# VIPoma

VIPomas are rare functioning neuroendocrine tumors. The majority arises as a non-?, non-? islet cell pancreatic tumor that has an excessive, unregulated secretion of vasoactive intestinal polypeptide (VIP). VIP is a neuropeptide that vasodilates and also regulates smooth muscle activity, epithelial cell secretion, and blood flow in the gastrointestinal tract. Clinical features include GI distress, watery diarrhea that persists with fasting, hypokalemia and achlorhydria. Management consists of giving somatostatin analogs (octreotide) to control the diarrhea and surgical resection if the tumor has not spread to other organs. VIPomas tend to occur as isolated tumors, but may occur in 5% of patients as part of the multiple endocrine neoplasia syndrome type 1.


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## Characteristics

**Pancreatic Neuroendocrine Tumor**
Pancreas with Neuron-indy-car and Tumor-guy
VIPomas are rare functioning neuroendocrine tumors and the majority arise as a non-?, non-? islet cell pancreatic tumor that have an excessive, unregulated secretion of vasoactive intestinal polypeptide (VIP).

**Secretes Vasoactive Intestinal Polypeptide (VIP)**
Secret-agent in VIP-area
VIP is a polypeptide that binds to receptors on intestinal epithelial cells, activating the production of adenylate cyclase and cAMP. VIP also induces vasodilation, inhibits gastric acid secretion, bone resorption, and enhanced glycogenolysis, causing flushing, hypochlorhydria, hypercalcemia, and hyperglycemia.

## Clinical Features

**GI Distress**
GI with Flare-gun
Patients with VIPoma can develop symptoms of GI distress, such as abdominal cramping, nausea, vomiting and diarrhea.

**Watery Diarrhea**
Toilet with Water-spilling out
VIPoma is characterized by watery diarrhea that persists with fasting. Stools are abundant (700-3000 mL/day), tea-colored and odorless. Patients with VIPomas develop this watery diarrhea due to the binding of VIP to receptors on intestinal epithelial cells, which activates the production of adenylate cyclase and cAMP, leading to a net fluid and electrolyte secretion into the lumen.

**Hypokalemia**
Hippo-banana
Hypokalemia is a serum potassium level of < 3.5 mEq/L (3.5 mmol/L). It is a potentially life-threatening imbalance. Hypokalemia seen in patients with VIPoma is due to the massive fecal loss of potassium.

**Achlorhydria**
Acorn-hydra
Achlorhydria is the absence of hydrochloric acid in gastric secretions. Patients with VIPomas develop achlorhydria due to the inhibition of gastrin secretion by VIP.

## Management

**Octreotide**

Octo-tree-ride

Treatment for VIPoma begins with IV fluid replacement and correction of electrolyte abnormalities. After this, somatostatin analogs, such as octreotide, are given to inhibit the secretion of vasoactive intestinal polypeptide and control the diarrhea in these patients.

**Surgical Resection**

Surgeon

Surgical resection of the tumor can cure the patient if it has not spread to other organs. However, in one third to one half of patients with VIPoma, the tumor has spread by the time of diagnosis and cannot be cured.

## Associations

**Multiple Endocrine Neoplasia Type 1 (MEN 1)**

Man with (1) Wand

VIPomas tend to occur as isolated tumors, but may occur in 5% of patients as part of the multiple endocrine neoplasia syndrome type 1 (MEN1) and are associated with parathyroid and pituitary tumors, gastrinoma, and other tumors.

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