



# **Statistics**

### · How common is pancreatic cancer?

- The American Cancer Society's estimates for pancreatic cancer in the United States for 2023 are:
  - About 64,050 people (33,130 men and 30,920 women) will be diagnosed with pancreatic cancer.
  - About 50,550 people (26,620 men and 23,930 women) will die of pancreatic cancer.
  - Pancreatic cancer accounts for about 3% of all cancers in the US and about 7% of all cancer deaths.
  - It is slightly more common in men than in women.
  - https://www.cancer.org/cancer/types/pancreatic-cancer/about/keystatistics.html



# Risk Factors

- · Tobacco use
- · Being overweight
- Diabetes
- Chronic pancreatitis
- Workplace exposure to certain chemicals
- Pancreatic Cancer Risk Factors | American Cancer Society

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### Survival Rates

### 5-year relative survival rates for pancreatic cancer

Based on people diagnosed with pancreatic cancer between 2012 and 2018.

| SEER* Stage              | 5-year Relative Survival Rate |
|--------------------------|-------------------------------|
| Localized                | 44%                           |
| Regional                 | 15%                           |
| Distant                  | 3%                            |
| All SEER stages combined | 12%                           |

 $^\star$  SEER = Surveillance, Epidemiology, and End Results

• <u>Survival Rates for Pancreatic Cancer | American Cancer Society</u>

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### Famous Personalities with pancreatic cancer **April Fritz** Alex Trebek Steve Jobs Sally Ride Ruth Bader Ginsburg Patrick Swayze Aretha Franklin Michael Landon John Hurt Joan Crawford Luciano Pavarotti Gene Upshaw (football) Sharon Jones (singer) Benjamin Orr (musician) Alan Rickman Charlotte Rae (actress) Bill Hicks (Comedian) Jerry Springer Jack Benny Donna Reed Marcello Mastroianni Dizzy Gillespie Pete Postlethwaite Count Basie Fred Gwynne (Herman Munster) Syd Barrett (musician Pink Floyd) Ben Gazzara Rex Harrison Alan Bates (Actor) Henry Mancini (composer) Keenan Wynn (Actor) Fernando Lamas (actor) NAACCR

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# **Clinical Trials**



- Clinical trials are carefully controlled research studies that are done to get a closer look at promising new treatments or procedures. Clinical trials are one way to get state-of-the art cancer treatment. In some cases they may be the only way to get access to newer treatments. They are also the best way for doctors to learn better methods to treat cancer.
- <a href="https://www.cancer.org/cancer/types/pancreatic-cancer/treating.html">https://www.cancer.org/cancer/types/pancreatic-cancer/treating.html</a>
- National Comprehensive Cancer Network Recommendations

Note: All recommendations are category 2A unless otherwise indicated.
Clinical Trials: NCCN believes that the best management of any patient with cancer is in a clinical trial. Participation in clinical trials is especially encouraged.

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# Current Clinical Trials for Pancreatic Cancer

- Testing the Use of the Usual Chemotherapy before and after Surgery for Removable Pancreatic Cancer
- Testing the Addition of Pembrolizumab, an Immunotherapy Cancer Drug to Olaparib Alone as Therapy for Patients with Pancreatic Cancer That Has Spread with Inherited BRCA Mutations
- APOLLO: A Randomized Phase II Double-Blind Study of Olaparib versus Placebo Following Curative Intent Therapy in Patients with Resected Pancreatic Cancer and a Pathogenic BRCA1, BRCA2 or PALB2 Mutation
- · Palbociclib and Binimetinib in RAS-Mutant Cancers, A ComboMATCH Treatment Trial
- CA-4948 Added to Standard Chemotherapy to Treat Metastatic or Unresectable Pancreatic Cancer
- The PLATINUM Trial: Optimizing Chemotherapy for the Second-Line Treatment of Metastatic BRCA1/2 or PALB2-Associated Metastatic Pancreatic Cancer
- 23 011 A Phase 1b Study of Odetiglucan with CDX-1140 Immunotherapy to Treat Metastatic Pancreatic Cancer
- 16 261 A Phase I Study of MVT-5873 Alone or with Chemotherapy in Patients with Pancreatic Cancer and Other CA19-9 Positive Tumors
- 20-481 A Phase II Study of Pembrolizumab Immunotherapy and OLApaRib (POLAR) Maintenance Therapy for Patients with Metastatic Pancreatic Cancer
- First in Human Phase 1/2 Trial of ELI-002 7P Immunotherapy as Treatment for Subjects with Kirsten Rat Sarcoma (KRAS)/Neuroblastoma RAS viral oncogene homolog (NRAS) Mutated Pancreatic Ductal Adenocarcinoma (PDAC) and Other Solid Timores

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# Poll 1

- Clinical trials should always be coded under "other" therapy.
- 1. True
- 2. False

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# **Coding Clinical Trials**

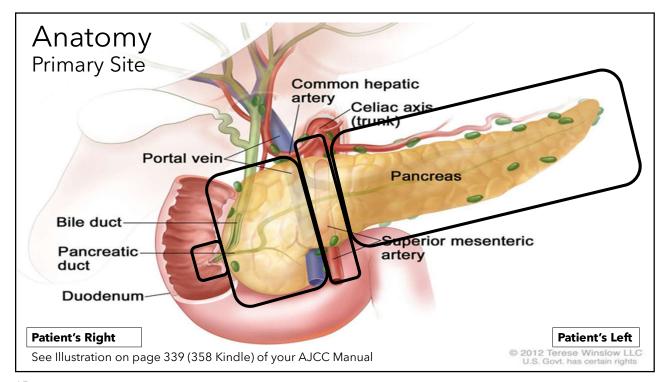
- If you KNOW the agents, code under that modality (i.e. chemotherapy, immunotherapy, hormone therapy, radiation).
- If the trial contains an agent not yet classified in SEER RX, then code it under Other therapy (1 or 2 other experimental)
- If it is double blind (neither the physician or the patient know what they are/are not receiving), then Other therapy 3.

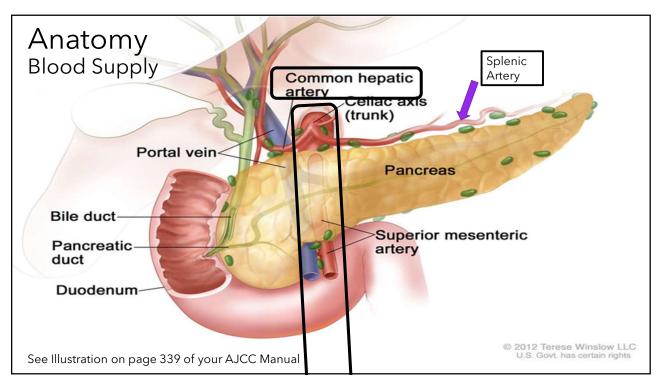
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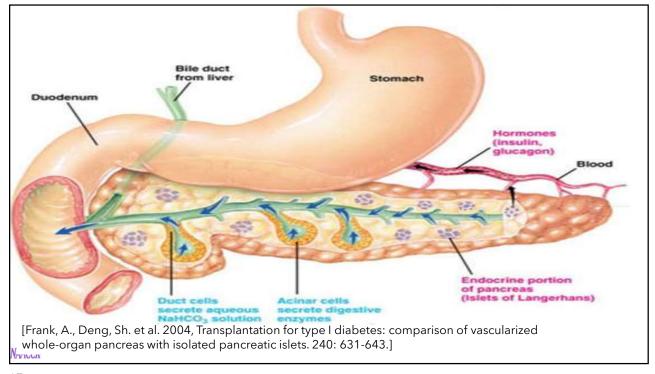
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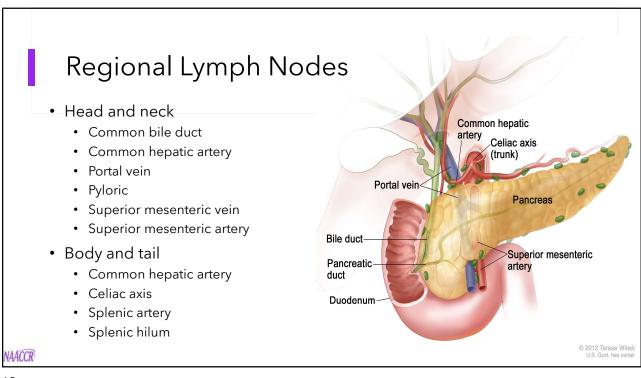
Overview

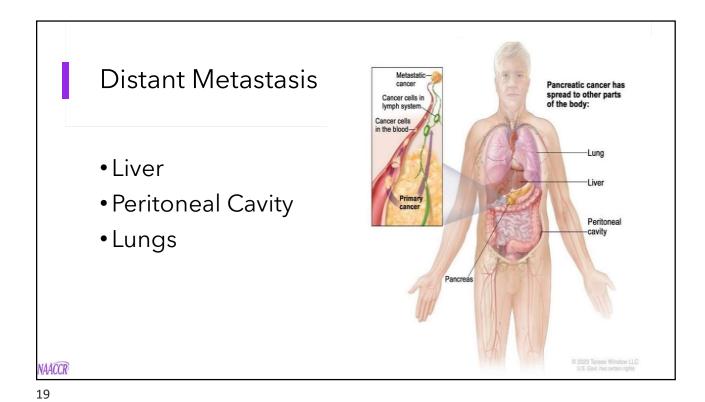
Anatomy, STR's, Staging, and Treatment











Histology-Exocrine Pancreas
Adenocarcinoma, NOS (8140/3)
Ductal adenocarcinoma (8500/3)
Most frequently occurs in head of pancreas
Arises in association with precursor lesions:
PanIN, IPMN or MCN
Acinar carcinoma (8550/3) or acinar cell cystadenocarcinoma (8551/3)
Adenosquamous (8560/3)
Neuroendocrine carcinoma (8246/3)

# Histology-NET Endocrine Pancreas

- Neuroendocrine tumor (8240/3)
  - Neuroendocrine tumor, grade 1
  - Neuroendocrine tumor, well differentiated
- Neuroendocrine tumor, grade 2 (8249/3)
- Neuroendocrine tumor, grade 3 (8249/3)

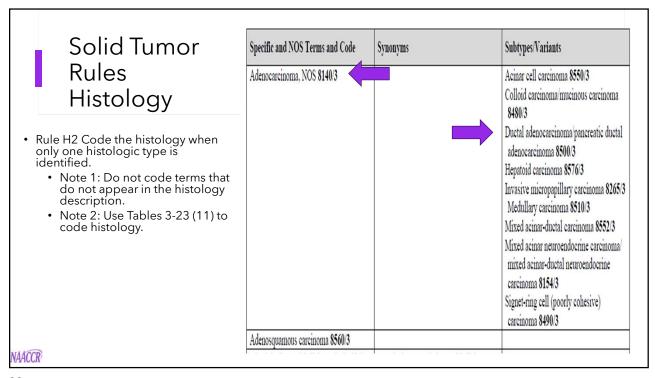
- Reportable as of 1/1/2021
  - Pancreatic neuroendocrine tumor, non-functioning (8150/3)
  - Insulinoma (8151/3)
  - Glucagonoma (8152/3)

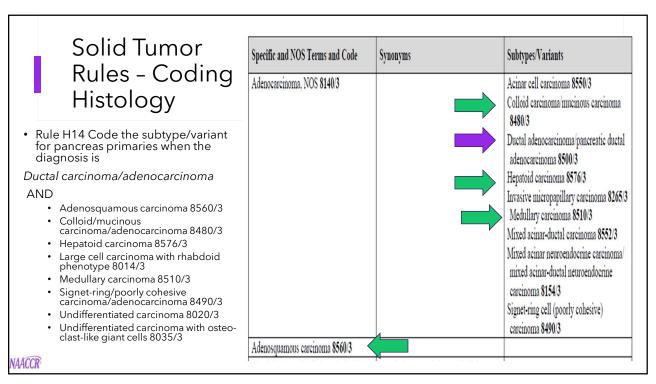
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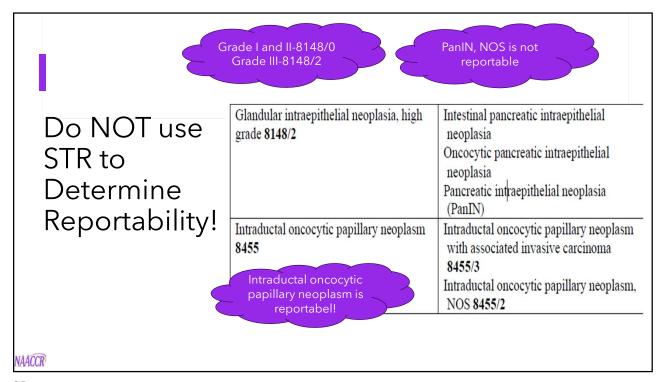
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# Solid Tumor Rules-Multiple Primary Rules

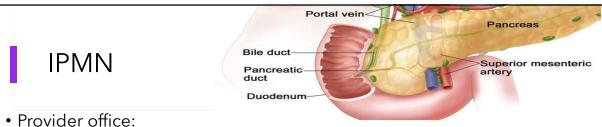
- **Rule M2** Abstract a single primary when there is a single tumor.
- **Rule M12** Abstract multiple primaries when the patient has a subsequent tumor after being clinically disease-free for greater than *one year* after the original diagnosis or recurrence.
- **Rule M17** Abstract multiple primaries when separate/non-contiguous tumors are two or more different *subtypes/variants in Column 3,* Table 3-23 in the Equivalent Terms and Definitions.
- **Rule M18** Abstract a single primary when synchronous, separate/non-contiguous tumors are on the *same row* in Table 3-23 in the Equivalent Terms and Definitions.
- **Rule M19** Abstract multiple primaries when separate/non-contiguous tumors are on *multiple rows* in Table 2-23 in the Equivalent Terms and Definitions. Timing is irrelevant
- Rule M20 Abstract multiple primaries when an invasive tumor occurs more than 60 days after an in situ tumor.
- **Rule M21** Abstract a *single primary* when there are multiple tumors that do not meet any of the above criteria.







|                     | Specific and NOS Terms and Code   | Synonyms  |
|---------------------|---|---|
|                     | Intraductal papillary mucinous neoplasm 8453  IPMN, NOS is not reportable | Intraductal papillary mucinous neoplasm with high grade-dysplasia 8453/2 High-grade IPMN 8453/2 Intraductal papillary mucinous carcinoma, non-invasive 8453/2 Intraductal papillary mucinous carcinoma, invasive 8453/3 Intraductal papillary mucinous neoplasm with associated invasive carcinoma 8453/3 |
|                     | Intraductal tubulopapillary neoplasm 8503                                 | Intraductal tubulopapillary neoplasm 8503/2 Intraductal tubulopapillary neoplasm with   |
| NAACCR <sup>®</sup> |   | Intrad  |

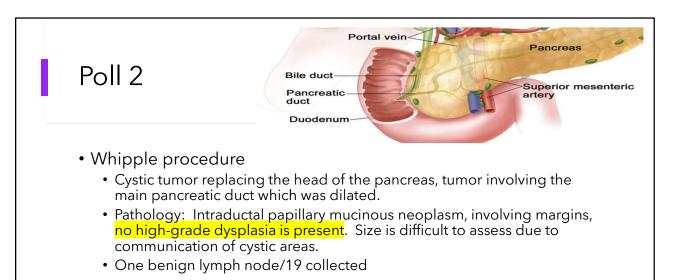


- - Patient underwent cholecystectomy in 2019.
  - Presents with abdominal pain, scans showed new onset of dilation of the pancreatic duct (we do not have scans).
  - Needs EUS.
- EUS:
  - Pancreas parenchyma was homogenous but appeared atrophic.
  - Pancreatic duct was dilated throughout the pancreas, up to 15 mm in the neck. Did not see a fish mouth sign.
  - There was a cystic change in the head of the pancreas 27 mm but think that was dilation of the pancreatic duct.
  - Did an EUS guided FNA but fluid was too thick and could only obtain a very small amount of fluid, did not see a mass.
  - Impression: IPMN (Intraductal papillary mucinous neoplasm)



# Fish Mouth Sign

- Patulous duodenal papilla with extrusion of mucus on endoscopic evaluation is a "fish mouth sign" and is reported with main duct and mixed IPMN
- https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6849671
- · Patulous wide open or distended



Is this case reportable?

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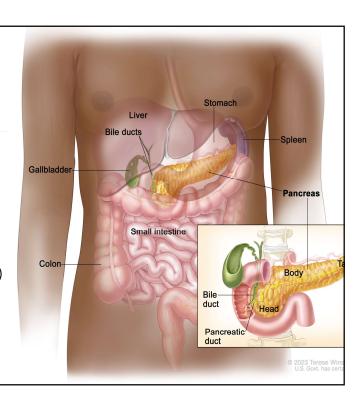
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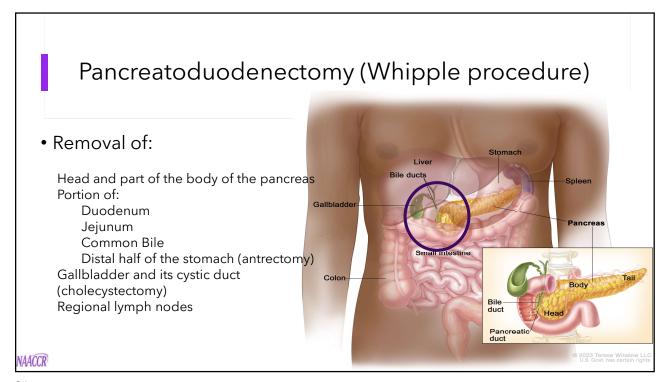
# Work-up

- Pancreatic protocol CT
- Magnetic Resonance (MR) imaging or MR cholangiopancreatography (MRCP)
- Endoscopic ultrasound (EUS)
- Endoscopic retrograde cholangiopancreatography (ERCP)
- Biopsy
  - CT guided
  - EUS guided (preferred)
- Laparoscopy





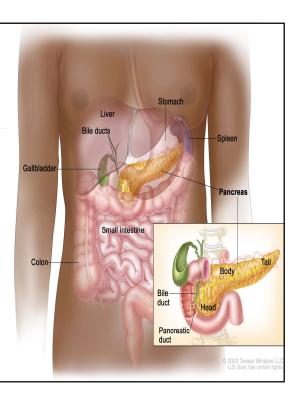


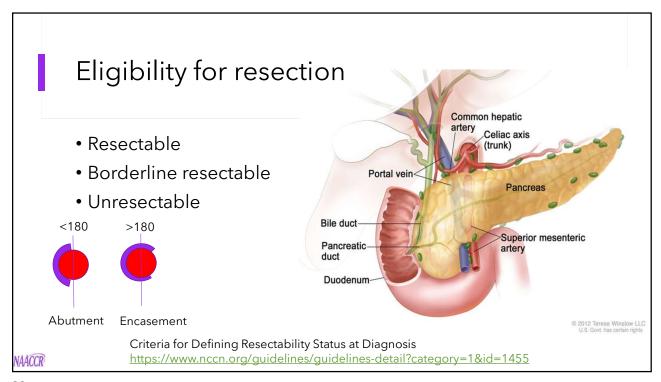


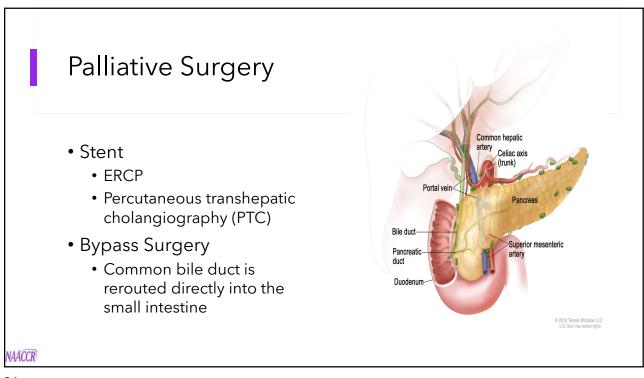
# Surgical Coding (2024)

- B300 Partial pancreatectomy, NOS; example: Distal pancreatectomy or subtotal pancreatectomy
- B350 Local or partial pancreatectomy and duodenectomy; example: Pancreaticoduodenectomy (Whipple Procedure)
  - B351 WITHOUT distal/partial gastrectomy, pylorus preserving Whipple
  - B352 WITH partial gastrectomy, Classic Whipple
  - Note: Use code B350 when it is not specified where the stomach was cut.
- B400 Total pancreatectomy
- B600 Total pancreatectomy and subtotal gastrectomy and/or duodenectomy, extended pancreatoduodenectomy

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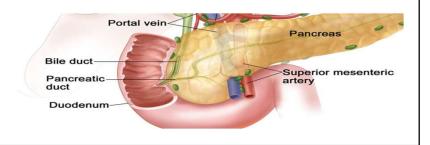






# Poll 3

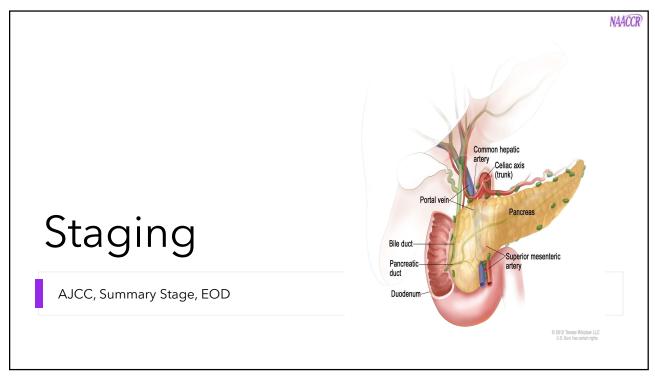
- A patient is taken to the OR for a Whipple procedure. Most likely the cancer is in what part of the pancreas?
- 1. Head
- 2. Body
- 3. Tail



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# Poll 4 • Nodes located along the portal vein would be regional for what part of the pancreas? • 1. Head/Neck • 2. Body/Tail • 3. Both

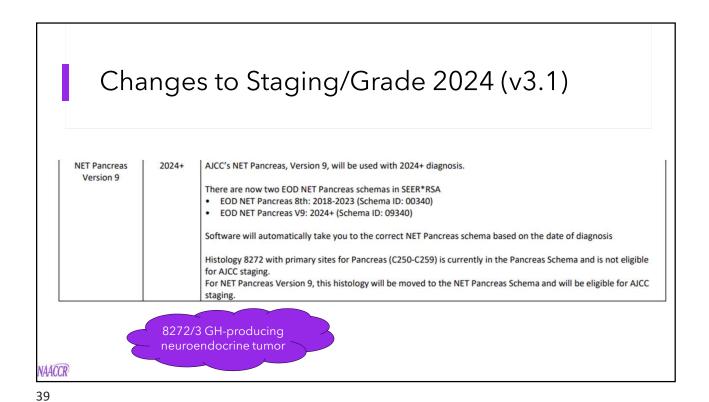


# New AJCC Version 9 Protocols

- 5.6 AJCC Version 9 Protocols AJCC Cancer Staging System will release seven Version 9 Protocols to go into effect with cases diagnosed January 1, 2024, and forward:
  - Vulva Version 9
  - Neuroendocrine Tumors of the Stomach Version 9
  - Neuroendocrine Tumors of the Duodenum and Ampulla of Vater Version 9
  - Neuroendocrine Tumors of the Jejunum and Ileum Version 9
  - Neuroendocrine Tumors of the Appendix Version 9
  - Neuroendocrine Tumors of the Colon and Rectum Version 9
  - Neuroendocrine Tumors of the Pancreas Version 9
- 2024-Implementation-Guidelines\_20230727.pdf (naaccr.org)

2024

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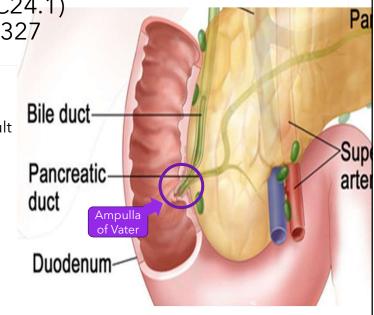
Ampulla of Vater (C24.1)
Chapter 27, page 327

• Highlights
• Primary site can be difficult

 Histology table
 8144-Adenocarcinoma, intestinal type

to determine

• 8163 Adenocarcinoma, pancreatobiliary type

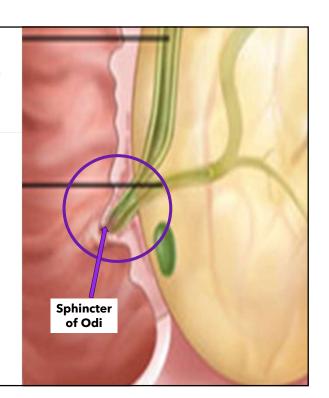


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# Ampulla of Vater (C24.1) Chapter 27, page 327

- T Values
  - Has the tumor spread beyond to ampulla of vater/sphincter of odi?
  - Has the tumor invaded into the duodenum?
    - How **deep** has the tumor penetrated the duodenum?
  - Has the tumor invaded into the pancreas?
    - How far has it invaded?
  - Is there invasion of major blood vessels?

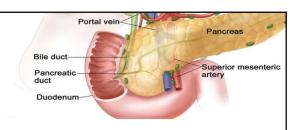


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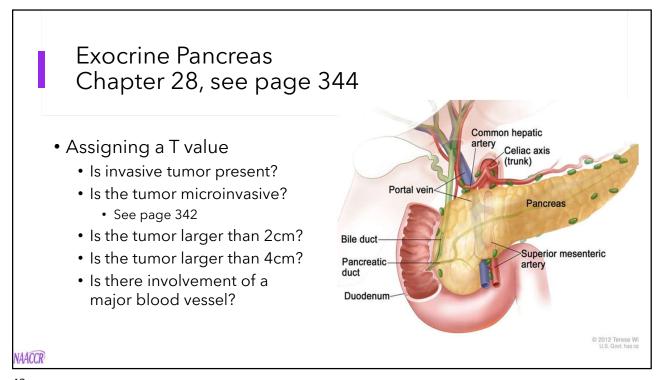
# Exocrine Pancreas Chapter 28, page 337

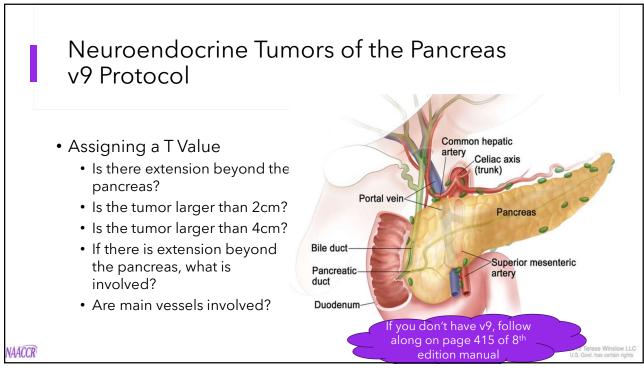
- Clinical Classification
  - Preoperative biopsy (pg 340 top of left column)
  - Abutment vs Encasement (pg 341 second paragraph)
  - Suggested Radiology Report Format



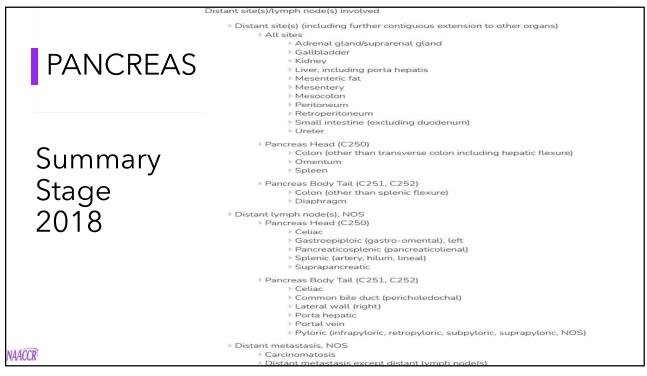
- Pathological Classification
  - Surgical Resection of the primary tumor and regional lymph nodes (pg 342)
  - Review of T categories (pg 342 right column, first full paragraph)
  - Review of N categories (pg 343, left column, second full paragraph)

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|                          | SS2018 | Description   |
|--------------------------|--------|---|
| PANCREAS                 | 0      | In situ, intraepithelial, noninvasive  > High-grade pancreatic intraepithelial neoplasia (PanIn-3)  > Intraductal papillary mucinous neoplasm with high grade dysplasia  > Intraductal tubulopapillary neoplasm with high grade neoplasm  > Mucinous cystic neoplasm with high-grade dysplasia  |
|                          | 1      | Localized only (localized, NOS)  > Confined to pancreas   |
| Summary<br>Stage<br>2018 | 2      | Regional by direct extension only  > All sites  > Ampulla of Vater  > Blood vessel(s) (major)  > Aortic artery  > Celiac artery  > Common hepatic artery  > Further contiguous extension to other major arteries  > Portal vein  > Superior mesenteric artery/vein  > Duodenum  > Extrahepatic bile duct(s)  > Fixation to adjacent structure(s), NOS  > Peripancreatic tissue, NOS  > Stomach  > Pancreas Head (C250)  > Adjacent stomach  > Blood vessel(s) (major)  > Gastroduodenal artery  > Transverse colon, including hepatic flexure  > Pancreas Body Tail (C251, C252)  > Spleen  > Spleen  > Splenic artery/vein |
| NAACCK'                  |        | > Splenic flexure   |



|                          | SS2018 | Description  |
|--------------------------|--------|--|
| NET<br>Pancreas          | 0      | In situ, intraepithelial, noninvasive  > High-grade pancreatic intraepithelial neoplasia (PanIn-3)  > Intraductal papillary mucinous neoplasm with high grade dysplasia  > Intraductal tubulopapillary neoplasm with high grade neoplasm  > Mucinous cystic neoplasm with high-grade dysplasia   |
|                          | 1      | Localized only (localized, NOS)  > Confined to pancreas  |
| Summary<br>Stage<br>2018 | 2      | Regional by direct extension only  All sites  Ampulla of Vater  Blood vessel(s) (major)  Acrtic artery  Celiac artery  Celiac artery  Common hepatic artery  Further contiguous extension to other major arteries  Portal vein  Superior mesenteric artery/vein  Dudenum  Extrahepatic bile duct(s)  Fixation to adjacent structure(s), NOS  Peripancreatic tissue, NOS  Stomach  Pancreas Head (C250)  Adjacent stomach  Blood vessel(s) (major)  Gastroduodenal artery  Transverse colon, including hepatic flexure  Pancreas Body Tail (C251, C252)  Spleen  Spleen  Spleenie artery/vein |
| NAACCR                   |        | > Splenic artery/vein  |

# Poll 5

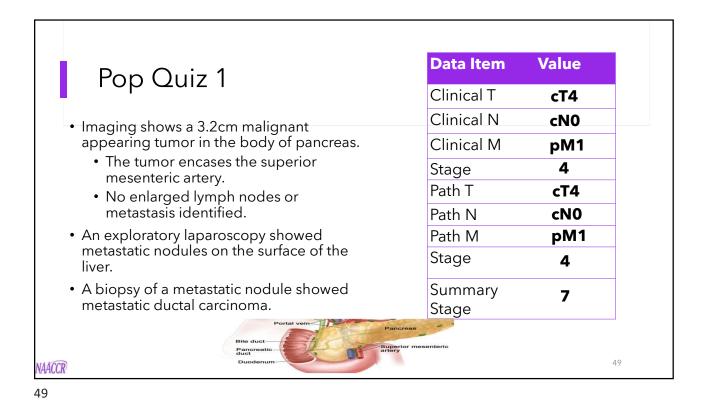
- Imaging shows a 3.2cm malignant appearing tumor in the body of pancreas.
  - The tumor encases the superior mesenteric artery.
  - No enlarged lymph nodes or metastasis identified.
- An exploratory laparotomy showed metastatic nodules on the surface of the liver.
- A biopsy of a metastatic nodule showed metastatic ductal carcinoma.



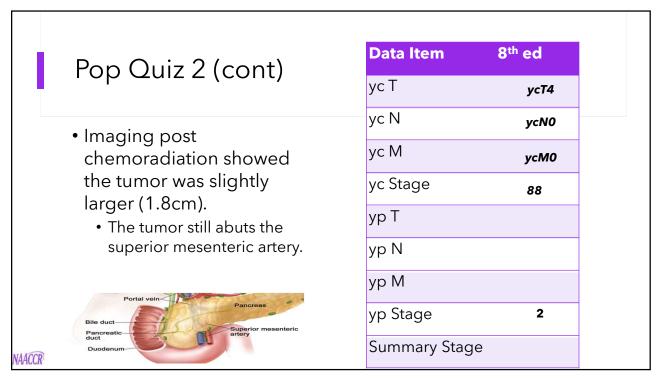
- "Encasement" identified on imaging indicates...
  - The tumor is near major artery and a clear margin of healthy tissue can be seen around the artery.
  - There is no space between the tumor and less than half the artery.
  - There is no space between the tumor and more than half the artery
  - None of the above.
- For staging purposes "Encasement" identified on imaging indicates...
  - The artery is involved
  - The artery is not involved

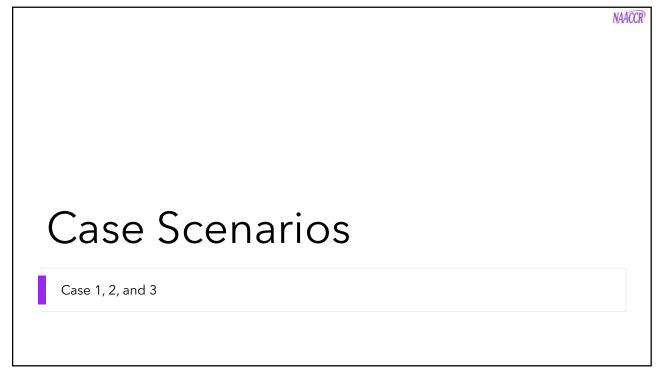
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**Data Item** Values Clinical T Pop Quiz 2 cT4 Clinical N cN0 Clinical M • Imaging shows a 1.4 cm tumor in the head of the pancreas. сМ0 • The tumor abuts the superior Stage mesenteric artery. There is less than 180° of involvement. No additional Pathological T blank arterial or celiac axis involvement. · No enlarged lymph nodes or Pathological N metastasis identified. blank An EUS-FNA confirms poorly Pathological M differentiated ductal adenocarcinoma blank The patient is treated with neoadjuvant Pathological chemoradiation. 99 Stage Summary Stage 2 NAACCR





Case #1

52 year old female went to the ER after an episode of hypotension, generalized abdominal pain and vomiting. She is otherwise in good health.

- CT abdomen/pelvis:
  - Well-circumscribed hypoenhancing 2.9 x 2.7 cm proximal pancreatic body mass, without upstream pancreatic ductal dilation or atrophy, possibly serous cystadenoma.
  - Neuroendocrine tumor considered less likely.

### MRI Abdomen:

- Enhancing mass identified in the pancreatic neck and proximal aspect of the pancreatic body measuring up to 2.7 cm.
- The lesion is not characteristic of an adenocarcinoma due to the lack of upstream dilatation of the main pancreatic duct and pancreatic atrophy. The pattern of enhancement is nonspecific. Correlation with an endoscopic ultrasound and tissue biopsy is advised.
- Differential diagnostic possibilities would include a neuroendocrine tumor and a pancreatic lymphoma although this is less likely due to the absence of lymphadenopathy and splenomegaly. No definitive evidence of liver metastases.

### • PET Scan:

- The focal lesion located in the pancreatic neck does not demonstrate significant somatostatin receptors radiotracer uptake, SUV 3.4 and measures approximately 2.4 x 2.4 cm.
- There is likely physiologic radiotracer activity in the pancreatic uncinate process with SUV 8.6

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Primary Site - what to do with conflicting information

CT abdomen/pelvis: Well-circumscribed hypoenhancing 2.9 x 2.7 cm proximal pancreatic body mass

MRI Abdomen: Enhancing mass identified in the pancreatic neck and proximal aspect of the pancreatic body measuring up to 2.7 cm.

PET Scan: The focal lesion located in the pancreatic neck

Operative Findings: Large mass at pancreatic head/neck overlying celiac axis

Path report: Tumor location - pancreatic body

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# Primary Site - conflicting information

### For Pancreatic Primaries:

- The "other" solid tumor rules do not provide guidance in coding primary site.
   The "general" STR do not provide guidance in coding primary site.
- The STORE manual refers you to ICD-O-3 to code primary site. Rules A through K do not provide information on what to do with conflicting information.
- SEER Program and Staging Manual does not provide information on what to do when there is conflicting information.

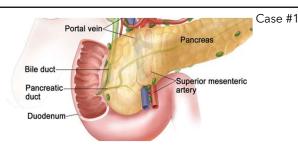
Must make your best judgement call based on all available information (including the type of surgery performed). Sometimes you may have an overlapping tumor (.8 subsite) or have very little information (.9 subsite).

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### • EUS:

- An irregular subtle poorly defined mass was identified in the genu of the pancreas.
- The mass was heterogeneous and solid. The echotexture was only slightly different compared to the pancreas parenchyma.
- The mass measured 33 mm x 24 mm in maximal cross-sectional diameter.
- The endosonographic borders were poorly defined.
- FNA:
  - Pancreas neck mass fine-needle, biopsy.
     Solid pseudopapillary neoplasm.
  - Note: Immunohistochemical stains supporting the diagnosis.



Per the Solid Tumor Rules and confirmed by SINQ (20140058), **Solid pseudopapillary neoplasm of the pancreas** is reportable.

Solid pseudopapillary neoplasm of pancreas 8452/3 Solid pseudopapillary carcinoma Solid pseudopapillary neoplasm with high-grade carcinoma

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Case #1

# Treatment Recommendations

- Second opinion at our facility:
  - Had a long discussion with the patient about her diagnosis, including pertinent anatomy, pathophysiology, differential diagnoses, treatment options.
  - Discussed differentials of pancreatic lesions including benign cysts, premalignant lesions, and malignant lesions.
  - I agreed with the other surgeon's evaluation in that she should undergo surgery.
  - However, I mentioned to the patient I would likely offer a laparoscopic subtotal pancreatectomy and splenectomy.
  - Discussed risks and benefits of both procedures, including among other things risk of pancreatic leak, diabetes, post splenectomy sepsis, bleeding, infection.

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# Surgery

Operative Procedure Performed:

- Laparoscopic subtotal pancreatectomy and splenectomy, intraoperative ultrasound
- Findings:
  - Large mass at pancreatic head/neck overlying celiac axis with associated desmoplasia, neovascularization, and distortion of tissue planes.
  - Ultrasound used to delineate medial edge, which was to the left of the GDA (gastroduodenal artery).
  - Subtotal pancreatectomy and splenectomy performed without complication.

# Pathology

**GROSS DESCRIPTION:** 

A. Specimen labeled distal pancreas and spleen is received fresh, excision and formalin time 1346/1404, cold ischemic time less than 1 hour and consists of a subtotal, left sided pancreas,  $12 \times 4 \times 2.1$  cm with a small, attached, fatty tissue at the tail and an impact spleen,  $7.7 \times 4.6 \times 2.8$  cm.

| * SYNOPTIC REPORT:                                 | Pancreas Exocrine tumor, invasive              |
|--|--|
| * Procedure:                                       | Subtotal pancreactectomy                       |
| * Tumor location:                                  | Pancreatic body                                |
| * Tumor size:                                      | Up to 2.7 cm                                   |
| * Type:  | Solid-pseudopapillary neoplasm                 |
| * Grade:   | Not applicable                                 |
| * Extent:  | Tumor is limited to pancreas                   |
| * Margins, invasive tumor:                         | Free 0.5 cm from the proximal resection margin |
| * Margins, dysplasia/intraepithelial<br>neoplasia: | Free   |
| * Treatment effect, primary site:                  | Not applicable                                 |
| * Lymphovascular invasion:                         | None   |
| * Perineural invasion:                             | Focally present                                |
| * Lymph nodes, # total:                            | 19   |
| * Lymph nodes, # involved:                         | 0  |
| * Distant metastases:                              | Not applicable                                 |
| * Stage (AJCC 8):                                  | p T2 N0 M (not applicable)                     |
| * MSI IHC and Interpretation:                      | Not applicable                                 |
| * Tumor block(s) for possible future studies:      | A4   |

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Case #1

Case #1

Grade Solid pseudopapillary neoplasm

| Code | Grade Description                      |
|------|--|
| 1    | G1: Well differentiated                |
| 2    | G2: Moderately differentiated          |
| 3    | G3: Poorly differentiated              |
| 9    | Grade cannot be assessed (GX); Unknown |

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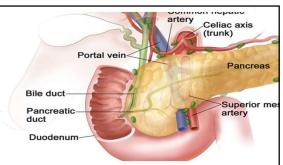
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# Additional Treatment

• Patient can follow-up with surveillance MRI in 6 months

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Celiac axis (trunk)

Pancrea

Superior mesenteric artery

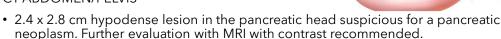
- Patient presented to the emergency department with complaints of diarrhea, abdominal pain, bloody urine, shortness of breath, and chest discomfort.
- Patient has a long-standing history of coronary artery disease with multiple stents as well as hypertension and hyperlipidemia status post angioplasty of the left anterior descending currently on aspirin. In addition, patient has a history of obstructive sleep apnea, COPD, diabetes and obesity as well as COVID in 2020.
- CT the abdomen performed on admission showed a subtle hypodense lesion in the pancreatic head measuring 2.8 cm suspicious for neoplasm.
- Intraductal biliary ductal dilatation with gallbladder distention was noted. Recommend outpatient follow-up with PET scan.

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# Imaging

### CT ABDOMEN/PELVIS



Portal vei

Pancreaticduct Duodenum

• Intrahepatic biliary ductal dilatation and gallbladder distention.

### MRI Abdomen:

- 3.7 x 2.8 x 3.6 cm mass in the superior pancreatic head/neck resulting in stricture in the common bile duct, inferior aspect situated approximately 2.7 cm from the level of the ampulla and spanning 1.2 cm in length; early narrowing of the main pancreatic duct.
- The mass abuts superior mesenteric vein, extrahepatic main portal vein, and gastroduodenal artery. No encasement of these vascular structures.
- No definitive intrahepatic metastases.

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Poll 6

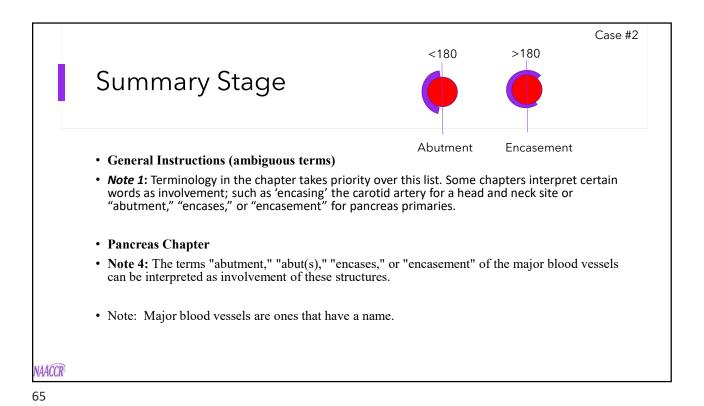
• For PANCREAS primaries - Does <u>abut</u> mean involved or not involved for Summary Stage?

• 1. Involved

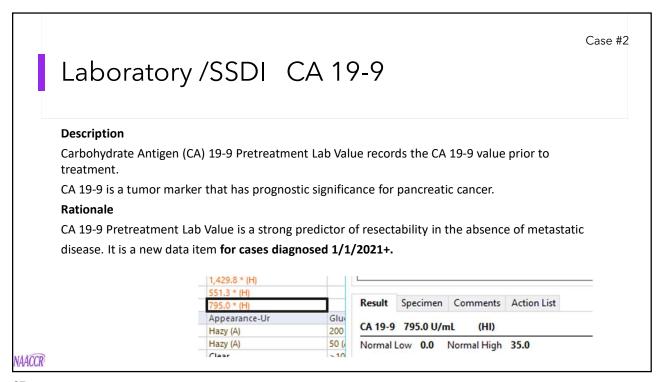
• 2. Not involved

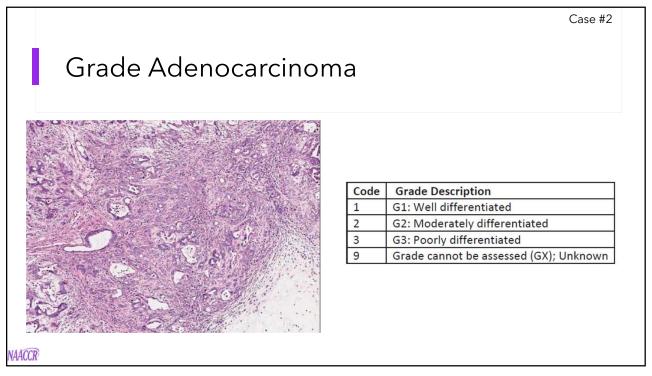
• 3. Not enough information

Case #2 Summary Stage Manual (ambiguous terms) Not Involved Abuts Extension to without invasion/involvement of Kiss/kissing Approaching Approximates Matted (except for lymph nodes) Attached Possible Cannot be excluded/ruled out Questionable Efface/effacing/effacement Reaching Encased/encasing Rule out Encompass(ed) Suggests Entrapped Very close to Equivocal Worrisome NAACCR



Case #2 Cancer-Type Relevant Biomarkers Office Artho Pathology report CNA-Seq: DNA-Tumoi Deletion Not Detected Exon 2 p.G12D DNA-Tumor Mutation Not Detected DNA-Tumos Deletion Not Detected Exon 12 p.W524C SMAD4 DNA-Tumor Deletion Not Detected DIAGNOSIS: DNA-Tumor Deletion Not Detected Mutation Not Detected PALB2 Mutation Not Detected DNA-Tumor · Pancreatic head mass fine DNA-Tumor Stable needle biopsy: PD-L1 (SP142) Protein Negative 0% Mismatch Renain · Invasive adenocarcinoma, moderately to poorly NTRK1/2/3 RNA-Tumor Fusion Not Detected CDKN2A DNA-Tumor differentiated. Exon 2 p.M52\_L64 Tumor Mutationa Low, 5 mut/Mb CNA-Seq DNA-Tumor Deletion Not Detected Exon 8 | p.E285K MTA : Mutation Not Detected DNA-Tumor Deletion Not Detected Mutation Not Detected NAACCR





• Oncology History:

Case #2

- Patient admitted with cholestasis found to have a mass on the head of the pancreas. EUS biopsy was performed and confirmed it to be adenocarcinoma, and a fully covered stent was placed. Admitted later with chest pain, and was found to have a subsegmental PE, also right inferior frontal gyrus infarction, now on Xarelto.
- Underwent neoadjuvant chemotherapy with FOLFIRINOX and completed 7 cycles, which also lead to cardiotoxicity. Ejection fraction is 25%, cardio cleared for surgery with a moderate risk and recommended intraoperative Swan-Ganz and fluid restriction.

### Multidisciplinary Tumor Board

• Tumor Board Treatment:

- Plan: Tumor board recommendation was to speak to the patient about ablative radiation therapy as a potential option. They also recommend speaking to risk management to be involved in the case, if needed.
- Discussion with Patient:
  - I explained to him that our typical institutional pathway for borderline resectable pancreas cancer includes induction chemotherapy followed by SBRT and then reassessment for surgery although given his high risk for surgery I would not recommend radiation therapy if he in fact did proceed with surgery as radiation could increase the risk of surgical complexity which would be detrimental to him given his cardiac condition.
  - He agreed to receive definitive ablative MR-guided SBRT instead of surgery.

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# **Treatment**



Case #2

- Treatment Summary Course # 1:
- Treatment was delivered to the pancreas, with photons and a(n) MR-guided SBRT technique on the MRIdian MRLinac (treatment unit). A total of 50 Gy was delivered at 10 Gy per fraction (treatment session). A total number of 5 fractions were delivered from the start date of XX/XX/XX to end date XX/XX/XX over 8 elapsed days.

| Treatment Site | Energy | Dose/Fx<br>(Gy) | #Fx | Total Dose<br>(Gy) | Start Date | End Date | Elapsed<br>Days |
|----------------|--------|-----------------|-----|--------------------|------------|----------|-----------------|
| Pancreas       | 6XFFF  | 10              | 5/5 | 50                 | 12/27/2022 | 1/4/2023 | 8               |

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# Additional Treatment Following FOLFIRINOX and SBRT

- ABDOMEN MRI W WO CON:
  - No significant change in pancreatic head mass with similar involvement of the GDA and common hepatic artery.
  - Multiple new rim-enhancing hepatic lesions with poorly defined margins favor to represent small abscesses over new metastatic disease.
  - Additional wedge-shaped regions of restricted diffusion in the liver may represent cholangitis. Consider short-term MRI after cholangitis treatment.
  - Unless otherwise specified, incidental findings in the body of the report may not need additional follow-up imaging.
- Tumor board: Reviewed liver lesions, given significantly elevated tumor marker 11,000 with new appearance of liver lesions, tumor board recommendations likely this is metastatic disease to liver now.
- Patient recommended to resume systemic treatment, but chose Hospice

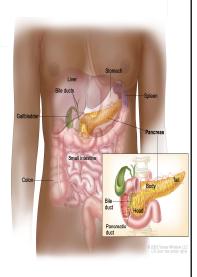
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Case #2

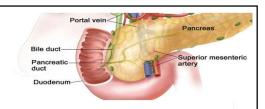
# Poll 7 Progression yes/no

- If the patient had chosen more treatment, would you code it?
  - Tumor board: Reviewed liver lesions, given significantly elevated tumor marker 11,000 with new appearance of liver lesions, tumor board recommendations likely is metastatic disease to liver now.
    - Remember the ambiguous terms list is only for histology and date of diagnosis not for staging, treatment, etc.
    - To determine progression, look for statement from treating physician, change in treatment, and consider all available information, not just a particular ambiguous term.



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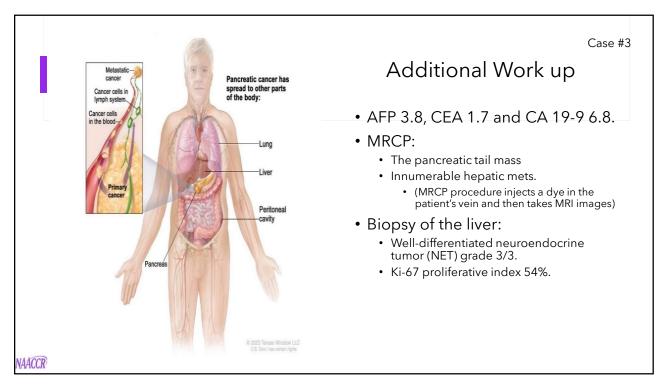
# Pancreas Case #3



- 56-year-old man with diabetes mellitus had progressive weakness, nausea, occasional vomiting, constipation, poor appetite and a 15-pound weight loss over the last 6 weeks. He presented to his PCP and had an abdominal ultrasound that identified multiple liver lesions
- CT Chest:
  - Gross hepatomegaly with widespread hepatic metastatic disease.
  - 5.4 x 4.1 x 5.3 cm mass lesion within or adjacent to the tail of the pancreas.
  - Splenic vein not well visualized and involvement cannot be excluded.
  - Obvious abdominal or retroperitoneal adenopathy is not identified.
- Ultrasound of the spleen:
  - Spleen is enlarged 15.4 cm, splenic vein is patent, no discrete splenic masses are seen.
- MRI:
  - Pancreatic tail mass 5.9 x 5.1 x 3.8 cm, no adenopathy.
  - Innumerable hepatic masses favoring mets.

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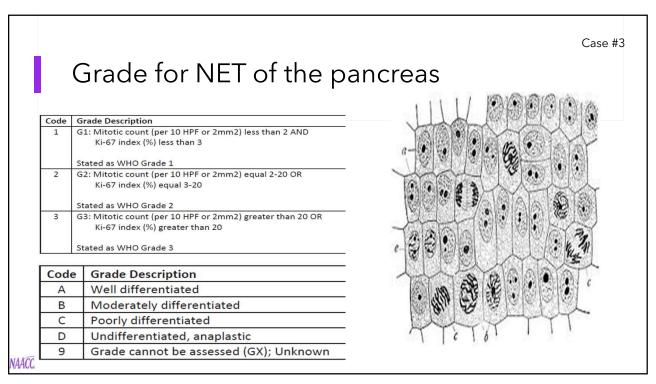
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### Case #3 SSDI NET Pancreas Code Description 0.0-100.0 0.0 to 100.0 percent positive: enter percent positive XXX.4 Ki-67 stated as less than 3% XXX.5 Ki-67 stated as 3%-20% XXX.6 Ki-67 stated as greater than 20% XXX.7 Test done; actual percentage not stated 8.XXX Not applicable: Information not collected for this case (If this item is required by your standard setter, use of code XXX.8 will result in an edit error.) XXX.9 Not documented in medical record Ki-67 (MIB-1) not assessed or unknown if assessed <Blank> N/A-Diagnosis year is prior to 2021

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Even though we do not collect BRCA2, note that many tumors now have genetic analysis to try and help guide treatment recommendations.

 Hepatic core biopsies: Metastatic well differentiated neuroendocrine tumor (NET) grade 3

| BIOMARKER | METHOD | ANALYTE   | RESULT                                   | THERAPY ASSOCIATION   | BIOMARKER<br>LEVEL*  |
|-----------|--------|-----------|--|---|--|
| BRCA2     | Seq    | DNA-Tumor | Pathogenic Variant<br>Exon 10   p.K43/fs | oxaliplatin olaparib A pathogenic or likely pathogenic BRCA2 muration, and/or del in the tumor for which germline status is negative or unavailab of therapy associations. The strongest evidence for DNA-dama PAPP inhibitors are platinum compounds comes from studies the predominantly germline mutations. Additionally, prescribing in consensus guidelines (e.g., NCCN) for PAPP inhibitors state are germline mutations. Therefore, the clinical benefit of these the comest of tumor/somatic-only mutations (including deletions) determined. | ole for interpretation<br>ging agents like<br>nat included<br>aformation and<br>quirement for<br>rapies in the |

Biomarker reporting classification: Level 1 – Companion diagnostic (CDX): Level 2 – Strong evidence of clinical significance or is endorsed by standard clinical guidelines.
 Level 3 – Potential clinical significance. Bolded benefit therapies, if present, highlight the most clinically significant findings.

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Case #3

### Treatment discussion

- This well-differentiated neuroendocrine tumor of the tail of the pancreas had a Ki-67 proliferative index of 54% which indicates a more aggressive behavior than well differentiated NET G1 or G2 but a better prognosis than neuroendocrine carcinomas of the pancreas (NEC).
- Patients with advanced NET G3 often have a relatively poor response to platinum plus etoposide
  regimens, and for this reason platinum-based chemotherapy may not be the most appropriate firstline treatment.
- My recommendation is to initiate chemotherapy with the CAPTEM regimen as soon as possible as the patient is symptomatic and has bulky liver disease.
- He would receive capecitabine 750 mg/m2 twice daily on days 1-14 and temozolomide 200 mg/m2 daily on days 10-14 of a 28-day cycle.
- I also recommend to order a Gallium Dotatate PET/CT scan to evaluate the uptake of the NET G3.
  - If there is Dotatate avidity, for second line treatment consideration should be given to therapy with Lutathera (PRRT).
  - Other potential treatments for second line upon disease progression include platinum based chemotherapy and immunotherapy with ipilimumab plus nivolumab.
- Sandostatin 30 mg IM every 4 weeks ordered to start AFTER patient completed PET DOTATATE (currently delayed due to limited insurance coverage). Port flush q8 weeks.

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## Gallium Dotatate PET scan

- Somatostatin receptor positive neuroendocrine tumors NET come from neuroendocrine cells, a large percentage of these cancers have receptors for somatostatin (a hormone). Octreotide is a protein that attaches to this somatostatin receptor.
- Gallium Dotatate a radioactive material, attached to octreotide and injected prior to PET scan. The labeled octreotide will attach to somatostatin receptors and help to delineate many neuroendocrine tumors as well as mets from the tumors.

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Case #3

# Explanation of treatment

- CAPTEM This regimen is not in SEER RX
- GOOGLE Per the NIH:
- Capecitabine and temozolomide (CAPTEM) regimen used for metastatic, well-differentiated neuroendocrine cancers
- Remember do not just look for the agents in the chemo flowsheet, but make sure you know everything included in the regimen in case there is an oral medication OR hormone that needs to be coded.

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# Lutathera

- SEER RX IMPORTANT INSTRUCTIONS: See STORE and <u>SEER</u> program manuals for <u>coding</u> instructions.
- Lutathera lutetium Lu 177 dotatate radioactive drug that attaches to the somatostatin receptors and the radiation helps to kill the cancer cells. (note: If the gallium dotatate PET scan indicates the cancer cells are taking the dye, then they are also likely to have the Lutathera attach and affect the cancer cells).
- Coded to isotopes NOS (13)
- Lutathera (lutetium LU 177) CAnswer Forum (facs.org)

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### Case #3 Name Octreotide Acetate Sandostatin Alternate Names L-Cysteinamide Minisomatostatin Sandostatin Sandostatin LAR Depot Abbreviations Code: Yes or No? Category Ancillary Agent Hormones and hormonal mechanisms Will start Sandostatin for carcinoid syndrome. I spoke to patient and Subcategory wife just now. I informed them of the Relief of symptoms NSC Number resulted 24H urine 5HIAA, serotonin level, CgA level, all elevated and **Primary Site** suggestive of carcinoid syndrome. None Interestingly, patient does not have Histology the classic carcinoid symptoms None (flushing, wheezing, diarrhea, Remarks abdominal pain), however, is still at $Remark\ added\ 11/18/2015: {\color{red}Sandostatin} \ us\ usually\ prescribed\ to\ treat\ side-effects/symptoms\ from\ TSH-secreting\ pituitary$ adenomas. Studies show this may also shrink tumors or inhibit further growth. If the physician states this agent is being risk for carcinoid heart prescribed to shrink or prohibit growth of the tumor, then code as hormone treatment Please note: not all drugs classified as hormone treat malignant neoplasms. NAACCR

# Additional Treatment

- Following 5 months on CAPTEM, repeat PET scan was done:
  - There is extensive disseminated FDG avid metastatic liver disease throughout both the right and left lobes of the liver which appears to have mildly increased in size when compared to prior PET/CT from 04/15/2022.
  - Numerous new or larger FDG avid lesions are identified.
  - Increased FDG activity corresponding to a lucent bone region in the medial right iliac bone with an SUV max of 2.8 that on prior study had an SUV max of 1.5, new 2.3 cm lytic lesion in the spinous process of the L3 vertebral body with an SUV max of 3.5, and new left femur lesion has an SUV max of 3.3.
- CAPTEM stopped due to progression and need for second line therapy.
- Second line therapy was Everolimus.

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# Coming UP...

- Boot Camp 1
  - Juliet Wilkins, MA, CTR
- Boot Camp 2
  - Nancy Etzold, CTR

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