Multiple Endocrine Neoplasia Type 1

National Endocrine and Metabolic Diseases Information Service

U.S. Department of Health and Human Services

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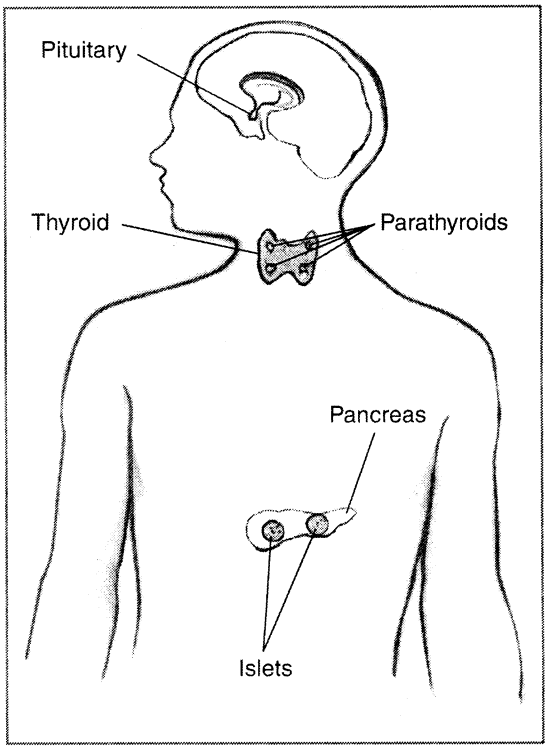
What is multiple endocrine neoplasia type 1 (MEN1)?

MEN1 is an inherited disorder that causes tumors in the endocrine glands and the duodenum, the first part of the small intestine. MEN1 is sometimes called multiple endocrine adenomatosis or Wermer's syndrome, after one of the first doctors to recognize it. MEN1 is rare, occurring in about one in 30,000 people.[1] The disorder affects both sexes equally and shows no geographical, racial, or ethnic preferences.

Endocrine glands release hormones into the bloodstream. Hormones are powerful chemicals that travel through the blood, controlling and instructing the functions of various organs. Normally, the hormones released by endocrine glands are carefully balanced to meet the body's needs.

In people with MEN1, multiple endocrine glands form tumors and become hormonally overactive, often at the same time. The overactive glands may include the parathyroids, pancreas, or pituitary. Most people who develop overactivity of only one endocrine gland do not have MEN1.



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[1]White ML, Doherty GM. Multiple endocrine neoplasia. *Surgical Oncology Clinics of North America.* 2008;17:439–459.

How does MEN1 affect the endocrine glands and the duodenum?

The Parathyroid Glands

The parathyroids are the endocrine glands earliest and most often affected by MEN1. The body normally has four parathyroid glands, which are located close to the thyroid gland in the front of the neck. The parathyroids release into the bloodstream a chemical called parathyroid hormone (PTH), which helps maintain a normal supply of calcium in the blood, bones, and urine.

Hyperparathyroidism

In MEN1, all four parathyroid glands tend to be overactive, causing hyperparathyroidism. The parathyroid glands form tumors that release too much PTH, leading to excess calcium in the blood. High blood calcium, known as hypercalcemia, can exist for many years before it is found by accident or through screening for MEN1. Unrecognized hypercalcemia can cause excess calcium to spill into the urine, leading to kidney stones or kidney damage. Also, the bones may lose calcium and weaken.

Nearly everyone who inherits a susceptibility to MEN1 will develop hyperparathyroidism by age 50, but the disorder can often be detected before age 20. Hyperparathyroidism may cause no problems for many years, or it may cause tiredness, weakness, muscle or bone pain, constipation, indigestion, kidney stones, or thinning of bones.

Doctors must decide whether hyperparathyroidism in MEN1 is severe enough to need treatment, especially in a person who has no symptoms. The usual treatment is an operation to remove most or all of the parathyroid glands. One option is to remove the three largest glands and all but a small part of the fourth. Another is to remove all four glands and at the same time transplant a small part of one gland into the forearm. By maintaining a portion of one gland, the parathyroid transplant continues to release PTH into the bloodstream to do its job.

After parathyroid surgery, regular testing of blood calcium should continue because often the small piece of remaining parathyroid tissue grows larger and causes recurrent hyperparathyroidism. If the remaining piece is in the forearm and additional surgery is needed to remove more parathyroid tissue, the arm operation can be performed under local anesthesia.

Sometimes all four glands are completely removed to prevent recurrence or may be unintentionally removed during parathyroid surgery. People whose parathyroid glands have been completely removed must take daily supplements of calcium and vitamin D or another related treatment to prevent hypocalcemia, or low blood calcium.