Collecting Cancer Data:
Gastrointestinal Stromal Tumor (GIST)
Gastrointestinal Neuroendocrine Tumors (NET)

2013-2014 NAACCR Webinar Series

January 9, 2014

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Q&A

- Please submit all questions concerning webinar content through the Q&A panel.

Reminder:

- If you have participants watching this webinar at your site, please collect their names and emails.
  - We will be distributing a Q&A document in about one week. This document will fully answer questions asked during the webinar and will contain any corrections that we may discover after the webinar.

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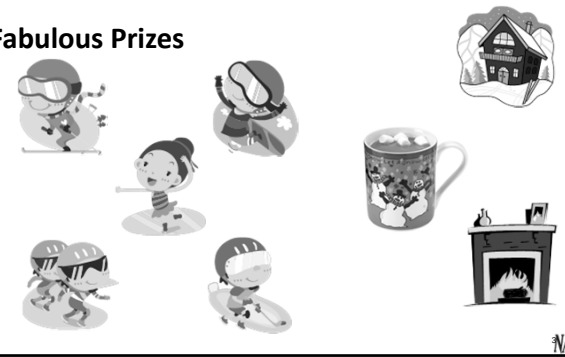
Fabulous Prizes



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Agenda

GIST
- Overview
- Collaborative Stage
- Treatment
- Quiz
- Case Scenarios 1-3

NET
- Overview
- Collaborative Stage
- Treatment
- Quiz
- Case Scenarios 4-5

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Gastrointestinal Stromal Tumors (GIST)

Overview

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Key Statistics

- Estimated new cases
  - 3,300 to 6,000
  - Compromise less than 1% of all gastrointestinal tumors

http://www.cancer.gov/cancertopics/types/soft-tissue-sarcoma

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What are GISTs?

- Rare type of soft tissue sarcoma
  - 4500-6000 adults (2009) – all sites
- Different from carcinomas
  - Develop in muscle layer of gut rather than mucosa
  - Grow outward (exophytic)
- Described as a distinct entity in 1998
  - Umbrella term for most mesenchymal tumors of stomach and intestine
  - Most tumors historically called leiomyosarcoma are now classified as GISTs

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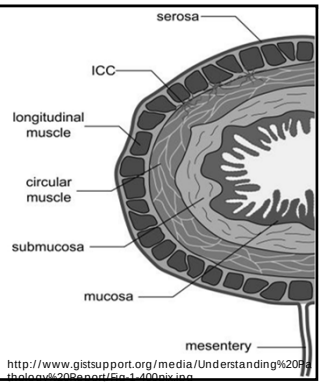
Proposed Cell of Origin

- Interstitial cells of Cajal
  - "Pacemaker cells of gut"
  - Send signals to muscles of GI tract to move food and liquid through system (peristalsis)



http://www.gistsupport.org/media/Understanding%20Pathology%20Report/Fig-1-400pix.jpg

Oncogenic Mutations

- ~85% of GIST contain oncogenic mutations in one of two receptor tyrosine kinases
  - KIT-Mutant GIST
  - PDGFRA (Platelet-derived Growth Factor Receptor Alpha)
- Wild Type GIST
  - ~12-15% of GIST contain no genetic mutation of KIT or PDGFRA.

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KIT (CD117)

- The KIT gene produces a tyrosine kinase enzyme that helps to regulate cellular activity (cell division)
  - A mutation in the KIT gene can produce enzymes that cause unregulated cell growth.
    - Mutations primarily of exon 11 and 9, and rarely of exons 13 and 17
  - An overproduction of CD117 can indicate a mutation of the KIT gene.
  - ~80% of all GIST contain a mutation in the KIT receptor tyrosine kinase

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PDGFRA

- ~5% to 8% of GIST harbor a mutation in PDGFRA
  - Like KIT, a mutation of PDGFRA can cause unrestricted cell growth

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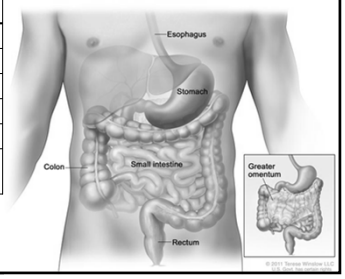
Tumor Location

Primary Site	% of all GIST's
Stomach	60%
Small Intestine	30%
Rectum	3%
Colon	1-2%
Esophagus	<1%
Omentum/ mesentery	Rare



Tumor Size

- T1 Tumor 2cm or less
- T2 Tumor more than 2cm, but not more than 5cm
- T3 Tumor more than 5cm, but not more than 10cm
- T4 Tumor more than 10cm in greatest dimension

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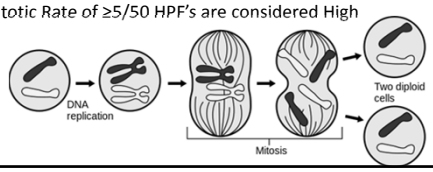
Mitotic Rate

- A measure of how fast cancer cells are dividing and growing. To find the mitotic rate, the number of cells dividing in a certain amount of cancer tissue is counted.
  - Mitotic Rate of ≤ 5/50 HPF's are considered Low
  - Mitotic Rate of ≥5/50 HPF's are considered High



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http://www.cancer.gov/cancertopics/pdq/treatment/gist/HealthProfessional/Table1

Table 1. Risk Stratification of Primary GIST by Mitotic Index, Tumor Size, and Tumor Location[a]

Mitotic Index, hpf	Size, cm	Site and Risk of Progressive Disease (%)			
		Gastric	Duodenum	Jejunum/Ileum	Rectum
≤5 per 50	≤2	None (0)	None (0)	None (0)	None (0)
	>2 ≤5	Very low (1.9)	Low (4.3)	Low (8.3)	Low (8.5)
	>5 ≤10	Low (3.6)	Moderate (24)	(Insufficient data)	(Insufficient data)
	>10	Moderate (10)	High (52)	High (34)	High (57)
>5 per 50	≤2	None	High[b]	(Insufficient data)	High (54)
	>2 ≤5	Moderate (16)	High (73)	High (50)	High (52)
	>5 ≤10	High (55)	High (85)	(Insufficient data)	(Insufficient data)
	>10	High (86)	High (90)	High (86)	High (7)

GIST = gastrointestinal stromal tumors; hpf = high-power field, assessed from an area that on initial screen appears to have the highest mitotic activity.

[a]Annual review of pathology by ANNUAL REVIEWS, INC. Reproduced with permission of ANNUAL REVIEWS, INC., in the format Internet posting via Copyright Clearance Center.[2]

[b]Small numbers of cases.

References

2. Corless CL, Heinrich MC: Molecular pathobiology of gastrointestinal stromal sarcomas. Annu Rev Pathol 3: 557-86, 2008. [PUBMED Abstract]

Gastrointestinal Stromal Tumors (GIST)

- What is the difference between GIST, NOS and a malignant GIST?
  - GIST, NOS 8936/1
  - Malignant GIST 8936/3

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Question

- Are there criteria other than a pathologist or clinician's statement that a registrar can use to determine reportability of gastrointestinal stromal tumors (GIST)?

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Answer

- Per SINQ 20091021 and 20021151, GIST cases are not reportable unless they are stated to be malignant.
  - A pathologist or clinician must confirm the diagnosis of cancer. There are cases that are not stated to be malignant in the pathology report or confirmed as such by a clinician; however, these cases do have information that for other primary sites would typically be taken into consideration when determining reportability.
    (SEER SINQ 20100014)

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Question

- Pathologists have used tumor size and mitotic activity to determine whether GISTS were benign or malignant. The 7th Edition AJCC Manual uses criteria for Stage 1 GIST which would otherwise be considered benign.
  - Could you clarify if we are to go by staging criteria to determine if a GIST is reportable?

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Answer

- The CoC requires to report all sites malignancies with behavior 2 and 3, except skin cancers 8000-8110, CIS, intraepithelial neoplasia grade III (8077/2) of the cervix (CIN III), prostate (PIN III), vulva (VIN III), vagina (VAIN III), and anus (AIN III).
  - Benign GIST is not reportable since the behavior is 0.
  - However, if your facility or your state requires to collect benign GIST, you should follow their requirements.
  - When you submit the data, make sure you do not include benign GIST in your data file submitted to NCDB (CoC), otherwise it will be rejected.

http://cancerbulletin.facs.org/forums/showthread.php?449-Is-benign-GIST-reportable-to-the-CoC&highlight=GIST

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Question

- Our cancer committee has decided that we should collect ALL GIST tumors.
  - In the event that a GIST that we have abstracted becomes malignant and thus is now reportable to NCDB, how should be handle this case?

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Question (cont)

- Would we create a new abstract with the date of diagnosis being the date the physician states the case is malignant and thus the patient would have two abstracts?
- The first would have a sequence code of 60 and a behavior code of 1?
- The second would have a sequence code of 00 (if it was the first malignancy) and behavior code of 3 and a different date of diagnosis?

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Answer

- Yes, to your last three questions.

http://cancerbulletin.facs.org/forums/showthread.php?3214-GIST-Tumors&highlight=GIST

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GIST Histology

- Stromal sarcoma, NOS 8935/3
- Gastrointestinal stromal sarcoma 8936/3
  - Gastrointestinal stromal tumor, malignant
  - GIST Malignant

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Multiple Primary and Histology Rules

- Use the site where the tumor originated to determine MPH chapter to use.

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GIST Multiple Primary Rules

Stomach	Other
Small intestine	Other
Esophagus	Other
Large intestine	Colon
Rectum	Other
Other (very rare)	Other

- Peritoneum, mesentery, omentum, liver, pancreas, ovaries, uterus, prostate

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Staging Systems

GIST

CS Schemas for GIST

- GIST Appendix
- GIST Colon
- GIST Esophagus
- GIST Peritoneum
- GIST Rectum
- GIST Small Intestine
- GIST Stomach

AJCC Cancer Staging: GIST

- 2 Anatomic Stage/Prognostic Groups
  - Gastric GIST
    - Stomach, omentum
  - Small Intestinal GIST
    - Small intestine, appendix, colon, rectum, esophagus, peritoneum (except omentum)
- Anatomic Stage/Prognostic Group
  - Determined by T, N, M, and mitotic rate
- Staging scheme includes all GIST
  - Staging does not determine reportability or ICD-O-3 behavior code

CS Tumor Size: GIST

- AJCC T categories based on tumor size
  - T1: 2 cm or less
  - T2: More than 2 cm but not more than 5 cm
  - T3: More than 5 cm but not more than 10 cm
  - T4: Tumor more than 10 cm in greatest dimension

CS Tumor Size: GIST

Code	Description
000	No mass/tumor found
001-988	001-988 mm; Exact size to nearest mm
989	989 mm or larger
990	Microscopic focus or foci only & no size of focus given
991	Described as less than 1 cm
992	Described as less than 2 cm or greater than 1 cm or between 1 & 2 cm Stated as T1 with no other information on size
993	Described as less than 3 cm or greater than 2 cm or between 2 & 3 cm
994	Described as less than 4 cm or greater than 3 cm or between 3 & 4 cm

CS Tumor Size: GIST

Code	Description
995	Described as less than 5 cm or greater than 4 cm or between 4 & 5 cm Stated as T2 with no other information on size
996	Described as less than 10 cm or greater than 5 cm or between 5 & 10 cm Stated as T3 with no other information on size
997	Described as greater than 10 cm Stated as T4 with no other information on size
999	Unknown; size not stated Size of tumor cannot be assessed Not documented in patient record

CS Extension: GIST

- CS Extension codes are based on depth of invasion
- AJCC does not include in situ category for GIST
  - CS Extension code 000 maps to TX
- Ignore intraluminal extension
- AJCC T categories for GIST are based on tumor size
- Use code 150 (invasive tumor in polyp) only if GIST is described as arising in polyp
  - Appendix, colon, rectum, small intestine, stomach

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CS Extension: GIST

- AJCC Cancer Stage
  - T1, T2, T3, T4 categories based on tumor size
    - CS Extension code = 100-800

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CS Extension: GIST
Appendix, Colon, Rectum, Small Intestine

- Summary Stage 2000:
  - In situ: Noninvasive; intraepithelial
    - CS Extension = 000
  - Localized (L): Confined to organ
    - CS Extension = 150-440
  - Regional by direct extension (RE): Extension to adjacent tissues and/or organs
    - CS Extension = 450-680
  - Distant extension (D): Extension to distant organs
    - CS Extension = 700-800

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CS Extension: GIST Esophagus

- Summary Stage 2000
  - In situ: Noninvasive; intraepithelial
    - CS Extension = 000
  - Localized (L): Confined to esophagus
    - CS Extension = 155-300
  - Regional by direct extension (RE): Extension to adjacent tissues and/or organs
    - CS Extension = 400-650, 730
  - Distant extension (D): Extension to distant organs
    - CS Extension = 660-680, 740-810

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CS Extension: GIST Peritoneum

- Summary Stage 2000
  - Localized (L): Confined to site of origin
    - CS Extension = 100-380
  - Regional by direct extension (RE): Extension to adjacent tissues and/or organs
    - CS Extension = 400-600
  - Distant extension (D): Extension to distant organs
    - CS Extension = 800

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CS Extension: GIST Stomach

- Summary Stage 2000
  - In situ: Noninvasive; intraepithelial
    - CS Extension = 000
  - Localized (L): Confined to stomach
    - CS Extension = 150-440
  - Regional by direct extension (RE): Extension to adjacent tissues and/or organs
    - CS Extension = 450-610
  - Distant extension (D): Extension to distant organs
    - CS Extension = 650-800

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CS Lymph Nodes: GIST

- Code regional node involvement
  - Lymph node metastasis is rare for GIST
  - Nodes considered regional are based on site of GIST
  - AJCC N1
  - Summary Stage 2000 RN

CS Mets at DX: GIST

- Record distant metastasis at time of diagnosis in distant lymph nodes and/or organs and tissues
  - Based on site of GIST
  - AJCC M1
  - Summary Stage 2000 D
- Record liver metastasis in liver parenchyma
  - Adherence to liver capsule is NOT recorded in CS Mets at DX
- Distant metastasis are relatively rare for GIST

CS Mets at DX: GIST

- Differentiate between
  - Intra-abdominal metastasis
    - Involvement in abdominal cavity outside of the main tumor mass in the peritoneum, omentum, serosae of organs, and cul-de-sac, among others
    - Record in CS Mets at DX
  - Tumor multiplicity
    - Anatomically separate multiple tumors of different sizes arising independently
      - Solitary omental or mesenteric tumor mass
    - DO NOT record in CS Mets at DX

Pop Quiz

- Exploratory laparotomy, hepatic wedge resection, hemigastrectomy, omentectomy.
- Operative findings: Gastric mass, 6 cm, on lesser curvature antral area of stomach.
- Path findings:
  - Stomach: Epithelioid pleomorphic GIST, malignant.
  - Omentum: Small epithelioid GIST (1 mm).
  - Liver: Negative for tumor.

Pop Quiz

- What is the code for CS Tumor Size?
  - 001
  - 060
  - 996: Described as "less than 10 cm," or "greater than 5 cm" or "between 5 cm and 10 cm"
  - 999: Unknown

Pop Quiz

- What is the code for CS Extension?
  - 300: Implants inside stomach; Localized NOS
  - 395: Stated as T3 with no other information on extension
  - 450: Extension to adjacent (connective) tissue WITHOUT perforation of visceral peritoneum: Gastric artery Ligaments: Gastrocolic, Gastrohepatic, Gastrosplenic; Omentum, NOS: Greater, Lesser; Perigastric fat
  - 999: Unknown

Pop Quiz

- What is the code for CS Lymph Nodes?
  - 000: No regional node involvement
  - 050: Nodule in perigastric fat
  - 999: Unknown

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Pop Quiz

- What is the code for CS Mets at DX?
  - 00: No distant metastasis
  - 40: Distant metastasis except distant lymph nodes including: peritoneal nodules, liver parenchymal nodules; carcinomatosis; malignant peritoneal cytology
  - 60: Distant metastasis NOS
  - 99: Unknown

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SSFs for GIST

- Mitotic Count
  - SSF5 for GIST Peritoneum
  - SSF6 for GIST Esophagus, GIST Small Intestine, GIST Stomach
  - SSF11 for GIST Appendix, GIST Colon, GIST Rectum
- KIT Gene Immunohistochemistry (IHC)
  - SSF6 for GIST Peritoneum
  - SSF7 for GIST Esophagus, GIST Small Intestine, GIST Stomach
  - SSF12 for GIST Appendix, GIST Colon, GIST Rectum
- Location of Primary Tumor
  - SSF10 for GIST Peritoneum

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Mitotic Count

- Describes the potential aggressiveness of the tumor
- Determines histologic grade
  - Low or high
- Used with T, N, and M categories to stage group
- Code the specific mitotic count per 50 HPF to the nearest tenth of a mitosis as documented in path report
- Use code 996 if denominator is something other than 50 HPF

KIT Gene Immunohistochemistry (IHC)

- KIT gene regulates cell growth and differentiation
- Presence of KIT gene
  - Confirms diagnosis of GIST
  - Indicates if a patient will respond to Gleevec or Sutent
- Record interpretation from KIT test IHC stains
  - Path report
    - Positive/elevated
    - Negative/normal
    - Borderline
  - May be called CD117

Pop Quiz

- Exploratory laparotomy and hemigastrectomy path report: 6 cm gastric tumor on lesser curvature of stomach, malignant GIST.
  - Addendum: IHC KIT is negative. High mitotic rate of 10/50 HPFs.

Pop Quiz

- What is the code for SSF6 (Mitotic Count)?
  - 001
  - 010
  - 100
  - 998: No histologic specimen from primary site
- What is the code for SSF7 (KIT Gene IHC)?
  - 010: Positive/elevated
  - 020: Negative/normal
  - 030: Borderline
  - 998: Test not done

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Location of Primary Tumor GIST Peritoneum

- All peritoneum structures coded to C48.1 but 2 stage tables derive TNM values
  - GISTStomach
  - GISTSmallIntestine

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Location of Primary Tumor GIST Peritoneum

Code	Description	Stage Table
010	Mesentery; Mesoappendix; Mesocolon	GISTSmallIntestine
020	Omentum	GISTStomach
030	Pelvic Peritoneum	GISTSmallIntestine
040	Rectouterine pouch Cul de sac; Pouch of Douglas	GISTSmallIntestine
988	Not applicable	ERROR
998	Other specified peritoneal site	GISTSmallIntestine
999	Unknown	GISTSmallIntestine

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GIST Work-up

- Imaging
  - Contrast enhanced CT
  - PET scan
- Biopsy of primary site preferred over percutaneous biopsy
  - Tumors are soft and fragile
  - Risk of hemorrhage and intra abdominal dissemination

GIST Treatment

- Surgical resection
  - Based on primary site and extent of disease
  - Complete surgical resection possible in 80+% of patients
- Chemotherapy-Tyrosine Kinase Inhibitor (TKI) therapy
  - Gleevec (imatinib)
    - May be neoadjuvant, adjuvant or primary treatment
  - Sutent (sunatinib) for Gleevec-refractory or intolerant cases
- For distant metastases
  - Liver: wedge resections, RFA, cryosurgery, chemoembolization

Case Scenarios

Scenarios 1-3

Gastrointestinal Neuroendocrine Tumor (NET)

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Key Statistics

- The age-adjusted incidence of carcinoid tumors worldwide is approximately 2 per 100,000 persons.
- The average age at diagnosis is 61.4 years.
- Carcinoid tumors represent about 0.5% of all newly diagnosed malignancies

http://www.cancer.gov/cancertopics/types/gastrointestinalcarcinoid

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Neuroendocrine Cells

- Cells do not form an organ
  - Single cells or small clusters scattered throughout other organs
    - Lungs, stomach, and intestines
  - Occur in aggregates or sheets within other organs
    - Islets in pancreas or medullary portion of adrenal
  - Form small collections of cells called “bodies”
    - Carotid body or glomus jugulare

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Neuroendocrine Tumors

- Derived from neuroendocrine cells
  - A bridge between the body's endocrine system and nervous system
  - Found in almost every organ
- Malignant neuroendocrine tumors may secrete hormones in excess, causing a variety of symptoms

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Carcinoid Histologies

- Neuroendocrine carcinoma, NOS (8246)
  - General term covering carcinoids and some adenocarcinomas
- Carcinoid, NOS (8240)
  - Typical carcinoid, low grade or well-differentiated neuroendocrine carcinoma
  - Usually occur in rectum, appendix; uncommon in colon
  - Rarely metastasizes
  - Favorable prognosis if size is < 2 cm

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Carcinoid Histologies

- Enterochromaffin (EC) cell carcinoid (8241)
  - Produces serotonin
- ECL cell tumor (Entero-Chromaffin-Like) (8242)
  - Non-secreting tumor
- Atypical carcinoid tumor (8249)
  - More aggressive than a typical carcinoid
  - Primarily found in the respiratory tract

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Other NETs

- Gastrinoma, malignant (8153/3)
  - May appear anywhere in the gastro intestinal tract
  - Over produce gastric acid and peptic ulceration

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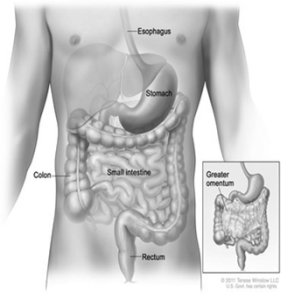
Location



- Foregut 25%
  - Lung, thymus, stomach, proximal duodenum
- Midgut 50%
  - Small intestine, appendix, or proximal colon, with the appendix being the most common site of origin
- Hindgut 15%
  - Distal colon or rectum
- Other
  - gallbladder, kidney, liver, pancreas, ovary, and testis

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Multiple Primary and Histology Rules

- Colon
- Other

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Colon MP/H

- Neuroendocrine carcinoma (8246): Neuroendocrine carcinoma is a group of carcinomas that include typical carcinoid tumor (8240), atypical carcinoid tumor (8249).

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Colon MP/H

- Rule H8
  - Code 8240 (carcinoid tumor, NOS) when the diagnosis is neuroendocrine carcinoma (8246) and carcinoid tumor (8240).
- Rule H9
  - Code 8244 (composite carcinoid) when the diagnosis is adenocarcinoma and carcinoid tumor.
- Rule H10
  - Code 8245 (adenocarcinoid) when the diagnosis is exactly "adenocarcinoid."

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Other MP/H

- Stomach
- small intestine
- Pancreas
- Thyroid gland
- Adrenal gland
- Thymus
- Heart
- Other sites that develop carcinoids and small cell carcinomas
- Skin

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Staging Systems

NET

CS Schemas for NET

- NET Ampulla
- NET Colon
- NET Rectum
- NET Small Intestine
- NET Stomach

CS: NET

- AJCC only stages well differentiated neuroendocrine tumors
  - Grade code not needed to select correct schema
- NET schemas used for carcinoid tumors and gastrinomas

CS Tumor Size: NET

- AJCC T categories based on tumor size
  - NET Ampulla
    - T1: 1 cm or less
    - T2: More than 1 cm
  - NET Colon, Net Rectum
    - T1: Invades lamina propria or submucosa and 2 cm or less
      - T1a: Less than 1 cm
      - T1b: 1-2 cm
  - NET Small Intestine
    - T1: Invades lamina propria or submucosa and 1 cm or less
  - NET Stomach
    - T1: Invades lamina propria or submucosa and 1 cm or less

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CS Tumor Size: NET

Code	Description
000	No mass/tumor found
001-988	001-988 mm; Exact size to nearest mm
989	989 mm or larger
990	Microscopic focus or foci only & no size of focus given
991	Described as less than 1 cm Stated as T1 with no other information on size (NET Ampulla, NET Small Intestine, NET Stomach) Stated as T1a with no other information on size (NET Colon, NET Rectum)

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CS Tumor Size: NET

Code	Description
992	Described as less than 2 cm or greater than 1 cm or between 1 & 2 cm Stated as T1b or T1 NOS with no other information on size (NET Colon, NET Rectum) Stated as T2 with no other information on size (NET Ampulla)
993	Described as less than 3 cm or greater than 2 cm or between 2 & 3 cm
994	Described as less than 4 cm or greater than 3 cm or between 3 & 4 cm
995	Described as less than 5 cm or greater than 4 cm or between 4 & 5 cm Stated as T2 with no other information on size
999	Unknown; size not stated Size of tumor cannot be assessed Not documented in patient record

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CS Extension: Ampulla NET

- AJCC Staging
  - T1: Tumor size 1 cm or less
  - T2: Tumor size more than 1 cm
  - T3: Invades pancreas or retroperitoneum or non-peritonealized tissues
    - CS Extension = 520-600
  - T4: Invades serosa or other organs
    - CS Extension = 610-810

For CS Extension codes 100-430 only, T category is based on value of CS Tumor Size

CS Extension: NET Colon, NET Rectum

- AJCC Staging
  - T1: Tumor invades lamina propria or submucosa AND size 2 cm or less
  - T2: Invades muscularis propria OR size greater than 2 cm with invasion of lamina propria or submucosa
    - CS Extension = 200-210
  - T3: Invades muscularis propria into subserosa or into non-peritonealized pericolic or perirectal tissues
    - CS Extension = 400-458
  - T4: Invades peritoneum or other organs
    - CS Extension = 500-810

For CS Extension codes 100-190 & 300 only, T category is based on value of CS Tumor Size

CS Extension: NET Small Intestine

- AJCC Staging
  - T1: Invades lamina propria or submucosa AND size 1 cm or less
  - T2: Invades muscularis propria OR size greater than 1cm
    - CS Extension = 200-210
  - T3: Invades through muscularis propria into subserosal tissue without penetration of serosa (jejunal or ileal tumors) or invades pancreas or retroperitoneum (duodenal tumors) or into non-peritonealized tissues
    - CS Extension = 400-488
  - T4: Invades serosa or other organs
    - CS Extension = 500-810

For CS Extension codes 100-170 & 300 only, T category is based on value of CS Tumor Size

CS Extension: NET Stomach

- AJCC Staging
  - Tis:
    - In situ; CS Extension = 000
    - Confined to mucosa; CS Extension = 100
  - T1: Invades lamina propria or submucosa AND size 1 cm or less
  - T2: Invades muscularis propria OR size more than 1 cm
    - CS Extension = 200, 390
  - T3: Penetrates subserosa
    - CS Extension = 400-480
  - T4: Invades serosa or other organs or adjacent structures
    - CS Extension = 500-810

For CS Extension codes 110-170 & 300 only, T category is based on value of CS Tumor Size

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CS Extension: NET Ampulla

- Summary Stage 2000
  - In situ: Noninvasive; intraepithelial
    - CS Extension = 000
  - Localized (L): Confined to organ of origin
    - CS Extension = 100-300, 430
  - Regional by direct extension (RE): Extension to adjacent tissues and/or organs
    - CS Extension = 310, 420, 520, 600-700, 810
  - Distant extension (D): Extension to distant organs
    - CS Extension = 550, 750-800

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CS Extension: NET Colon, NET Rectum

- Summary Stage 2000
  - In situ: Noninvasive; intraepithelial
    - CS Extension = 000
  - Localized (L): Confined to organ of origin
    - CS Extension = 100-410
  - Regional by direct extension (RE): Extension to adjacent tissues and/or organs
    - CS Extension = 450-660, 810
  - Distant extension (D): Extension to distant organs
    - CS Extension = 7000-800

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CS Extension: NET Small Intestine

- Summary Stage 2000
  - In situ: Noninvasive; intraepithelial
    - CS Extension = 000
  - Localized (L): Confined to small intestine
    - CS Extension = 100-400
  - Regional by direct extension (RE): Extension to adjacent tissues and/or organs
    - CS Extension = 410-680, 810
  - Distant extension (D): Extension to distant organs
    - CS Extension = 700-800

CS Extension: NET Stomach

- Summary Stage 2000
  - In situ: Noninvasive; intraepithelial
    - CS Extension = 000
  - Localized (L): Confined to stomach
    - CS Extension = 100-400
  - Regional by direct extension (RE): Extension to adjacent tissues and/or organs
    - CS Extension = 450-610, 810
  - Distant extension (D): Extension to distant organs
    - CS Extension = 650-800

Pop Quiz

- Colonoscopy with rectal polypectomy: 1 cm polyp with carcinoid tumor.
- Abdominal perineal resection (APR): No residual tumor.
- What is the code for CS Tumor Size?
  - 010
  - 999
- What is the code for CS Extension?
  - 110: Invades lamina propria, including lamina propria in the stalk of a polyp
  - 120: Confined to and not through the muscularis mucosae, including muscularis mucosae in the stalk of a polyp
  - 160: Submucosa (superficial invasion), including submucosa in the stalk of a polyp
  - 300: Localized NOS; confined to rectum

CS Lymph Nodes: NET

- Code regional node involvement
  - NET has an affinity for spread through lymphatic system
  - Nodes considered regional are based on site of NET
  - AJCC N1
  - Summary Stage 2000 RN

CS Mets at DX: NET

- Record distant metastasis at time of diagnosis in distant lymph nodes and/or organs and tissues
  - Based on site of NET
  - AJCC M1
  - Summary Stage 2000 D
- Most common metastatic sites
  - Lymph nodes, liver, peritoneum, pancreas

SSFs for NET

- Clinical Assessment of Regional Lymph Nodes
  - SSF1 for NET Stomach
  - SSF2 for NET Colon & NET Rectum
- Serum Chromogranin A (CgA) Lab Value
  - SSF5 for NET Ampulla
  - SSF11 for NET Small Intestine & NET Stomach
  - SSF16 for NET Colon & NET Rectum
- Urinary 5-HIAA Lab Value
  - SSF6 for NET Ampulla
  - SSF12 for NET Small Intestine & NET Stomach
  - SSF17 for NET Colon & NET Rectum

Clinical Assessment of Regional Lymph Nodes

- Record clinically involved regional lymph nodes
  - Based on imaging or physical exam
    - Endoscopic procedures are excluded
  - Do NOT record pathologically determined information

Serum CgA Lab Value

- Record highest CgA lab value prior to treatment to the nearest ng/ml
- Chromogranin
  - Protein released from neuroendocrine cells
  - Elevated levels are a marker for neuroendocrine tumors

Question

- Can the serum CgA value be coded from a metastatic site such as a liver biopsy, or must the value be coded from the primary site?

Urinary 5-Hydroxyindoleacetic Acid (5-HIAA) Lab Value

- Record highest 5-HIAA lab value prior to treatment to nearest mg
- Carcinoids release excessive serotonin
  - Metabolized serotonin (5-HIAA) released in urine

Work-Up

- CT and MRI
- Radiolabeled somatostatin receptor scintigraphy
- Depending on the primary site:
  - Colonoscopy
  - Endoscopic ultrasound (EUS)
  - Esophogastroduodenoscopy (EGD)

Treatment

- Surgical resection for locoregional tumors
  - Tumor 2cm or less may have endoscopic resection
  - For larger tumor or tumors not accessible by endoscope a surgical resection of the bowel with regional lymphadenctomy
  - Prophylactic cholecystectomy may also be done if the patient is to receive adjuvant octreotide.
- For distant metastases
  - Liver: wedge resections, RFA, cryosurgery, chemoembolization
  - Palliation: combination chemotherapy or radiation
  - Somatostatin analogs for symptom control

Surveillance

- Chromagranin A
  - Elevated levels after initial treatment have been associated with recurrence
- 5-Hydroxyindoleacetic acid (5-HIAA)
  - Decreasing levels indicate a response to treatment
  - Increasing levels indicate treatment has not been successful

Case Scenarios

Coming Up…

- Collecting Cancer Data: Treatment Data
  - February 6, 2014
- Abstracting & Coding Boot Camp
  - March 6, 2014

And the Winners are…..





CE Certificate Quiz/Survey

- Phrase
- Link
  - http://www.surveygizmo.com/s3/1496306/GIST

Thank You!!!!

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