

This is for those going for the gold. I include it because it was my first patient in residency. It was also likely my last encounter with it.

MEN1 Syndrome

Also known as **Wermer's Syndrome**, this is an **autosomal dominant** mutation of **MEN1 gene** that causes hyperplasia or adenomas of the "**3 Ps:**" **P**ituitary **A**denomas, **P**arathyroid **A**denomas, and **P**ancreatic **A**denomas. There's a strong association with Gastric Ulcers (Zollinger-Ellison syndrome from the pancreatic adenomas), Hypoglycemia (Insulinoma), and Hypercalcemia (PTH).

MEN2A and MEN2B

These are essentially the same disease and aren't clearly separated. Both are caused by a mutation in the **RET proto-oncogene**. They cause endocrine tumors everywhere except the 3ps. Look for **Pheochromocytomas** and **thyroid adenomas**. The parathyroid gland can also be involved, but isn't classic. Really, the only difference between 2A and 2B is the presence of **neuronal tumors** found in MEN2B.

MEN1 = Pituitary + Pancreas + Parathyroid

MEN2A = Pheochromocytomas + Thyroid + Parathyroid

MEN2B = Pheochromocytomas + Thyroid + Neuronal

Real life vs test

This is rare (~1 in 50,000) – you're not going to see it. If there's a combination of:

- recurrent endocrine neoplasias +
- age <40 +
- family history +
- multiple organ systems affected

You'd suspect something was up and do a **mutational analysis** of the MEN1/2 genes.