What you should know about Multiple Endocrine Neoplasia Type 1 (MEN1)

MEN1 is a condition that causes a person to develop tumors in their endocrine glands, as well as non-endocrine tumors. MEN1 is caused by mutations in the MEN1 gene. MEN1 is defined as an individual with at least two of the three main MEN1 endocrine tumors (parathyroid, pituitary, and pancreatic). Individuals who do not meet this definition may also have MEN1.

The risk for tumors associated with MEN1

MEN1 is associated with the following risks for endocrine tumors:

- Parathyroid tumors (100% risk by age 50)
  Tumors that develop in the parathyroid glands in individuals with MEN1 are typically not cancerous, but they can produce excessive amounts of parathyroid hormone. This causes calcium to be moved from the bone to the blood, which causes the bones to become weak and can cause kidney stones to develop.
- Pituitary tumor (10-60% lifetime risk)
- Pancreatic gastrinoma (40% lifetime risk)
- Pancreatic insulinoma (10% lifetime risk)
- Carcinoid (bronchial and thymic cancer) (10% lifetime risk)
- Adrenocortical cancer (20-40% lifetime risk)

MEN1 is also associated with non-endocrine tumors, including facial angiofibromas, collagenomas, and lipomas.

The risks to family members

MEN1 is inherited in an autosomal dominant fashion. This means that the children, brothers, sisters, and parents of individuals with MEN1 have a 50% risk to have MEN1. Approximately 10% of individuals with MEN1 do not have a family history of the disease, and thus have a new mutation.

Managing the risk

The following recommendations can help lower the risk of developing a tumor or catch a developing tumor in an earlier stage:

Biochemical testing:
- Annual serum prolactin, beginning at age 5
- Annual fasting total serum calcium concentration, beginning at age 8
- Fasting serum gastrin concentration, beginning at age 20

Imaging:
- Head MRI, every 3-5 years, beginning at age 5
- Abdominal CT or MRI, every 3-5 years, beginning at age 20

Surgery:
- Sub-total or total parathyroidectomy is usually recommended