

CNETS Patient Conference

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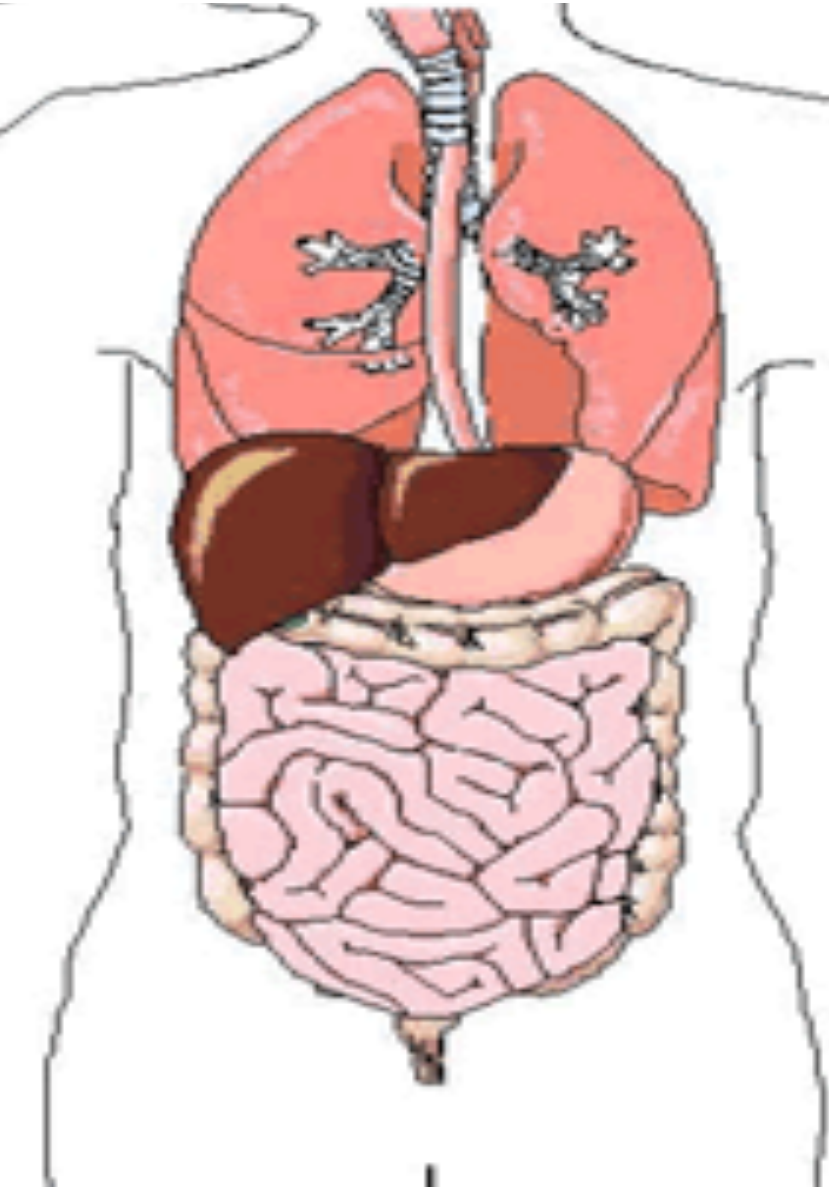
Medullary Thyroid Cancer

A primer for patients with a rare NE cancer

Neuroendocrine Cells & the Neuroendocrine System

- ▶ What are neuroendocrine cells?
 - Cells scattered throughout the body **like a web** that share a common embryologic origin and function as cellular “messengers”.
 - Communicate by releasing proteins.
 - This web of communicator cells make up the **Diffuse Neuroendocrine System**

The Diffuse Neuroendocrine System



Embryologic Derivation*
Foregut (33%):

lungs, thymus, **THYROID**,
esophagus, stomach,
duodenum, pancreas

Midgut (34%):

jejunum, ileum, appendix,
cecum, ascending colon

Hindgut (14%):

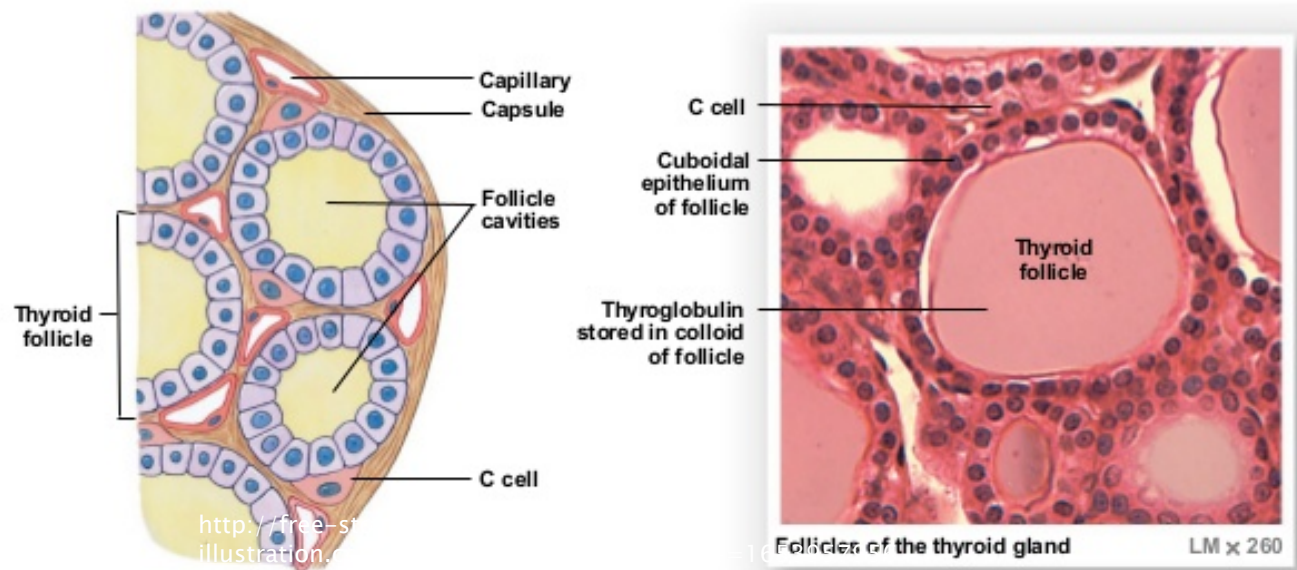
Descending colon, rectum

*NE cancers are cancers that
arise from these cells

Thyroid physiology

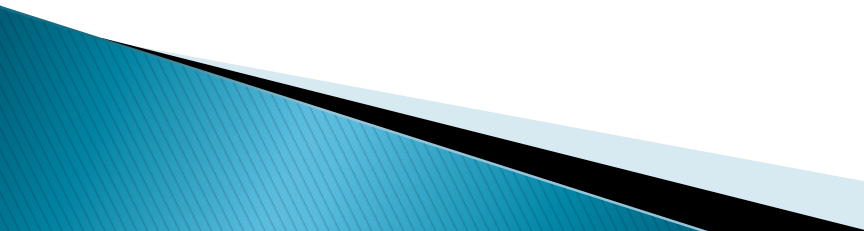
- Parafollicular C cells are responsible for the production of a hormone called Calcitonin.

- C Cells produce Calcitonin (CT)
- regulates calcium in blood



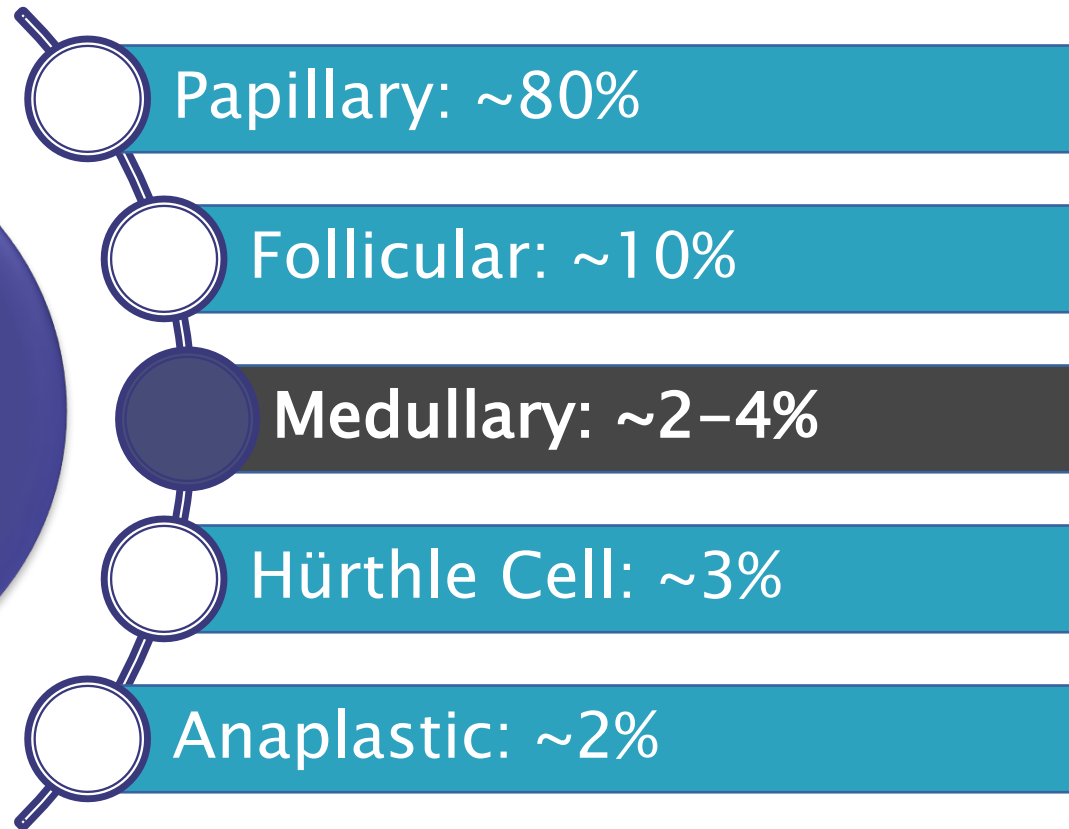
c Histological details of the thyroid gland showing thyroid follicles and both of the cell types in the follicular epithelium *ATLAS: Plate 18c*

Medullary Thyroid Cancer (MTC)

- ▶ A rare form of Neuroendocrine Cancer that arises from malignant change and growth of Parafollicular C cells.
 - ▶ MTC cells also produce Calcitonin.
 - ▶ Elevated levels of Calcitonin can produce some of the symptoms (diarrhea, flushing, itching) associated with medullary thyroid cancer.
- 

TYPES OF THYROID CANCER

9000 new cases of MTC per year globally



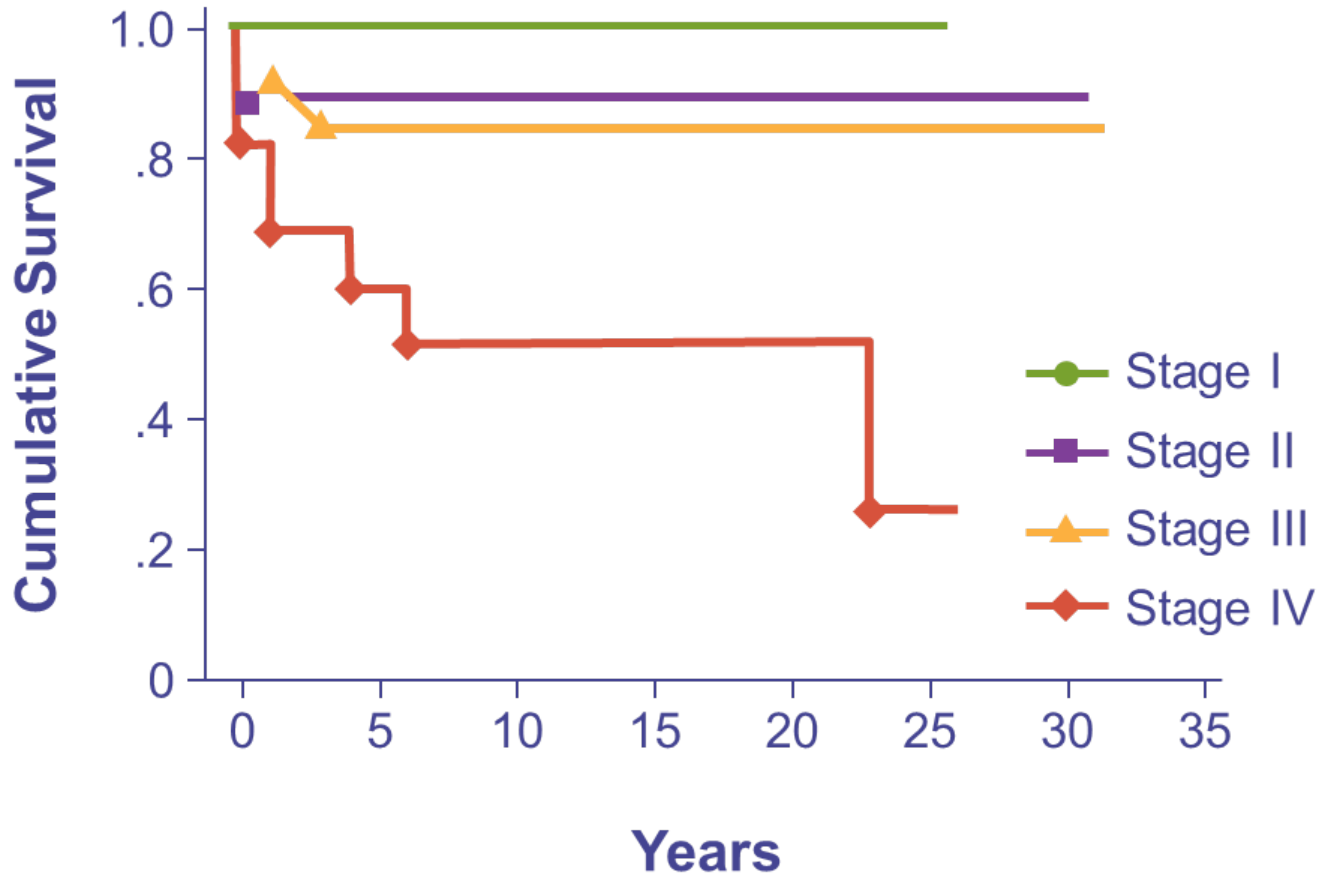
Medullary Thyroid Cancer

- ▶ MTC is different than more common forms of thyroid cancer in important ways.
 - The importance of family history.
- Sporadic: 75%
 - Thyroid nodule, distant metastatic disease, diarrhea, flushing or itching
- Hereditary/Familial MTC: 25%
 - Similar presentation
 - Family history of associated cancers and other syndromes that should prompt genetic testing.

Medullary Thyroid Cancer

- ▶ Many patients with MTC live with their disease for a long time.
 - The 10 year overall survival rate in patients with MTC is approximately 75%
 - In patients with locally advanced or metastatic disease 10 year survival is 40%
- ▶ The treatment of advanced disease has been frustrating due to a lack of good treatment options until recent years

MTC PATIENT SURVIVAL BY STAGE



TNM, tumor, node, metastasis.

Kebebew E, et al. *Cancer*. 2000; 88:1139-1148.

Tumor Markers in MTC

▶ Calcitonin

- Produced by parafollicular C cells and MTC
- Helpful in making the diagnosis
- Measured over time as part of strategy to monitor disease stability/ progression and monitor effects of treatment.

▶ Carcinoembryonic Antigen (CEA)

- A tumor marker (not specific for MTC) that can be used to assist with monitoring and follow up

Treatment of MTC

- ▶ Surgical resection is the most important treatment modality
 - For cure in local disease
 - To assist in disease management in advanced disease
- ▶ Radioactive Iodine treatment has no role in MTC (unlike more common types of thyroid cancer)
- ▶ In advanced (incurable) disease we have new treatments that can be used to manage the disease and associated symptoms, improve patients QOL and potentially extend survival.

Treatment of Advanced MTC

- ▶ Surgery
 - where feasible to manage recurrent disease in the neck and in some patients, to treat disease in other parts of the body.
- ▶ External beam radiation treatment
 - Tumors in lungs or bones

Treatment of Advanced MTC

▶ Systemic treatment

- Symptomatic Management
 - Imodium
 - Narcotics
- Somatostatin analogues (Octreotide) ?
 - To manage diarrhea and flushing
- MIBG
 - A form of “targeted” radiation delivered by intravenous injection

Treatment of Advanced MTC

- ▶ Systemic treatment
 - New drugs
 - Vandetanib
 - Cabozatinib
- ▶ Belong to a class of drugs called Tyrosine Kinase Inhibitors (TKI's).
 - Both of these agents can be used to delay progression of disease, manage symptoms and improve Quality of Life.

Treatment of MTC

- ▶ The importance of Multidisciplinary Care
 - Enhanced coordination and communication within the health care team and with our patients. Timely, patient focused care.
 - Use of local therapies (surgery, radiation therapy) for both curative intent and palliative treatment.
 - Choosing the right time for a patient to use systemic (drug) treatments to manage advanced MTC.
 - Educating and supporting patients living with a “chronic” cancer

Treatment of MTC

- ▶ The importance of Multidisciplinary Care
 - Rehabilitation
 - Speech, swallowing
 - Access to medical genetics
 - Assessment of patients and family members
 - Support to families and care givers