Endocrine Disorders for the USMLE, Step One:

Multiple Endocrine Neoplasia Type 1 (MEN-1 Syndrome)

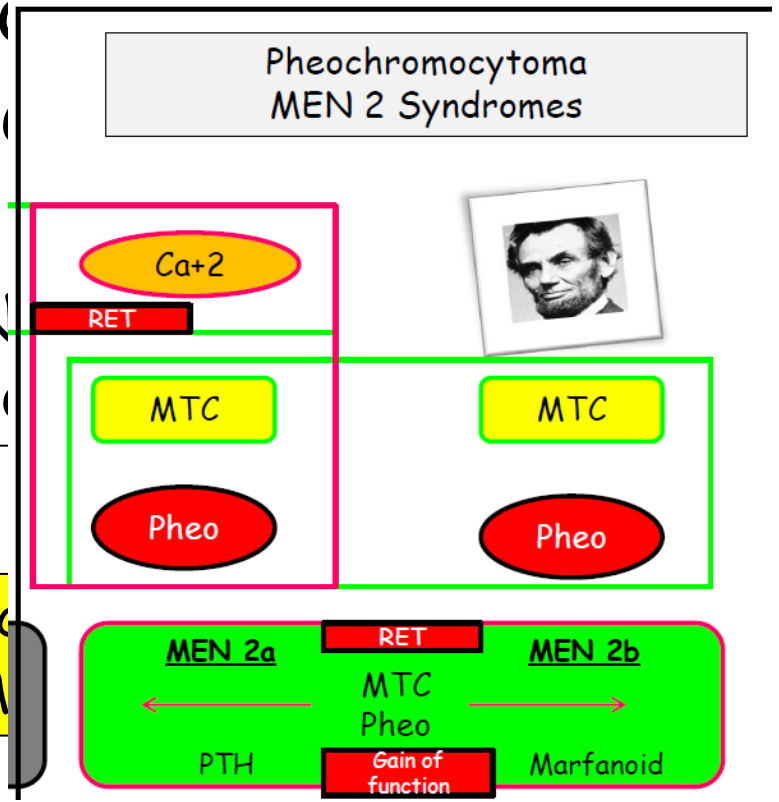
Howard J. Sachs, MD
www.12DaysinMarch.com

Endocrine Disor
USMLE, St

Multiple Endocrine N
(M

View MEN2 Syndromes
BEFORE
this video

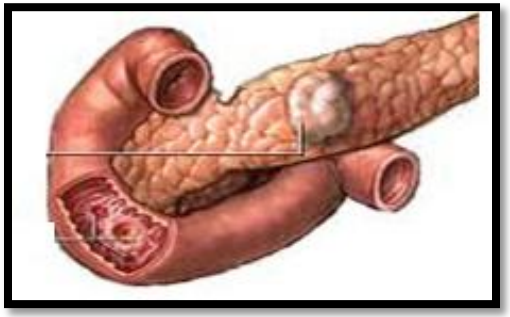
Howard J. Sac
www.12DaysinM



MEN-1 Syndrome

Pituitary

Parathyroid

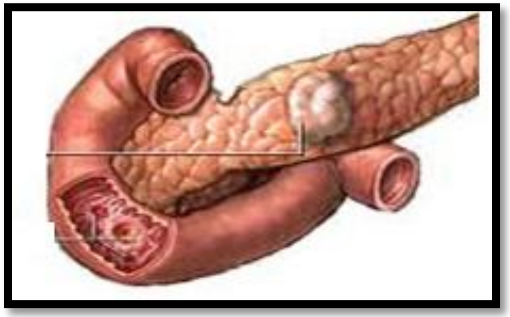


Pancreas

MEN-1 Syndrome

Pituitary

Parathyroid



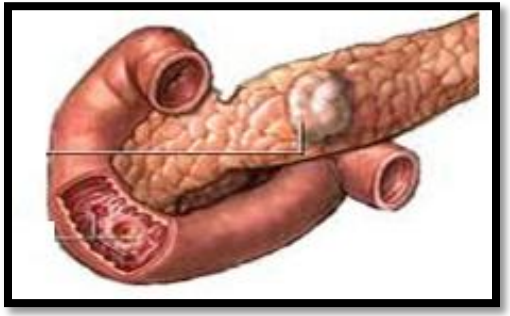
Pancreas

3-P's

MEN-1 Syndrome

Pituitary

Parathyroid



Pancreas

3-P's

MEN-1 Syndrome

Before proceeding, we need to change the name into something more memorable and informative.

Once we move from MEN-1 and the 3-P's, the rest is a breeze.

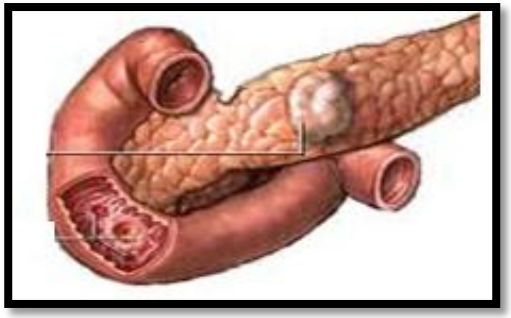
Really.

3-P's

MEN-1 Syndrome

Parathyroid

~100%



Pancreas

~60%

Pituitary

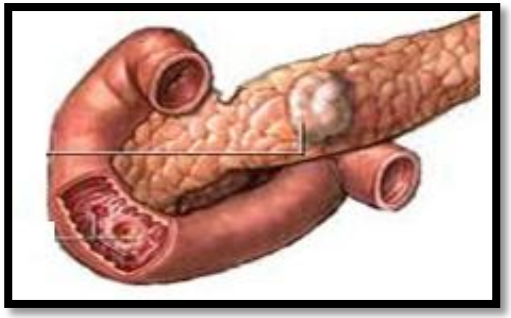
~20%

MEN-1 Syndrome

Parathyroid

~100%

Multiple Adenomas



Pancreas

~60%

Gastrinoma (ZE)

Pituitary

~20%

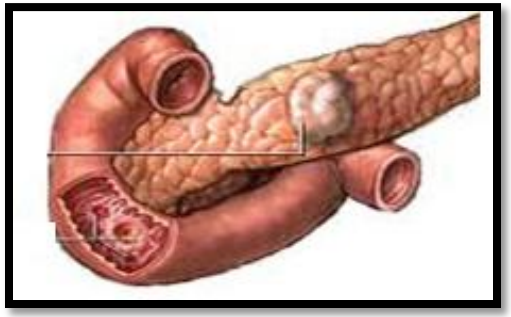
Prolactinoma

MEN-1 Syndrome

Pituitary

~20%

Prolactinoma



Pancreas

~60%

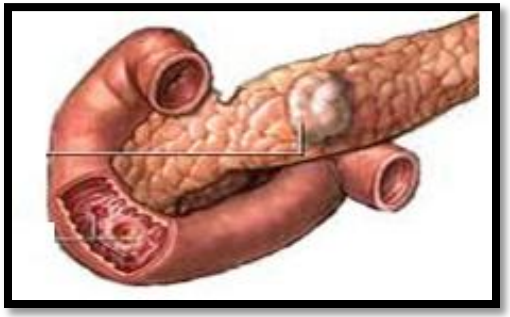
Gastrinoma (ZE)

Parathyroid

~100%

↑ Ca^{+2}

MEN-1 Syndrome



Pituitary	Pancreas	Parathyroid
~20%	~60%	~100%
Prolactinoma	Gastrinoma (ZE)	↑ Ca^{+2}

Pro-ZE-Ca Syndrome

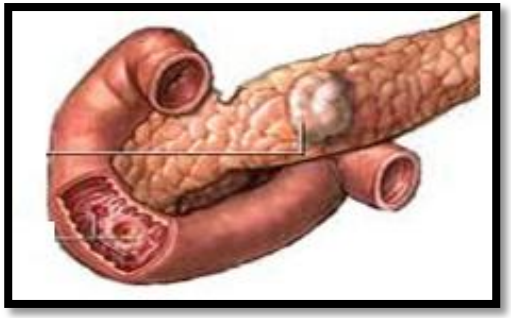
MEN 1: Pro-ZE-Ca Syndrome

Pituitary

~20%

Prolactinoma

Mass Effect
Hormonal Dysfunction



Pancreas

~60%

Gastrinoma (ZE)

Multiple/Atypical
Ulcers

Parathyroid

~100%

Multiple Adenomas

↑ Ca^{+2}

MEN 1: Pro-ZE-Ca Syndrome

What are the take homes?

Background:

A. High penetrance
B. Initial presention in majority of MEN
C. Compared with sporadic:
   - Multiple adenomas and early age

Presentation:

D. Same as any other patient with hyperPTH
   - ↑ Ca, ↑ PTH, ↓ $PO4^-$
   - Stones, Constipation
   - X-ray: subperiosteal bone resorption

Parathyroid

~100%

Multiple Adenomas

↑ Ca^{+2}

MEN 1: Pro-ZE-Ca Syndrome

What are the take homes?

Background:

A. High penetrance
B. Initial presention in majority of MEN
C. Compared with sporadic:
   - [illegible]
D. Same as any other patient with hyperPTH
   - ↑ Ca, ↑ PTH, ↓ $PO4^-$
   - Stones, Constipation
   - X-ray: subperiosteal bone resorption

On USMLE, any patient diagnosed or presenting with HyperPTH, you need be HyperVigilent for an MEN Syndrome (Type 1 or 2a)

[illegible]thyroid

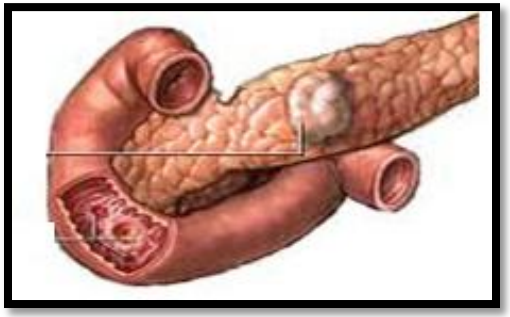
[illegible]00%

Multiple Adenomas

↑ Ca^{+2}

MEN 1: Pro-ZE-Ca Syndrome

What are the take homes?



Pancreas

~60%

Gastrinoma (ZE)

Multiple/Atypical Ulcers

Background (Pancreatic Endocrine Tumors):

A. Gastrinoma most common
   - May present in duodenal location

B. Insulinoma, VIPoma, Glucagonoma
   - Hypoglycemia, WDHA, NME (bronze skin)

C. Carcinoid (bronchial, thymus)

Presentation:

D. Multiple or Atypical Ulcers
   - Duodenal beyond D1
   - Epigastric pain despite rx

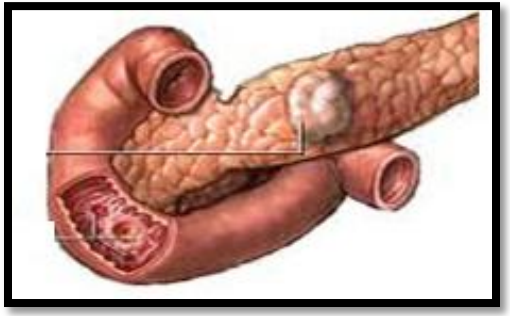
E. Diarrhea (inactivates lipase/acidic lumen)

MEN 1: Pro-ZE-Ca Syndrome

What are the take homes?

<u>Diagnostics (Gastrinoma)</u>:

1. High gastric basal acid secretion
2. ↑ Gastrin Level
3. Secretin Stimulation Test
    - Rise in Gastrin Level



Pancreas

~60%

Gastrinoma (ZE)

Multiple/Atypical Ulcers

MEN 1: Pro-ZE-Ca Syndrome

1. High gastri
2. ↑ Gastrin L
3. Secretin S
   - Rise in

Be on the lookout with any question that includes Gastrinoma, VIPoma, or Insulinoma.

If they include ↑ Ca^{+2}, you have two components of MEN-1…

…they will most assuredly come after you on the third component…

the Pituitary

ncreas

~60%

inoma (ZE)

Multiple/Atypical Ulcers

MEN 1: Pro-ZE-Ca Syndrome

Pituitary

~20%

Prolactinoma

Mass Effect
Hormonal Dysfunction

What are the take homes?

Background:

1. Adenomas of Anterior Pituitary follow same pattern as in non-MEN syndromes
   - Prolactin > Growth Hormone > Null
   - Hormone product versus Mass Effect

Presentation:

1. Pituitary mass lesion: HA, visual field cut
2. Prolactinoma: hypogonadotropic hypogonadism
   - Male: ↓ libido/ED
   - Female: Δ menses, galactorrhea

MEN 1: Pro-ZE-Ca Syndrome

Pituitary

~20%

Prolactinoma

Mass Effect
Hormonal Dysfunction

What are the take homes?

<u>Diagnostics</u>:

1. ↑ Prolactin level
2. Men: ↓ LH, testosterone
3. Imaging for mass effect or ↑ PRL

<u>Note</u>:

Frequency of pituitary lesion in MEN-1 depends on how hard you look (20% clinical, 60% by MRI screen)

MEN 1: Pro-ZE-Ca Syndrome

What are the take homes?

Interestingly, I've mentioned you should be hypervigilant looking for MEN1 in patients with hyperPTH and/or pancreatic masses or neuroendocrine tumors.

On the other hand, questions involving pituitary adenomas are usually focused on the adenoma and secretions.

20%

Prolactinoma

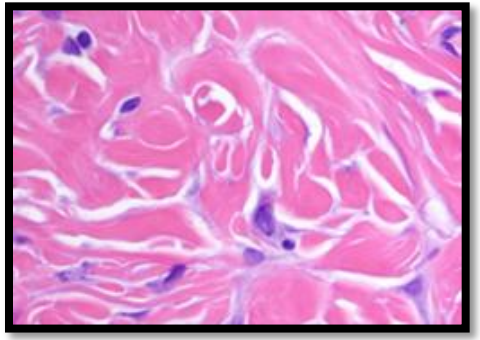
Mass Effect
Hormonal Dysfunction

Note:
Frequency of pituitary lesion in MEN-1 depends on how hard you look
(20% clinical, 60% by MRI screen)

MEN-1 Syndrome

- Reviewed major components
  - Endocrinopathy (PTH, Pancreas, Pituitary)
  - Derm Manifestations

Angiofibroma (90%)



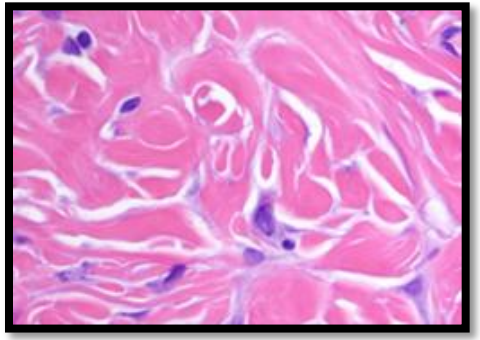
Collagenoma

Lipoma

Lip/Tongue Neuromas seen in MEN2b

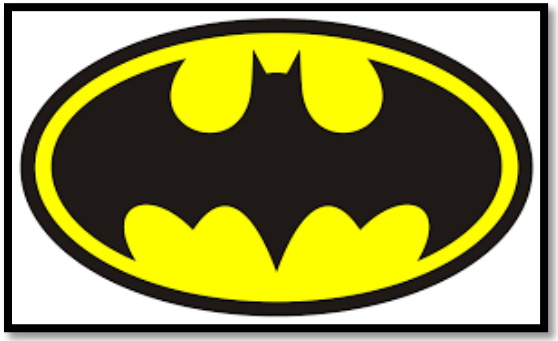
- Derm Manifestations

Angiofibroma (90%)



Collagenoma

Lipoma



MEN-1
(Pro-ZE-Ca)

1. Pituitary (Prolactinoma)
2. Pancreas (Gastrinoma; ZE)
3. Parathyroid (100%; ↑ Ca^{+2})

MEN-2

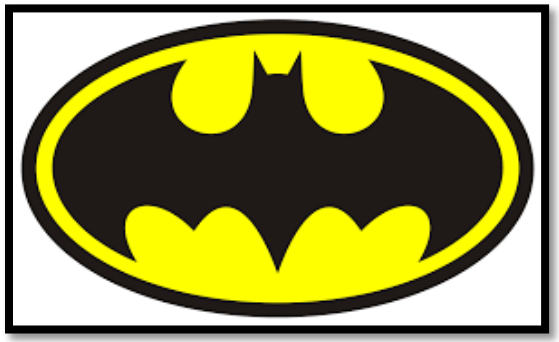
1. MTC (100%)
2. Pheochromocytoma (50%)

2a
HyperPTH

2b
Marfanoid, Neuromas

The most difficult part of an MEN Syndrome is knowing you are in one…

Autosomal Dominant



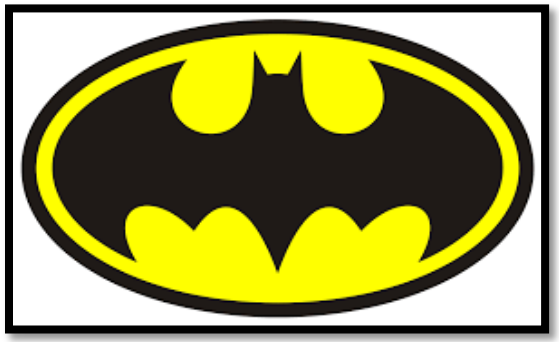
MEN-1
(Pro-ZE-Ca)

1. Pituitary (Prolactinoma)
2. Pancreas (Gastrinoma; ZE)
3. Parathyroid (100%; ↑ Ca^{+2})

MEN-2
1. MTC (100%)
2. Pheochromocytoma (50%)

2a	2b
HyperPTH	Marfanoid, Neuromas

Autosomal Dominant



MEN-1
(Pro-ZE-Ca)

1. Pituitary (Prolactinoma)
2. Pancreas (Gastrinoma; ZE)
3. Parathyroid (100%; ↑ Ca^{+2})

MEN-2
1. MTC (100%)
2. Pheochromocytoma (50%)

2a
HyperPTH

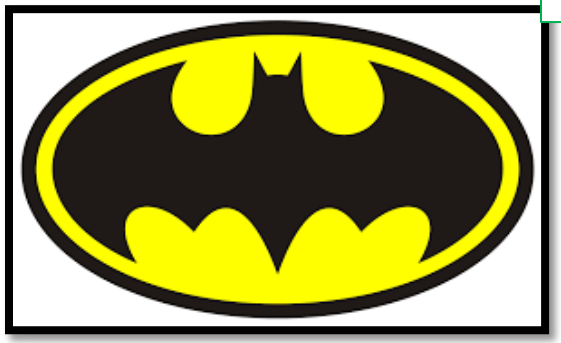
2b
Marfanoid, Neuromas

Loss of Suppressor

MEN1 Mutation

Gain of function

RET Mutation



MEN-1
(Pro-ZE-Ca)

1. Pituitary (Prolactinoma)
2. Pancreas (Gastrinoma; ZE)
3. Parathyroid (100%; ↑ Ca^{+2})

MEN-2

1. MTC (100%)
2. Pheochromocytoma (50%)

2a
HyperPTH

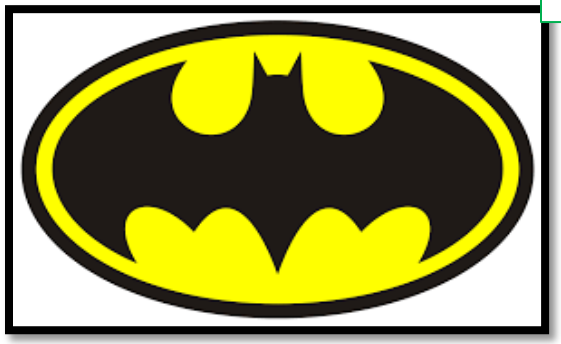
2b
Marfanoid, Neuromas

Loss of Suppressor

MEN1 Mutation

Gain of function

RET Mutation



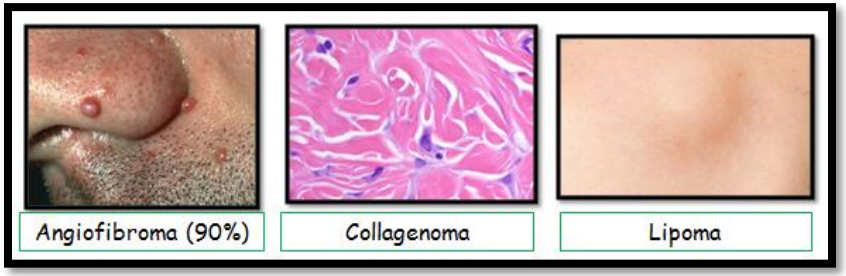
MEN-1
(Pro-ZE-Ca)

1. Pituitary (Prolactinoma)
2. Pancreas (Gastrinoma; ZE)
3. Parathyroid (100%; ↑ Ca^{+2})

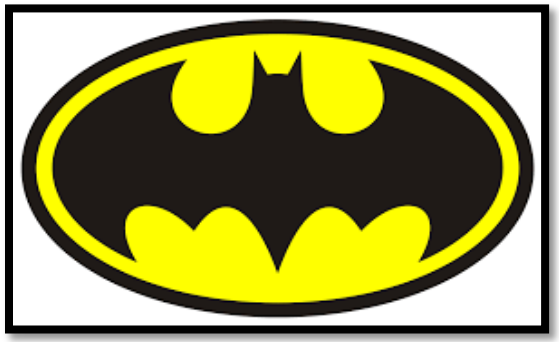
MEN-2
1. MTC (100%)
2. Pheochromocytoma (50%)

2a
HyperPTH

2b
Marfanoid, Neuromas



Autosomal Dominant



MEN-1
(Pro-ZE-Ca)

1. Pituitary (Prolactinoma)
2. Pancreas (Gastrinoma; ZE)
3. Parathyroid (100%; ↑ Ca^{+2})

MEN-2
1. MTC (100%)
2. Pheochromocytoma (50%)

2a	2b
HyperPTH	Marfanoid, Neuromas

Endocrine Disorders for the USMLE, Step One:

Multiple Endocrine Neoplasia Type 1 (Pro-ZE-Ca or MEN-1 Syndrome)

PROZAC®

Howard J. Sachs, MD
Email: Howard@12DaysinMarch.com