## **Condition:** Multiple endocrine neoplasia (MEN 1, 2)

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### Inheritance:
Autosomal Dominant**.**

### Genetic etiology:
MEN1: MEN1 gene on chromosome 11; MEN2: RET gene.

### Frequency:
Approximately 1/30,000 for either MEN1 or MEN2.

### Clinical features:
Patients with MEN1 are at risk of parathyroid adenomas, pituitary tumors, GI tumors (insulinoma, VIPoma, gastrinoma, glucagonoma), carcinoid, adrenal cortical tumors, and pheochromocytoma. MEN2 is divided into subclasses: MEN2a includes parathyroid adenoma, medullary carcinoma of thyroid, pheochromocytoma; MEN2b includes mucosal neuromas, Marfanoid habitus, and medullary carcinoma of the thyroid. Some RET mutations are associated with medullary carcinoma of thyoid as sole manifestation.

### Management:
Symptomatic management and monitoring for tumors. Prophylactic thyroidectomy is recommended for patients with MEN2.

### Genetic counseling:
Affected individuals at 50% risk of transmitting a gene mutation; both familial and de novo cases occur. Genetic testing is available.

*Human Genetics and Genomics*, Fourth Edition. Bruce R. Korf and Mira B. Irons.