



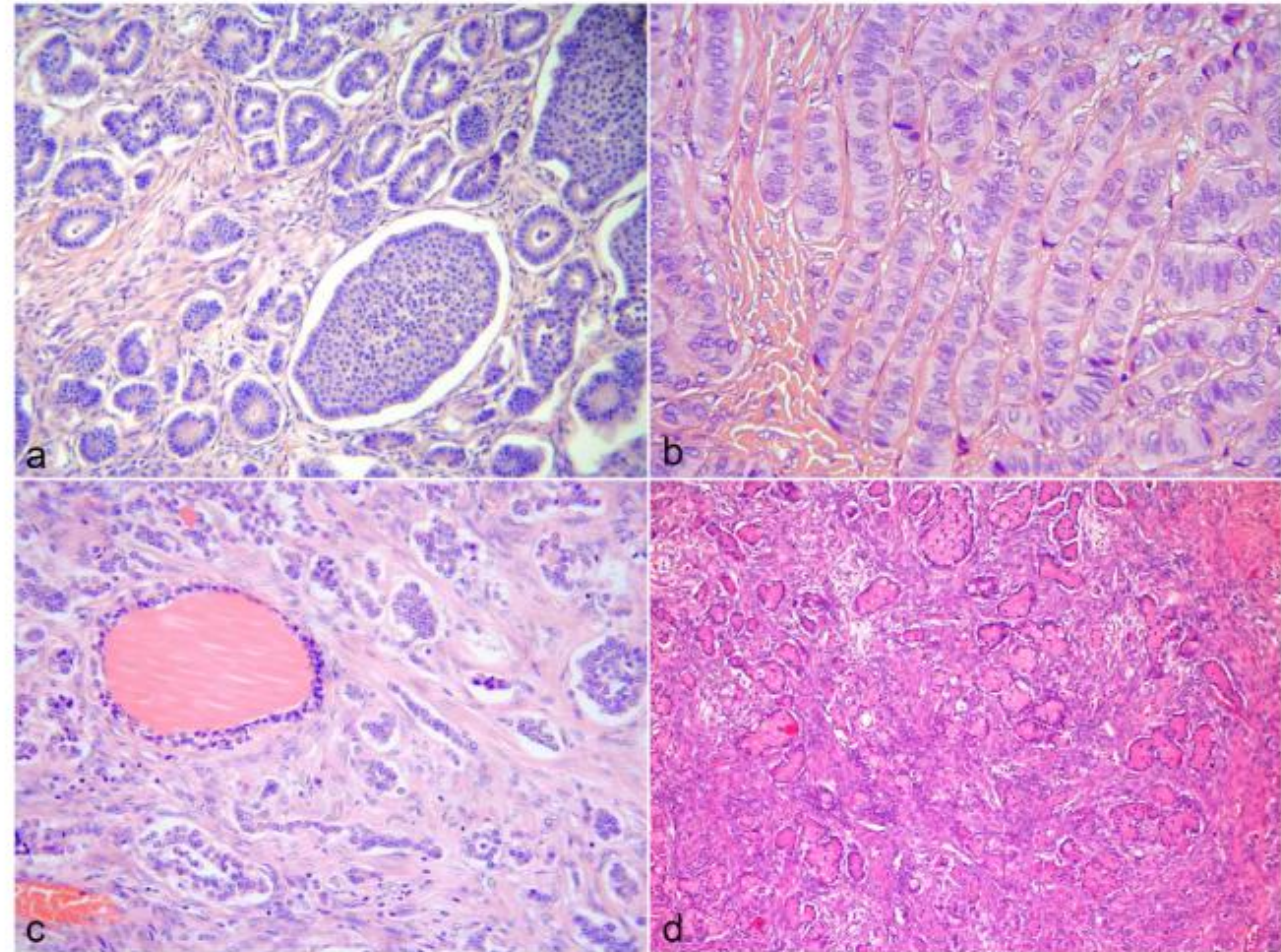
Case 3

Primary Ovarian Neuroendocrine Neoplasm



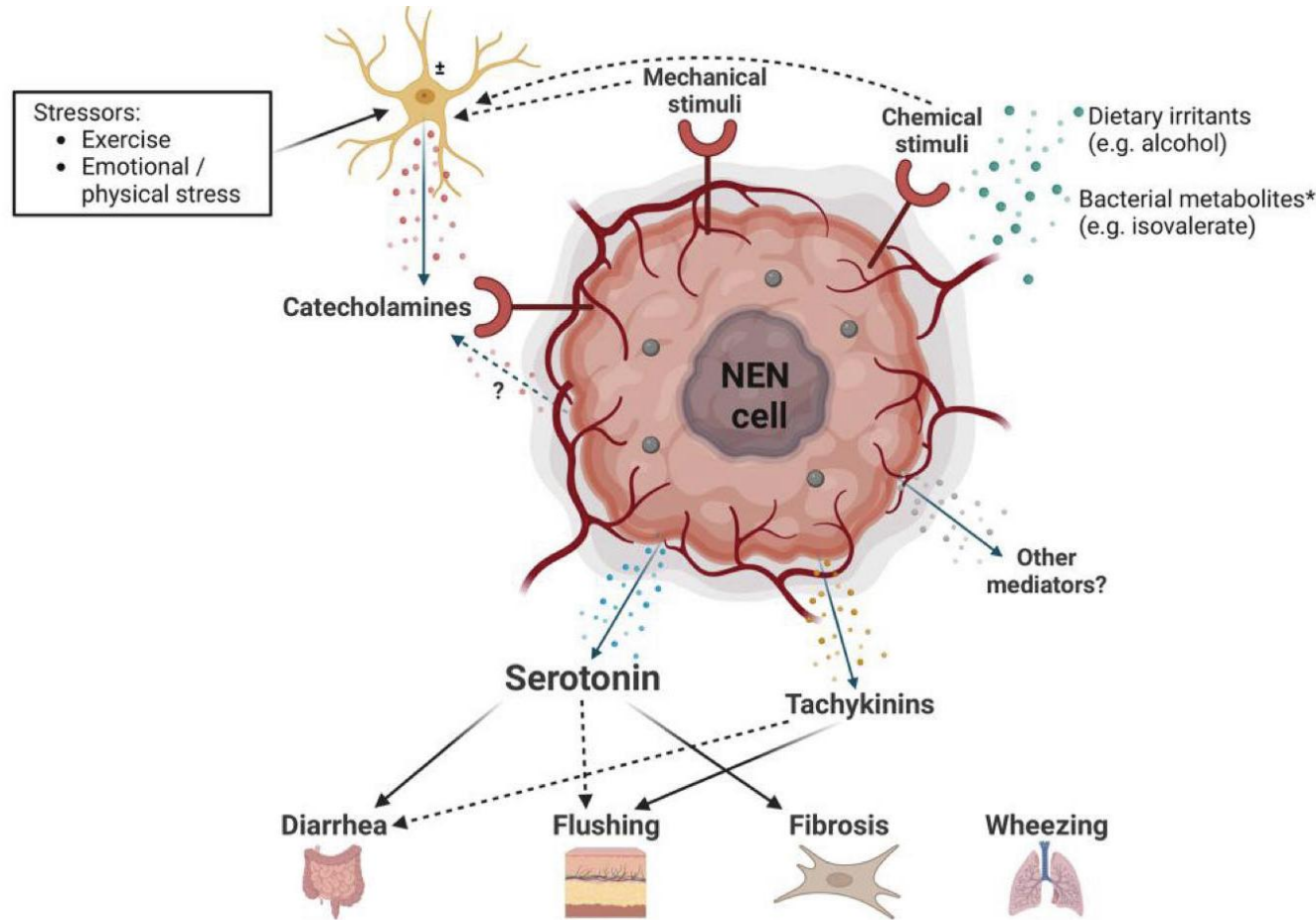
Primary Ovarian Neuroendocrine Neoplasm

- ❑ The majority of NENs arise in the midgut (small intestine, appendix, proximal colon), or lung/pancreas
- ❑ Minority will arise in other locations
- ❑ Primary ovarian NENs are extremely rare and only account for approximately 1% of NENs
- ❑ Primary ovarian NETs develop in either **pure** form or in **association with teratomas**
- ❑ Subtypes based on the pattern of their histopathology: insular (a), trabecular (b), stromal (c), mucinous (d), or mixed type





Ovarian NEN with Carcinoid syndrome



- ❑ **Carcinoid syndrome (CS) affect 19% of NENs** : associated with the secretion of several vasoactive amines: **serotonin**, catecholamines, tachykinins, bradykinin, kallikrein, histamine, prostaglandin
- ❑ In extensive liver metastases, a large amount of tumor-secreted substances are not completely metabolized by hepatic or pulmonary cells and enter the systemic circulation, causing CS

Flushing 90%

CHD (Rt >> Lt) 17-21%

Diarrhea 60-80%

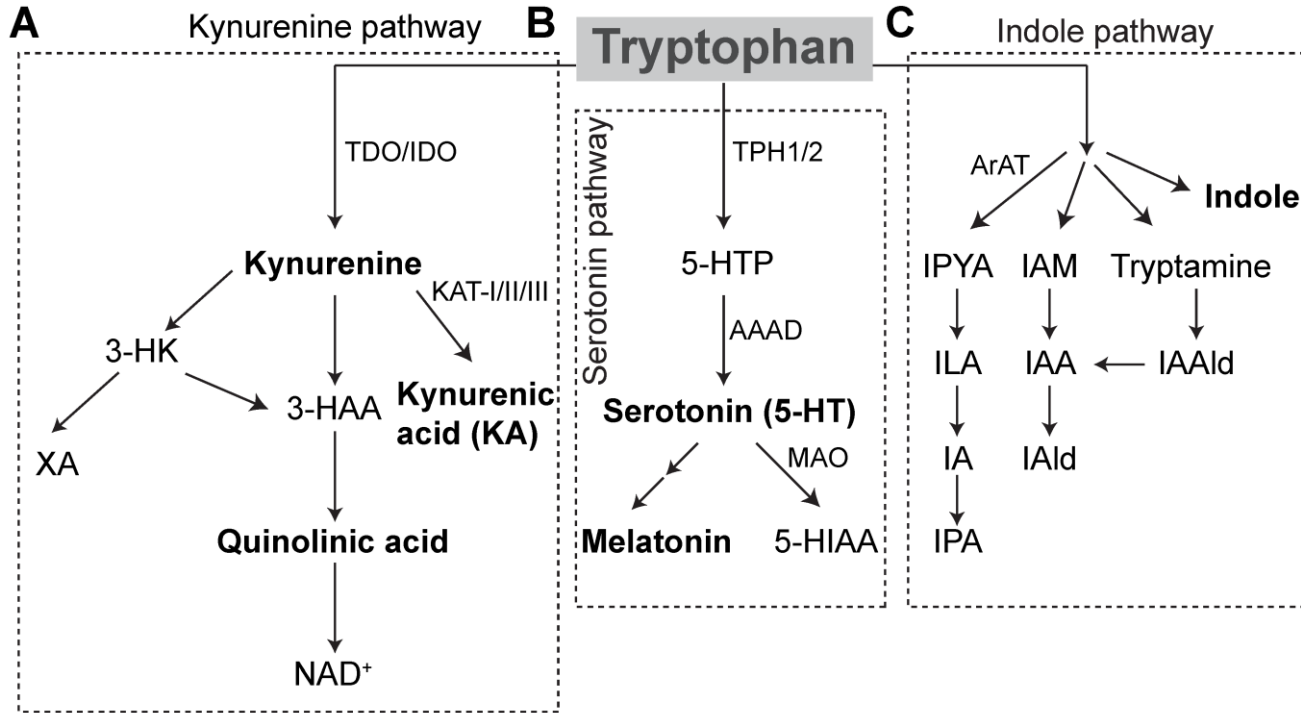
Bronchospasm 15%

Mesenteric fibrosis 50%

Pellagra 5%



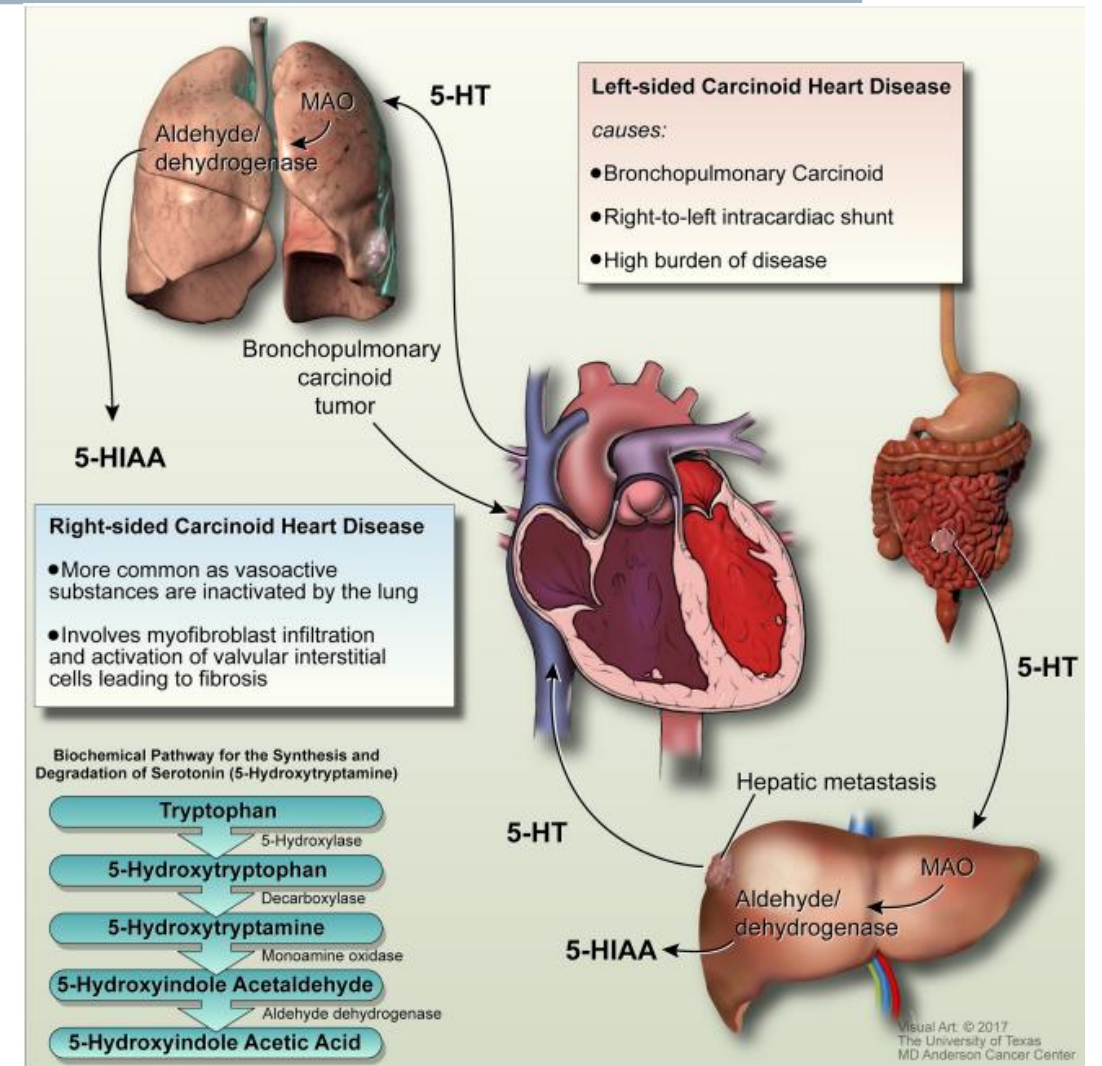
Tryptophan & Serotonin Metabolism



**Vitamin B3
(99% of pathway)**

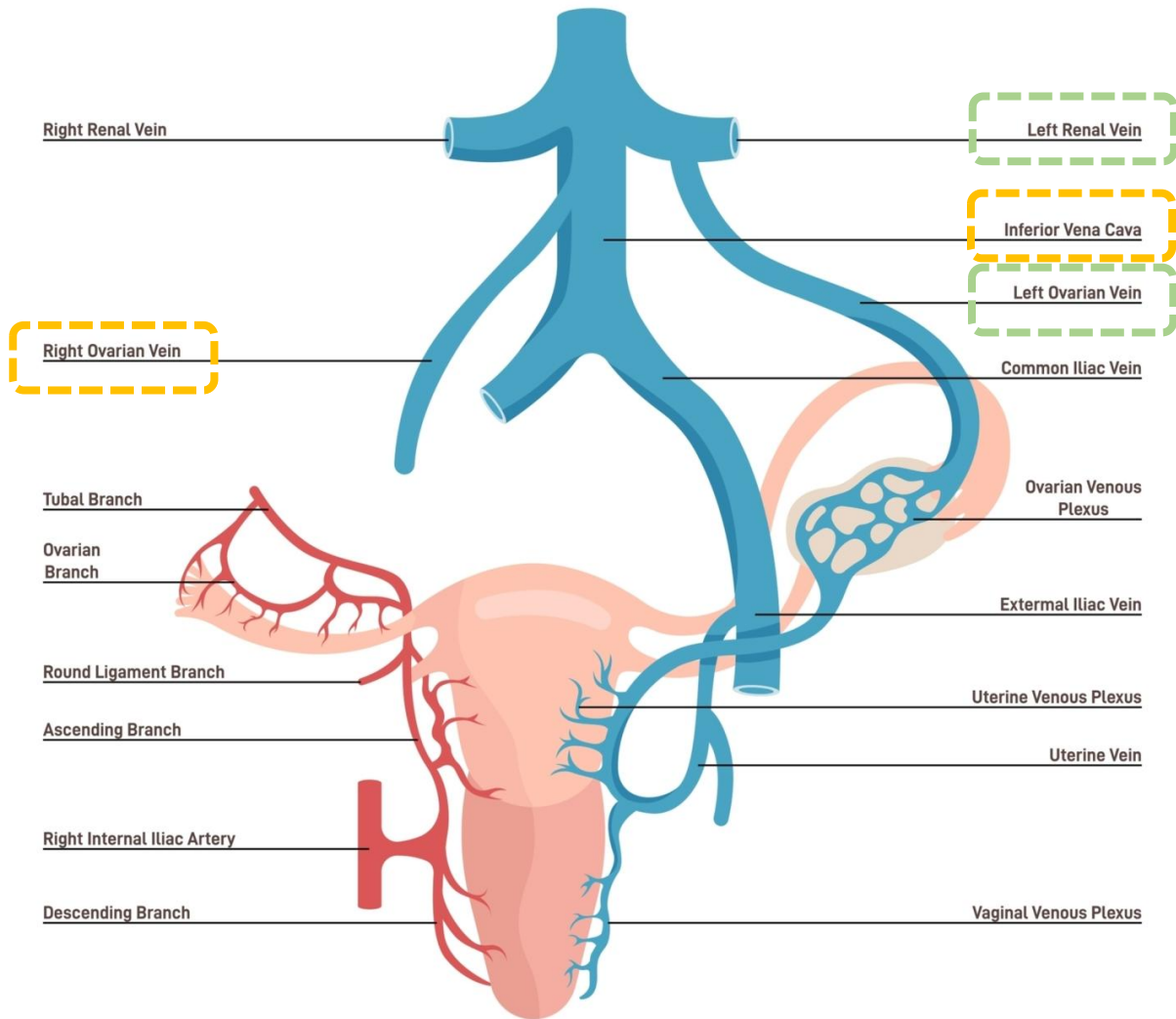
**Carcinoid
syndrome**

**Gut
Microbiota**





Ovarian NEN with Carcinoid syndrome



- ❑ CS may appear without pre-existing liver metastases: ovarian NENs, extensive retroperitoneal lymph node metastases (**by pass hepatic-portal circulation, drain directly into the systemic circulation**)
- ❑ Ovarian NEN present with CS independent of liver metastases and thus may present relatively early
- ❑ Biomarker: plasma chromogranin A, 24 h urinary 5-HIAA (metabolite of serotonin)



Management



- Extrapolated from that for the more common NENs: Sx+- adjuvant therapy
- For the young age group, take into account the possibility of future fertility
- No indication for any adjuvant treatment in well-differentiated tumors completely excised
- Niacin supplementation of 25–50 mg a day
- In case of residual/metastatic disease or pre-operative treatment

SSA: Octreotide LAR 20-30 mg IM q 4 wk
Lanreotide 120 mg SC q 4wk
→ reducing the release of hormones by tumor cells

mTOR inhibitor (Everolimus): combined with SSA
→ reduction of urinary 5-HIAA

Tryptophan hydroxylase inhibitor (Telotristat ethyl)
→ reduction of urinary 5-HIAA and diarrhea

Hepatectomy or TACE for liver metastasis
PRRT with Lu-177 or Y-90 for metastatic disease

Cytotoxic CMT is **not** recommended due to poorly response



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Thank You