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Clinician Management Resource for Hereditary Pheochromocytoma/Paraganglioma Syndrome

This overview of clinical management guidelines is based on this patient's positive test result. Unless otherwise stated, medical management guidelines used here are limited to those issued by the National Comprehensive Cancer Network (NCCN) in the U.S. Please consult the referenced guideline for complete details and further information.

Clinical correlation with the patient's past medical history, treatments, surgeries, and family history may lead to changes in clinical management decisions; therefore, other management recommendations may be considered. Genetic testing results and medical society guidelines help inform medical management decision but do not constitute formal recommendations. Discussions of medical management decisions and individualized treatment plans should be made in consultation between each patient and his or her healthcare provider and may change.

SCREENING CONSIDERATIONS[1, ^, *]	AGE TO START	FREQUENCY
Pheochromocytoma/Paraganglioma (PCC/PGL)-specific screening recommendations for patients with confirmed germline hereditary PCC/PGL syndrome		
Blood pressure monitoring.	6-10 years old for patients with pathogenic or likely pathogenic *SDHB* variants 10-15 years for patients with all other forms of hereditary PCC/PGL	At all medical visits
Measurement of plasma free metanephrines or 24-hour urine for fractionated metanephrines.	6-10 years old for patients with pathogenic or likely pathogenic *SDHB* variants 10-15 years for patients with all other forms of hereditary PCC/PGL	Annually
Cross-sectional imaging of skull base to pelvis. Whole body MRI or other non-radiation-containing imaging procedures. If whole body MRI not available, may consider abdominal MRI, skull base and neck MRI, and chest CT.**	6-10 years old for patients with pathogenic or likely pathogenic *SDHB* variants 10-15 years for patients with all other forms of hereditary PCC/PGL	Every 2–3 years

^ Patients with pathogenic *SDHD*, *SDHAF2*, or *MAX* variants are most at risk if the pathogenic variant was paternally inherited. Recommend following the above recommendations if the parent of origin is unknown. Consider screening for patients with maternally inherited variants as case reports of tumor occurrence exist.

* If asymptomatic and without a prior history of elevation, annual follow-up and testing can be omitted or done with imaging every 2-3 years. Since SDH genes have variability in their tumor penetrance and risk for malignancy, consideration can be given to modified screening intervals, especially for less penetrant genes such as *SDHA*.

** Available data suggest that patients with pathogenic *SDHAF2* variants are primarily at risk for head and neck tumors and patients with pathogenic *MAX* variants are primarily at risk for adrenal tumors. Therefore, consideration can be given to more targeted imaging in these cohorts.

1. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) for Neuroendocrine and Adrenal Tumors v1.2026. National Comprehensive Cancer Network, Inc. 2026. All rights reserved. Accessed April 28, 2026. To view the most recent and complete version of the guideline, go to NCCN.org. NCCN makes no warranties of any kind whatsoever regarding their content, use or application and disclaims any responsibility for their application or use in any way.

Understanding Your Positive *MAX* Genetic Test Result

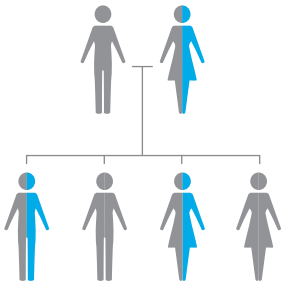
INFORMATION FOR PATIENTS WITH A PATHOGENIC OR LIKELY PATHOGENIC VARIANT

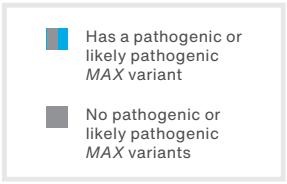
4 Things to Know

1	Result	Your testing shows that you have a pathogenic or likely pathogenic variant in the *MAX* gene.
2	Tumor risks	You have an increased chance to develop pheochromocytomas (PCCs), paragangliomas (PGLs), and possibly kidney tumors or neuroendocrine tumors (tumors that affect the nervous system or hormone-producing glands).
3	What you can do	Risk management decisions are very personal. There are options to detect cancer early or lower the risk to develop cancer. It is important to discuss these options with your healthcare provider and decide on a plan that works for you.
4	Family	Family members may also be at risk – they can be tested for the pathogenic or likely pathogenic *MAX* variant that was found in you. It is recommended that you share this information with your family members so they can learn more and discuss with their healthcare providers.

MAX in the Family

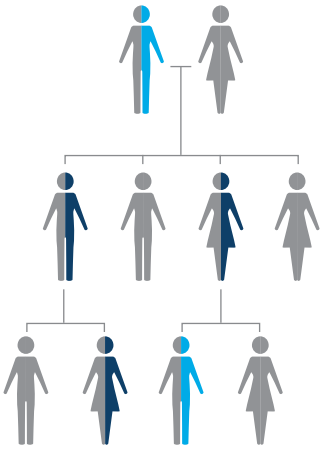
There is a 50/50 random chance to pass on the pathogenic or likely pathogenic *MAX* variant to each of your children.

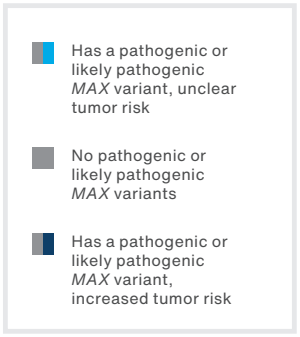




MAX Tumor Risk in the Family

Pathogenic and likely pathogenic *MAX* variants are equally inherited from either parent. Evidence suggests that the risk for tumors may be higher when the pathogenic or likely pathogenic *MAX* variant is inherited from your father. The image below demonstrates this.





RESOURCES

- Pheo Para Alliance pheopara.org
- National Society of Genetic Counselors nsgc.org
- Canadian Society of Genetic Counsellors cagc-accg.ca

Please discuss this information with your healthcare provider. The cancer genetics field is continuously evolving, so updates related to your *MAX* result, medical recommendations, and/or potential treatments may be available over time. This information is not meant to replace a discussion with a healthcare provider, and should not be considered or interpreted as medical advice.

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