MassGeneral Hospital for Children™

Multiple Endocrine Neoplasia Type 1: What You Need to Know

Multiple endocrine neoplasia type 1 (MEN-1) is a rare disorder of the endocrine system that causes multiple tumors associated with an increased secretion of certain hormones. In this handout, learn the causes and symptoms of MEN-1 and how doctors diagnose and treat the disorder.

WHAT IS MULTIPLE ENDOCRINE NEOPLASIA TYPE 1 (MEN-1)?

Multiple endocrine neoplasia is a group of disorders that leads to **tumors** (growth of tissue) in different **endocrine glands** (the system that both makes and regulates hormones). There are several types of multiple endocrine neoplasia. One common type of this disorder is **multiple endocrine neoplasia type 1 (or MEN-1)**. It is also known as **Wermer syndrome**.

MEN-1 is a genetic condition caused by **mutations** (changes) in the *MEN1* gene. This gene usually prevents the growth of tumors. In people with MEN-1, the mutation prevents the gene from working properly. This can lead to the growth of multiple tumors in the endocrine system and elsewhere.

Did you know…?

Not all tumors are dangerous. Tumors can be benign (non-cancerous) or malignant (cancerous). The tumors caused by MEN-1 usually lead to an increased secretion of certain hormones. They may or may not be malignant.

WHAT ARE THE SIGNS AND SYMPTOMS OF MEN-1?

There are many symptoms related to MEN-1. Most occur because of increased **secretion** (production) of certain hormones.

Common conditions associated with MEN-1 include:

- Endocrine tumors in the pancreas, pituitary gland and parathyroid glands
- Hyperparathyroidism, a condition in which the parathyroid glands (four small glands that lie behind the thyroid gland, a butterfly-shaped gland in the front of the neck) become overactive and disrupt the balance of calcium and phosphorus in blood, causing high levels of calcium and low levels of phosphorus. This may in turn be associated with:
    - Kidney stones
    - Fragile bones
- Pituitary tumors, which commonly include tumors that make too much prolactin (**prolactinomas**), or **ACTH** (a hormone that stimulates that adrenal glands to make cortisol), or sometimes growth hormone. Symptoms are related to increased secretion of the specific hormone
- Tumors in the stomach, duodenum or pancreas
    - Zollinger-Ellison syndrome, a condition where the tumor (known as a **gastrinoma**) causes the stomach to make too much acid. This in turn causes **ulcers** (sores) in the stomach or upper intestine
    - Pancreatic tumors which include tumors that make too much insulin (**insulinomas**), as well as other hormone secreting tumors

More signs and symptoms on the back! >>>

WHAT ARE THE SIGNS AND SYMPTOMS OF MEN-1? (CONTINUED)

- Non-endocrine tumors
    - Collagenomas
    - Lipomas, leiomyomas, meningiomas, and ependymomas
    - Facial angiofibromas, which are small red bumps that grow on the face, especially around the nose and cheeks

HOW DO DOCTORS DIAGNOSE MEN-1?

If your child is showing symptoms of MEN-1, one or more of the following are used to diagnose MEN-1:

- Review of the family history
- Physical exam to look for signs of excess hormone
- Blood tests to check hormone levels
- Various imaging tests to check for endocrine tumors
- *MEN-1* genetic testing

WHAT TREATMENTS ARE AVAILABLE FOR MEN-1?

There is currently no cure for MEN1. Early diagnosis and treatment can help manage symptoms and remove tumors. Without proper treatment, MEN-1 can be life-threatening.

Common treatments for MEN-1 include:

- Surgery to remove endocrine tumors
- Medications to help reduce the production of prolactin and the size of tumors making prolactin
- Radiation therapy to shrink or eliminate certain tumors
- Chemotherapy if necessary

In some cases, your child's doctor may suggest waiting and watching for symptoms before recommending surgery, medications or radiation.

Pediatric Endocrinology and Diabetes Center

Mass General Hospital *for* Children
55 Fruit Street, Suite 6C
Boston, MA 02114

www.massgeneralforchildren.org

For more information please call

617-726-2909