The complex endocrinology of Von Hippel Lindau

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Von Hippel Lindau is not considered an endocrine tumor syndrome, but has multiple endocrine manifestions. Pancreatic neuroendocrine tumors, paragangliomas, and pheochromocytomas are the most common endocrine sites. Adrenal insufficiency after bilateral pheochromocytoma resection and supratentorial hemangioblastomas in the pituitary region are also a direct consequence of the disease.

The more widespread use of nuclear imaging such as ^{68}GA-DOTATATE PET/CT imaging has given insight in another endocrine related phenomenon; somatostatin receptor expression is seen in generally all VHL related tumors. Is the endocrinology in VHL underestimated or more complex than assumed?