (plasma) tests are used if pheo or para is suspected.

Pheo is often referred to as the "great mimic". Receiving a diagnosis and distinguishing pheo para from other conditions can be a challenge. In a recent survey, 35% of respondents indicated it took 4+ years to receive a diagnosis.

Imaging, taking pictures of the inside of the body, is often used once biochemical tests indicate a pheo or para. Imaging will help to identify where, how many, and size of the tumor(s). CT/MRI are often used, but functional imaging involving the use of a radioactive substance to take pictures of the body may also be used.

connecting with others who have pheo para can be comforting.

@pheopara

TREATMENT

If possible, the treatment of choice is surgery to remove the tumor(s). Surgical treatment usually returns blood pressure to normal and alleviates symptoms. Alternatively, an experienced doctor may suggest only regular monitoring of the tumor(s) if they are not secreting catecholamines, there are no symptoms and the tumor(s) are stable (not growing).

Radiation, chemotherapy or radionuclide therapy may be used if surgery is not possible. More information about these treatments can be found at pheopara.org.

Before undergoing surgery, patients must be adequately "blocked" with medication. Anesthesia and manipulation of the tumor(s) can cause the release of catecholamines during surgery, which may result in a hypertensive crisis and even death. This is why it is usually not recommended for patients to have a biopsy of the tumor if pheo para is suspected.

It is critical that patients are "blocked" with medication before undergoing surgery.

A urine test or plasma test should be repeated 4-8 weeks after surgery to check for any remaining disease. Urine or plasma tests should be performed annually to detect remaining disease, return of the disease, or the development of metastases.