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# NEN of the Appendix The patient

- 34 years old
- Appendectomy last month
- Yesterday, surgical clinic visit
- "you have a tumor in the appendix, a neuroendocrine tumor of 2 cm, you must have another surgery to resect larger part of the colon and the lymph nodes"
- What to do now??

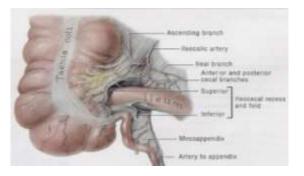






#### What is NEN of the Appendix?

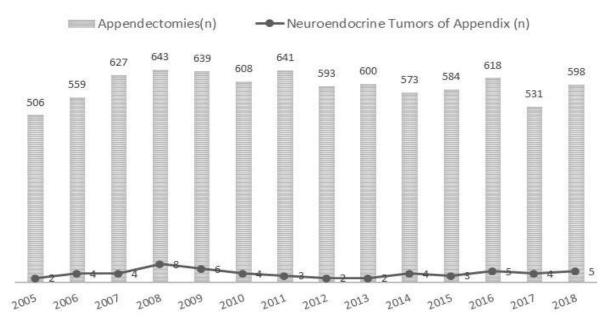
- Tumor originating from the appendix
- Usually found incidentally at appendectomy
- Female: male= 70:30
- May appear at any age (8-80 years)
- Prognosis is excellent
- Hormone secretion is extremely rare







#### NEN of the Appendix Incidence



Twito et al, 2020

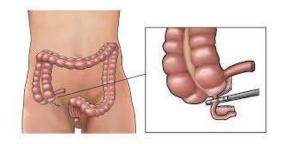
- 1-5/1,000,000 persons per year
- approximately 1/200 appendectomies





## NEN of the Appendix Treatment

- Most of ANEN are localized to the appendix
- Appendectomy is curative in majority of patients
   However...
- A portion of patients develop lymph node metastases (5-30%?) or distant metastases (4-10%??)





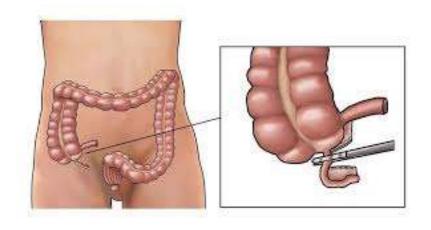




## NEN of the Appendix Treatment

#### **CLINICAL DILLEMAS:**

- HOW TO IDENTEFY PATIENTS WITH METASTATIC DISEASE?
- CAN WE PREVENT THE DEVELOPMENT OF METASTATIC DISEASE? (medical treatment, surgery)









## NEN of the Appendix Treatment

## HOW TO IDENTEFY PATIENTS WITH METASTATIC DISEASE?

- Imaging studies (CT, MRI, PET)
- Biochemical markers (blood, urine)
- Risk stratification
  - ✓ Tumor size>2cm
  - √ Special subtypes(e.g. goblet cell carcinoid)
  - ✓ Tumor invasion
  - ✓ Unclear surgical margins





# NEN of the Appendix International guidelines

**ENETS Consensus Guidelines** 



Neuroendocrinology 2012;95:135-156 DOI: 10.1159/000335629 Published online: February 15, 2012

NAN

ENETS Consensus Guidelines for the Management of Patients with Neuroendocrine Neoplasms from the Jejuno-Ileum and the Appendix Including Goblet Cell Carcinomas

The NANETS Consensus Management of Ulrich-Frank Pape<sup>a</sup> Aurel Perren<sup>b</sup> Bruno Niederle<sup>c</sup> David Gross<sup>d</sup> Thomas Gress<sup>e</sup> Frederico Costa<sup>f</sup> Rudolf Arnold<sup>e</sup> Timm Denecke<sup>g</sup> Ursula Plöckinger<sup>a</sup> Ramon Salazar<sup>h</sup> Ashley Grossman<sup>i</sup> and all other Barcelona Consensus Conference participants<sup>1</sup>

Well-Differentiated Neuroendocrine Tumors of the Jejunum, Ileum, Appendix, and Cecum

J. Philip Boudreaux, MD, \* David S. Klimstra, MD, † Manal M. Hassan, MD, PhD,‡

Eugene A. Woltering, MD,\* Robert T. Jensen, MD, § Stanley J. Goldsmith, MD, || Charles Nutting, DO,¶

David L. Bushnell, MD,# Martyn E. Caplin, MD,\*\* and James C. Yao, MD;‡

Right hemicolectomy is recommended for ANEN>2cm or ANEN with high risk features





Neuro endocrinology

Neuroendocrinology 2019;109:179–186 DOI: 10.1159/000499381 Received: January 21, 2019 Accepted after revision: March 5, 2019 Published online: May 6, 2019

### The Carc

Challenging the Current Risk Factors of Appendiceal Neuroendocrine Neoplasms: Can

Michail Ga

They Accurately Predict Local Lymph Nodal Invasion? Results from a Large Case Series

Endo: https:

Michail Galanopoulos<sup>a</sup> Rory McFadyen<sup>b</sup> Ioanna Drami<sup>c</sup> Rishi Naik<sup>b</sup>

ME

Nicholas Evans<sup>b</sup> Tu Vinh Luong<sup>d</sup> Jennifer Watkins<sup>d</sup> Martyn Caplin<sup>a</sup> Christos Toumpanakis<sup>a</sup>

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neoplasms: when to perform a right hemicolectomy? A systematic review and meta-analysis

Claudio Ricci<sup>1,2</sup> · Carlo Ingaldi<sup>1,2</sup> · Laura Alberici<sup>1,2</sup> · Nicole Brighi<sup>2,3</sup> · Donatella Santini<sup>2,4</sup> · Cristina Mosconi<sup>2,5</sup> · Valentina Ambrosini<sup>2,3</sup> · Davide Campana<sup>1,2</sup> · Francesco Minni<sup>1,2</sup> · Riccardo Casadei<sup>1,2</sup>





#### **Pros**

- Guidelines supporting hemicolectomy for high risk patients
- No long-term follow-up data for conservative treatment
- Hemicolectomy is a simple procedure

#### Cons

- criteria for hemicolectomy are questionable
- Long-term benefit of hemicolectomy is not clear
- Surgical procedure with possible significant complications





#### **SUMMARY**

- NEN of the appendix are relatively rare
- Simple appendectomy is usually curative
- In rare cases, local or distant metastases appear
- Hemicolectomy is needed for "high risk" patients only
- Patient specific decision making by a NET expert

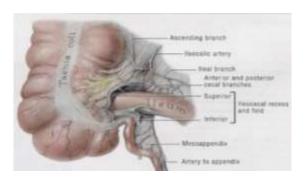


# NEN of the Appendix The patient

- 34 years old
- 2cm ANEN



- Imaging of the abdomen & pelvis
- Revision of the pathology specimen
- Shared decision making







### THANK YOU

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