A collaboration between Penn Gastroenterology and the Penn Renal, Electrolyte and Hypertension results in an interdisciplinary program to diagnose, stage and treat neuroendocrine tumors.

The Penn Neuroendocrine Tumor Treatment Program provides a comprehensive, interdisciplinary approach to the diagnosis, staging, and medical and surgical treatment of gastroenteropancreatic neuroendocrine tumors (GEP-NETs) and pheochromocytomas. GEP-NETs are a rare (~2 percent of all GI tumors), heterogeneous group of malignancies occurring in the digestive tract. Pheochromocytomas are tumors of neuroendocrine chromaffin cells, and are found in both the adrenal glands and in extra-adrenal locations. They are rarer (~2/100,000 persons) than GEP-NETs. With early diagnosis and treatment, tumors in both neuroendocrine classes are potentially curable and manageable for the long term.

Originating in cells having both nervous and endocrine properties, GEP-NETs are classified by histology as either alimentary tract carcinoid lesions (NETs) or pancreatic endocrine tumors (PETs) and subcategorized by whether or not they secrete neuroamines, hormones or peptides at levels sufficient to cause a syndromic response. A recent standard WHO classification has proposed that GEP-NETs be assigned to one of three categories (well-differentiated tumor, well-differentiated carcinoma, and poorly differentiated carcinoma) based on histology, size and proliferative indices.

A Foundation in Experience: The Penn NET Treatment Program

As a result of the rarity and indolent character of GEP-NETs and pheochromocytomas, their early diagnosis depends largely on the experience and expertise of treating clinicians and access to advanced imaging and laboratory facilities—a combination of advantages unique to Penn in the Philadelphia region.

Both Dr. Metz and Dr. Cohen have researched and published on NETs and both have wide experience in the long-term management of NET patients within their respective specialties. In addition, the Penn Neuroendocrine Tumor Treatment Program was developed under the direction of Debbie Cohen, MD (Nephrology) and David Metz, MD (Gastroenterology), shown above with NET Program patient coordinator Bonnie Bennett, BSN.

The Ruth and Raymond Perlman Center for Advanced Medicine
3400 Civic Center Boulevard
Philadelphia, PA 19104
215.349.9222

Penn Presbyterian Medical Center
38th and Market Streets
218 Wright-Saunders Building
Philadelphia, PA 19104
215.662.8900

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Case Study 1: A 49-year-old female with metastatic carcinoid tumors of the liver

Mrs. G, a 49-year-old female, was referred to the Penn Neuroendocrine Tumor Treatment Program for carcinoid tumor surgery. Several months before presenting at Penn, she had developed lower leg edema, flushing and diarrhea. These symptoms led her to visit her ob/gyn, who ordered an abdominal CT scan that found widely dispersed tumors in her liver. At Penn, a 24-hour urine test for 5-hydroxyindolacetic acid (5-HIAA) showed the main urinary metabolite of serotonin, measured >150 mg/day (normal =<6 mg/day), an assessment of chromogranin A (CgA), a NET marker, found levels >100 u/mL (normal range = 2-18 u/L). An octreoscan identified a primary tumor in the terminal ileum and an extensive tumor burden in the right lobes of her liver but no metastases beyond the liver. Mrs. G was diagnosed with widely metastatic carcinoid tumors in her liver and carcinoid syndrome and began octreotide LAR, 20 mg/ month, which improved, but did not resolve her symptoms. Her dose was increased to 30mg/month and following an interdisciplinary review of her tests and scans, it was recommended that Mrs. G have chemoembolization of the tumors in her right liver followed by debulking surgery. Following two visits to interventional radiology for chemoembolization, she had liver resection surgery in the division of gastroenterological surgery. She recovered from these procedures without incident. At this time, her 5-HIAA and CgA levels were within normal levels. Six months post-surgery, her octreoscan revealed no new hepatic lesions and no new metastases. At one year, Mrs. G’s status remains stable on octreotide maintenance therapy.

Case Study 2: Bilateral adrenal pheochromocytoma

Mr. R presented at age 12 with headaches and diarrhea; he was diagnosed with a right adrenal pheochromocytoma and underwent right adrenalectomy. At age 37, Mr. R was seen at the Penn Center for Complex Hypertension with recurrence of diarrhea and headaches. His BP was 132/80 mm Hg and he was not on any antihypertensive medications. He was found to have a left adrenal mass consistent with pheochromocytoma and was scheduled for a second adrenalectomy. He was treated with dibenzpyrene for prooperative alpha blockade and alpha methyl-butyrate. A left-sided adrenalectomy was performed, now able to produce endogenous steroids. Mr. R began a regimen of hydrocortisone and fludrocortisone. Because certain genetic mutations are associated with bilateral adrenal pheochromocytoma, Mr. R was referred for genetic testing. Genotyping studies were positive for Von Hippel Lindau VHL mutation, an autosomal dominant trait with a 50 percent risk of inheritance. Mr. R’s 18-year-old son, JW, was also found to be carrying the VHL mutation. JW had no symptoms; supine and sitting BP were 120/78 mm Hg and 110/82 mm Hg. Standing blood pressure was 96/74 mm Hg with a heart rate of 120 beats/minute. Three studies show elevated normetanephrine levels. MRI of the abdomen showed a left adrenal mass and laparoscopic adrenocortical sparing surgery was performed. While his BP remained normal with home BP monitoring, JW’s plasma and urine metanephrines never “normalized.” A repeat MRI performed a year later revealed a new tumor in the right adrenal gland and a second adrenal cortex sparing surgery for his second pheochromocytoma was performed. Both Mr. R and JW remain disease-free several years later with yearly surveillance with blood tests and imaging for recurrent pheochromocytoma.