

About *MEN1* Gene Mutations

About Genes

Genes are in every cell in our bodies. Genes are made of DNA, which gives instructions to cells about how to grow and work together. We have two copies of each gene in each cell—one from our mother and one from our father. When genes work right, they help stop cancer from developing. If one copy of a gene has a mutation or change, the gene doesn't work well and there is more risk of cancer.

When it works right, the *MEN1* gene controls how much certain tissues grow. When *MEN1* has a mutation, cells copy themselves too much and cancer can develop.

When someone inherits a copy of *MEN1* with a mutation, they have a syndrome called Multiple Endocrine Neoplasia Type 1 (*MEN1*).

People with *MEN1* have a high risk of getting non-cancerous (benign) tumors in part of the endocrine system, which makes hormones. These hormones help control body functions such as heart rate, body temperature, blood pressure, and metabolism. In people with *MEN1*, benign tumors are most likely to develop in the endocrine glands of the neck (parathyroid), abdomen (pancreas), and brain (pituitary). Sometimes these tumors can become cancer.

MEN1 Mutations and Tumor Risk

Parathyroid Hormone Levels and Tumors

There are four small parathyroid glands in the neck. These glands make a hormone that helps regulate how much calcium is in the body. Most people with *MEN1* make too much of this hormone. Higher parathyroid hormone levels are usually seen by age 50. Tumors can also form in the parathyroid gland.

Abdominal and Pancreas Tumors

The pancreas is located behind the stomach. It makes hormones such as insulin, which regulates blood sugar, and other hormones. It also helps digestion. People with *MEN1* can develop tumors in the pancreas. Tumors may also grow in the stomach or the upper part of the small intestine.

Pituitary Tumors

The pituitary is a small gland at the base of the brain. It acts as a "master gland" that makes sure other glands in the body are working. The pituitary produces many hormones, including prolactin, which affects fertility and helps make breast milk. It also makes a growth hormone that controls how the body grows, especially during adolescence. People with *MEN1* are more likely to get tumors in their pituitary gland.

<i>MEN1</i> Health Concern	Risk with <i>MEN1</i>
High parathyroid hormone	10 out of 10
Abdominal/pancreas tumors	5 out of 10
Pituitary tumors	4 out of 10
Parathyroid tumors	9 out of 10

Symptoms of *MEN1*

Hormone changes and tumors in *MEN1* can cause different symptoms. Parathyroid tumors can cause tiredness, weakness, muscle or bone pain, kidney stones, and thinning of bones. Pancreas tumors can cause ulcers, trouble breathing or swallowing, diarrhea, and stomach pain. People with pituitary tumors may have trouble with sex and fertility, breast milk production, and pink or purple stretch marks. They may also have weight gain around the stomach, upper back, and face, as well as weight loss in legs and arms. If you have *MEN1* and have these symptoms that cannot be explained by other things, speak with your doctor.

Recommendations

WOMEN AND MEN

People with *MEN1* have a higher risk for many different tumors. Specific cancer screenings are recommended.

Type of Bloodwork	How Often	Starting Age
Glucose, insulin, prolactin, IGF-1	Once a year	Age 5
Calcium, parathyroid hormone	Once a year	Age 8
Chromogranin, gastrin, glucagon, proinsulin	Once a year	Age 20
Type of MRI	How Often	Starting Age
Brain	Every 3 years	Age 5
Chest and abdomen	Every 1-2 years	Ages 10-20

KIDS AND SIBLINGS

Siblings and children of people with *MEN1* have a 1 in 2 chance of also having it. Children in families with *MEN1* should get tested at or before age 5.

It is important to determine which side of the family carries *MEN1* to know which family members are at risk for tumors. A genetic counselor can help you know who in your family should be tested.

Do you have questions about your risk for cancer?

Our doctors and genetic counselors can help find the cancer screening plan you need.

Call Huntsman Cancer Institute's Family Cancer Assessment Clinic to learn more: 801-587-9555.