

The two adrenal glands are located in the abdomen, one atop of each kidney, and produce a variety of hormones including adrenaline, aldosterone, and cortisol. Benign (noncancerous) adrenal tumors are relatively common in general population. However, a rare tumor of the adrenal gland called a pheochromocytoma (PCC) will secrete hormones into the bloodstream and can cause anxiety and uncontrollable high blood pressure.

A tumor that is similar to a PCC is called a paraganglioma (PGL). Unlike PCCs, PGLs are not limited to just the adrenal glands, and are commonly found in the head and neck region of the body. Adrenocortical carcinoma (ACC), or cancer of the adrenal cortex, is a rare type of adrenal cancer.

Compared with other types of tumors, PGLs and PCCs are very rare. However, the proportion of these rare tumors that are hereditary is much higher. As many as 25-40% of PCC and PGL tumors are considered to be due to a hereditary cause. Similarly, compared with other types of cancers, ACC is very rare. It is estimated that 10% of all cases of ACC are due to a hereditary cause. Children with ACC have a higher likelihood of having a hereditary cause than adults with ACC.

Because of the increased chance for these tumors and cancers to be hereditary, it is recommended that all patients consider genetic counseling regardless of what age they were diagnosed with PCC, PGL, or ACC, and whether or not they have a family history of cancer. There are currently at least a dozen genes that are known to contribute to PGL, PCC, and ACC, some of which overlap with other types of hereditary cancer syndromes. Some hereditary cancer syndromes that can include adrenal cancer are:

- [Multiple Endocrine Neoplasia, type 2 \(RET\)](#)
- [Li-Fraumeni syndrome \(TP53\)](#)
- [Von-Hippel Lindau syndrome \(VHL\)](#)
- [Neurofibromatosis, type 1 \(NF1\)](#)
- Carney complex (PRKAR1A)
- Hereditary paraganglioma-pheochromocytoma (SDHD, SDHAF2, SDHC, SDHB)

Click [here](#) to learn more about scheduling a genetic counseling appointment for questions about hereditary cancer predisposition.