Multiple Endocrine Neoplasia

National Cancer Institute

Source

National Cancer Institute. Multiple Endocrine Neoplasia. NCI Thesaurus. Code C6432.

An autosomal dominant inherited neoplastic syndrome characterized by the development of various endocrine neoplasms and abnormalities in various anatomic sites. There are four types recognized: type 1 (MEN 1), caused by inactivation of the tumor suppressor gene MEN-1, type 2A (MEN 2A), caused by mutation of the RET gene, type 2B (MEN 2B) also caused by mutation of the RET gene, and type 4 (MEN 4) caused by mutation of the CDKN1B gene. Patients with MEN 1 may develop hyperparathyroidism and parathyroid gland adenomas, pituitary gland adenomas, pancreatic islet cell neoplasms, and carcinoid tumors. Patients with MEN 2A develop medullary thyroid carcinomas and may also develop pheochromocytomas and parathyroid gland hyperplasia. Patients with MEN 2B develop medullary thyroid carcinomas and numerous neural defects including neuromas. Patients with MEN 4 develop endocrine neoplasms, particularly in the parathyroid glands, pituitary, and pancreas.