What you need to know

MEN1 Gene

What does it mean to have a *MEN1* pathogenic variant?

Having a *MEN1* pathogenic variant, also known as a mutation, causes a condition called Multiple Endocrine Neoplasia Type 1 (MEN1) syndrome. A similar condition, called Multiple Endocrine Neoplasia Type 4 (MEN4) syndrome, is caused by pathogenic variants in the *CDKN1B* gene.

Do I have an increased risk for cancer if I have a *MEN1* pathogenic variant?

If you have a *MEN1* pathogenic variant, you have a greater risk of developing certain types of benign tumors. Benign tumors are not cancerous and do not spread whereas cancerous (also called malignant) tumors can spread (metastasize) to nearby healthy tissue and organs. Cancerous tumor cells also have the potential to spread to more distant sites of the body. Benign and malignant tumors are often treated in different ways.

While cancer can occur in people with MEN1 syndrome, the likelihood of developing a cancer due to a *MEN1* pathogenic variant is low.

What type of tumors am I at risk for if I have a *MEN1* pathogenic variant?

People with MEN1 syndrome have an increased risk of developing tumors in the endocrine system as compared to the general population. The endocrine system is made up of endocrine glands, which secrete hormones to control important functions in the body such as mood, growth and development, and metabolism.

People with MEN1 syndrome are at highest risk for the following endocrine diseases:

- **Parathyroid glands:** About 95% of people with MEN1 will develop a symptom called primary hyperparathyroidism by the age of 50. This means that the parathyroid glands are overactive, causing high calcium level in the bloodstream (hypercalcemia).
- **Pituitary gland:** A benign tumor occurs in the pituitary gland in about 30-40% of people with MEN1. This benign tumor can cause the pituitary gland to overproduce hormones or cause symptoms, such as vision changes or headaches.
- **Neuroendocrine tumors of the gastro-entero-pancreatic (GEP) tract:** 20-80% of people with MEN1 develop benign tumors in their pancreas or elsewhere in the glands of the gastrointestinal tract. Sometimes, these neuroendocrine tumors can become malignant (cancerous).

Other features of MEN1 include tumors in the thymus gland, lung, small intestine and stomach called carcinoids. Carcinoids are endocrine tumors and are most often benign, but some can become cancerous. People with MEN1 can also develop adrenocortical tumors.

If I have a *MEN1* pathogenic variant, what is the chance my family members will have it too?

There is a 50% chance that a person with a pathogenic variant will pass it on to each of their children. In most cases, siblings of a person with a pathogenic variant each have a 50% chance to have the pathogenic variant. Additionally, other family members are at risk to have the pathogenic variant.