COLLECTING CANCER DATA: GIST/SARCOMA
2017-2018 NAACCR WEBINAR SERIES

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.
Fabulous Prizes

AGENDA
- Overview
- Epi Moment
- Quiz 1
- Staging
  - Bone
  - Soft Tissue
  - GIST
- Treatment
- Quiz 2
- Case Scenarios
SOFT TISSUE SARcoma

• Soft tissues include:
  • Muscles
  • Tendons
  • Fat
  • Blood vessels
  • Lymph vessels
  • Nerves
  • Tissues around joints
SOFT TISSUE SARCOMA

• Over 50 different types
  • Angiosarcoma – blood vessels or lymph vessels; following radiation
  • Undifferentiated pleomorphic sarcoma – most often in arms or legs (previously malignant fibrous histiocytoma MFH)
  • Spindle cell sarcoma – descriptive name based on the appearance of the cells
  • Liposarcoma – fat cells; most frequently seen

SOFT TISSUE SARCOMA

• Pleomorphic dermal sarcoma
  • Arise in sun-damaged skin
    • Head, Neck, Scalp
    • Negative: S100, Multiple CK, Desmin, CD34
    • Behavior likely more aggressive but limited follow up due to patient advanced age at presentation

• Atypical Fibroxanthoma (AFX)
  • Arise in sun-damaged skin
    • Ear, Nose, Forehead, Cheek
    • Negative: S100, multiple CK, Desmin, CD34
    • Behavior is almost invariably a benign behavior with only rare local recurrence
SOFT TISSUE SARCOMA

• Question

• What is the appropriate histology code for a final diagnosis or undifferentiated pleomorphic sarcoma and/or pleomorphic sarcoma, undifferentiated? Does the Other Sites MP/H Rule H17 apply in this case, which results in coding higher histology 8805/3? Or does the “undifferentiated” statement only refer to grade, which results in coding histology to 8802/3 (pleomorphic sarcoma)?

ANSWER

• SINQ 20160044

• Assign 8802/34 to pleomorphic cell sarcoma/undifferentiated pleomorphic sarcoma. Pleomorphic is more important than undifferentiated when choosing the histology code in this case. Undifferentiated can be captured in the grade code.
SARCOMA

• 2018 Histology – New Terms
  • 8571/3 Carcinoma with chondroid differentiation (C50._)
  • Carcinoma with osseous differentiation (C50._)
  • Metaplastic carcinoma with chondroid differentiation (C50._)
  • Metaplastic carcinoma with osseous differentiation (C50._)

• 2018 Histology – New Terms
  • 8801/3 Undifferentiated spindle cell sarcoma
  • 8802/3 Undifferentiated pleomorphic sarcoma
  • 8803/3 Undifferentiated round cell sarcoma
  • 8804/3 Undifferentiated epithelioid sarcoma
  • 8805/3 Undifferentiated uterine sarcoma
  • 8830/3 Undifferentiated high-grade pleomorphic sarcoma
OSTEOSARCOMA

- Most common malignant bone tumor
  - Arise from osteoblasts
- Typically occurs in long bones
- Mutation in TP53 are most common
- Distant metastasis occur in ~20%
  - Lung is most common site

OSTEOSARCOMA

- Periosteum
  - 2 layers
    - Fibrous connective tissue
    - Inner osteogenic
- Medullary cavity
  - Bone marrow

SOFT TISSUE SARCOMA GRADE

- FNCLCC grading system
  - Differentiation
  - Necrosis
  - Mitotic rate

- Histologic Grade:

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<thead>
<tr>
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<td>G1</td>
<td>FNCLCC grade score of 2 or 3</td>
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<tr>
<td>G2</td>
<td>FNCLCC grade score of 4 or 5</td>
</tr>
<tr>
<td>G3</td>
<td>FNCLCC grade score of 6, 7, or 8</td>
</tr>
</tbody>
</table>

GIST

ANATOMY
**WHAT IS GIST?**

- Rare type of soft tissue sarcoma
  - 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

**GIST**

- Interstitial cells of Cajal
  - “Pacemaker cells”
  - Sends signals to move food and liquid through system (peristalsis)

[Source Image](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% GIST contain no genetic mutation of KIT or PDGFRA

TUMOR LOCATION

<table>
<thead>
<tr>
<th>Location</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Stomach</td>
<td>60%</td>
</tr>
<tr>
<td>Small Intestine</td>
<td>30%</td>
</tr>
<tr>
<td>Rectum</td>
<td>3%</td>
</tr>
<tr>
<td>Colon</td>
<td>1-2%</td>
</tr>
<tr>
<td>Esophagus</td>
<td>&lt;1%</td>
</tr>
<tr>
<td>Omentum/Mesentery</td>
<td>Rare</td>
</tr>
</tbody>
</table>
GIST

• How do you determine if a GIST is malignant i.e. reportable?
  • GIST, NOS is a borderline tumor (/1)
    • If your state or facility requires collection of these GISTs, you should follow their requirements
  • Don’t determine reportability based on staging
    • AJCC staging forms are used on all GISTs

COLLECTING CANCER DATA: SARCOMA

Theme Song: The Skeleton Dance
**Epidemiology of Sarcomas**

- Large grouping of distinct cancers
  - 50+ distinct histologies; putative mesenchymal origin
  - Combined & studied as group
- Rare in adults (<2%); Top 5 for Pedi (21%)
- Majority soft tissue (87%); malignant bone (13%)
  - Soft: muscles, joints, fat, nerves, deep skin, blood vessels
  - Bone: commonly in cartilage
- Prognosis generally poor; esp soft
  - Delayed diagnosis: arise anywhere, lack of specific symptoms
  - No population based screening
  - Poor survival adults; better for pedi

**Pediatric Sarcomas (Soft)**

- Rhabdomyosarcoma – most common soft tissue for peds
  - Skeletal muscle
  - 50% occur <10; slightly more common in males
    - Often presents as painless mass; risk factor Li-Fraumeni syndrome
  - 5 year survival 70%; dependent upon location, stage, and histology—often lymph node involvement
    - Embryonal better prognosis than alveolar subtype; 20% present metastatic with 5 year survival 30-40% vs 80% for local
- Other
  - Fibrosarcoma – historically 2/3rds of sarcomas
    - now 12% due to better classification (proportion changed but not risk)
  - Liposarcoma – often large tumors; common among adults but <5% of ped sarcomas
  - Synovial sarcoma – 4th most common; 2x more common in males
    - origin mesenchymal not synovium; largely genetic
  - Malignant peripheral nerve sheath tumors – grouped into Brain CNS category for Epi
  - Alveolar Soft Part Sarcoma (ASPS) – rare, slow growing; generally mets at dx
  - Mesenchymoma – rare but highly aggressive
PEDIATRIC SARCOMAS (BONE)

- Osteosarcoma – most common
  - Generally on edges of “long” bones; 2nd most common location upper arm near shoulder
  - Surgery & Chemo
- Chondrosarcomas
- Ewing Family – 10-15% of bone sarcomas in pediatrics
  - Ewing sarcoma, extraosseous Ewing, PNET, Askin
  - Impacts teens; responsive to radiotherapy

Sarcomas (Bone), Rate per 1,000,000, CiNA 2014

RISK FACTORS

- Varied causes; distinct disease; limited studies & understanding
  - Environmental, genetic (synergistic)
- Genetic
  - Age: Soft: Rate high <5 for soft, lower but steady increase 6-49; 50+ high; Bone: Rate stable across ages; Rate high in YA (osteosarcoma, Ewing)
  - Race: Ewing (9x more common white vs black); but soft tissue higher for blacks
  - Hx of hernia - Ewing sarcoma (children)
  - Growth “spurts” – osteosarcoma (children)
  - Genetic syndromes (Li-Fraumeni; neurofibromatosis/von Recklinghausen dx, retinoblastoma)
    - Non currently described for Ewing but likely genetic
- Environmental
  - HIV+ for KS (but HHV8 is causal, HIV & EBV)
  - Radiation exposure - Bone cancer; Tx and atomic (Japanese)
  - Occupational exposures –herbicides (soft tissue)
THE GIST ON G.I.S.T. h/t Brad Wohler, FCDS

- 2001 ICD-O-3 specific code
- Rare, digestive tract soft tissue sarcoma
- Adult cancer; 50+
- Used to believe origin was nerve or muscle cells
  - Now understood to arise from interstitial cells of Cajal (ICC) or precursor—the “pacemakers” of digestion; occur from esoph to anus but over ½ in stomach
- Moving away from “benign” designation = increase rates but not risk

![Graph showing GIST over time (CINA 1995-2014)]

APC 7.8%*

QUESTIONS?

QUIZ 1
SUMMARY STAGE 2000-MUSCULOSKELETAL SYSTEM

- Bones, joints, and articular cartilage
  - C40.0-C40.3, C40.8-C40.9, C41.0-C41.4, C41.8-C41.9

- Peripheral nerves and autonomic nervous system; connective, subcutaneous, and other soft tissues
  - C47.0-C47.6, C47.8-C47.9, C49.0-C49.6, C49.8-C49.9

- Retroperitoneum and peritoneum
  - C48.0-C48.2, C48.8


SUMMARY STAGE 2000-GIST

- Use location of the tumor to determine to summary stage chapter.
  - GIST of the stomach
    - Use summary stage chapter Stomach
  - GIST of the Ileum
    - Use Summary Stage chapter Small Intestine
SUMMARY STAGE 2018
• GIST will have it’s own chapter
• Sarcoma chapters have not yet been defined

AJCC STAGE
REVIEW OF 7TH & 8TH EDITION
## BONE

### 7TH EDITION CHAPTERS

<table>
<thead>
<tr>
<th>Chapter Title</th>
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<tr>
<td>Gastrointestinal Stromal Tumor</td>
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<td>Soft Tissue Sarcoma</td>
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### 8th Edition Chapters

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<td>Trunk and Extremities</td>
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<tr>
<td>Abdomen and Thoracic Visceral Organs</td>
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<td>GIST</td>
<td>43</td>
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<td>Retroperitoneum</td>
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### Bone-Rules for Classification

- **Clinical**
  - Imaging (MRI) followed by biopsy
- **Pathologic**
  - Resection of the primary tumor
  - "cN may be used to assign the pathologic stage"
7th Edition - Size of the Primary Tumor

- Is the tumor 8cm or less?
- Are there discontinuous tumors present in the bone?

See page 286

https://basicmedicalkey.com/osteosarcoma-4/

8th Edition - Location, Location, Location

- Where is the tumor?
  - Appendicular skeleton, trunk, skull, facial bones
  - Spine
  - Pelvis

See page 476

http://www.connectedkansaskids.com/diagnoses/osteosarcoma.html
APPENDICULAR SKELETON, TRUNK, SKULL, FACIAL BONES

- Is the tumor 8cm or less?
- Are there discontinuous tumors present in the bone?

SPINE

- How many vertebral segments are involved?
- Is there spinal canal involvement?
- Is there involvement of the great vessels?

See page 476

https://en.wikipedia.org/wiki/Vertebral_column
PELVIS

- How many segments of the pelvis are involved?
  - See fig 38.2 on page 474
- Is the tumor 8cm or less?
- Does the tumor cross the sacroiliac joint?
- Does tumor encase the external iliac vessel or cause tumor thrombus?
  See page 476

https://en.wikipedia.org/wiki/Pelvis

METASTASIS

- Lymph node metastasis is rare
  - cN values may be used in the pN data item
- Distant metastasis
  - Lung
    - Solitary tumor
    - Multiple tumors
  - Secondary bone
  - Other
GRADE

- Cases diagnosed ≤ 2017 use the instructions for Coding Grade for 2014+
  - Two grade
    - Low-2
    - High-4
  - Four grade

GRADE

- Cases diagnosed ≥ 2018 code Clinical, Pathologic, Post-Therapy Grade

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<th>Grade Description</th>
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<tr>
<td>2</td>
<td>G2: Moderately differentiated, high grade</td>
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<tr>
<td>3</td>
<td>G3: Poorly differentiated, high grade</td>
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<tr>
<td>H</td>
<td>Stated as “high grade” only</td>
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<tr>
<td>9</td>
<td>Grade cannot be assessed (GX); Unknown; Not applicable</td>
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STAGE GROUPING

- Appendicular Skeleton, Trunk, Skull, and Facial Bones
  - Grade is part of stage grouping
- Spine and Pelvis do not have stage group tables.
  - Stage group is 88

SSF 3/SSDI-PERCENT NECROSIS POST NEOADJUVANT

- Record percentage value of the tumor necrosis post neoadjuvant chemotherapy as recorded in the pathology report from resection of the primary tumor.

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<td>(Percentage of tumor necrosis to nearest tenth of a percent)</td>
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<td>No neoadjuvant therapy</td>
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<tr>
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<td>No surgical resection of primary site is performed</td>
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**POP QUIZ 1**

- A 73 year old male presented with a mass on his left femur.
  - An MRI showed a single 9cm mass confined to the femur.
  - A biopsy confirmed high grade chondrosarcoma.
  - The patient received neoadjuvant chemotherapy followed by surgical resection of the tumor.
  - Imaging showed post-therapy tumor size of 7cm.
  - The pathology report from the resected specimen showed a 7cm chondrosarcoma grade I. The extent of tumor necrosis was 95%.

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<td>Percent Necrosis Post Neo adjunct/ SSF3</td>
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<td>95.0</td>
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</table>
SOFT TISSUE

7TH EDITION CHAPTER 28-SOFT TISSUE SARCOMA

- Applies to all soft tissue sarcomas except:
  - Kaposi Sarcoma
  - GIST (Chapter 16 Gastrointestinal Stromal Tumor)
  - Fibromatosis
  - Infantile fibrosarcoma

- Sarcoma’s arising from the following sites are not “optimally” staged by this system
  - Sarcomas arising in dura mater
  - Sarcomas arising in parenchymal organs
  - Sarcomas arising in visceral hollow organs
**SOFT TISSUE SARCOMA-RULES FOR CLASSIFICATION**

- **Clinical Staging**
  - Based on imaging and clinical evaluation prior to any treatment.
  - Tumor size can be measured clinically or radiographically (MRI or CT).
  - Evaluation for metastasis should be based on imaging. Most likely spot for distant metastasis is lungs.

- **Pathologic Staging**
  - Based on resection of the primary tumor and clinical/radiologic evaluation for regional and distant metastasis.
  - Tumor size can be based on imaging if an accurate tumor size cannot be obtained from the resected specimen.
  - If no lymph nodes removed, cN value may be used in the pN data item.
7TH EDITION CHAPTER 28-SOFT TISSUE SARCOMA

- Is the primary tumor ≤ 5cm?
- Is the tumor superficial or deep?
- No T3 or T4

SUPERFICIAL VS DEEP

- Superficial
  - Located entirely in the subcutaneous tissues without any degree of extension through muscular fascia or into underlying muscle
- Deep
  - Tumor arising within subcutaneous tissue with invasion into or through the superficial fascia
  - Tumor entirely beneath the superficial fascia
  - Tumor arising beneath the deep fascia with invasion into or through the superficial fascia

8TH EDITION SOFT TISSUE SARCOMA

- Chapter 39-Introduction
- Chapter 40-Head and Neck
- Chapter 41-Trunk and extremities
- Chapter 42-Abdomen and Thoracic Visceral Organs
- Chapter 44-Retroperitoneum
- Chapter 45-Unusual Histologies and Sites

SOFT TISSUE SARCOMA-RULES FOR CLASSIFICATION

- Clinical Staging
  - Based on imaging and clinical evaluation prior to any treatment.
    - Tumor size can be measured clinically or radiographically (MRI or CT)
    - Evaluation for metastasis should be based on imaging.
SOFT TISSUE SARCOMA-RULES FOR CLASSIFICATION

- Pathologic Staging
  - Based on resection of the primary tumor and clinical/radiologic evaluation for regional and distant metastasis.
  - Tumor size can be based on imaging if an accurate tumor size cannot be obtained from the resected specimen
  - If no lymph nodes removed, cN value may be used in the pN data item

SOFT TISSUE SARCOMA

Head and Neck
- C47.0 Peripheral nerves of head and neck
- C49.0 Connective, subcutaneous, and other soft tissues of head and neck
- C32.9 Larynx
- C02.3 Anterior 2/3 of tongue
- ...

Pg 500

Trunk and Extremities
- C47.1 Peripheral nerves of the upper limb and shoulder
- C49.1 Connective, subcutaneous, and other soft tissues of the upper limb and shoulder
- C50.9 Breast

Pg 500
SOFT TISSUE SARCOMA

Head and Neck
- Is the tumor ≤ 2cm?
- Is the tumor ≤ 4cm?
- Is the tumor invading adjacent structures?
- Stage group 88

Trunk and Extremities
- Is the tumor ≤ 5cm?
- Is the tumor ≤ 10cm?
- Is the tumor ≤ 15cm?
- Is the tumor more than 15cm?

Abdomen and Thoracic Visceral Organs
- C47.0 Peripheral nerves thorax
- C49.0 Connective, subcutaneous, and other soft tissues of thorax
- C15-C26 Digestive organs
- C34-C37 Intrathoracic organs

Retroperitoneum
- C48.0-C48.8 Retroperitoneum

Superficial vs Deep not a factor
SOFT TISSUE SARCOMA

Abdomen and Thoracic Visceral Organs

• Is the primary tumor confined to the organ of origin?
• Does the tumor invade into or through the serosa or visceral peritoneum?
• Does tumor invade another organ?
• Are there multiple tumors?
• Stage group 88

Retroperitoneum

• T, N, M similar to Trunk and extremities, but the stage group slightly different

GRADE-CLIN, PATH, POST-THERAPY

• Codes 1-3 take priority over A-D.
• Codes A-D are equivalent to a GX when assigning AJCC Stage Group.

<table>
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<tr>
<th>Code</th>
<th>Grade Description</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>G1: Sum of differentiation score, mitotic count score and necrosis score equals 2 or 3</td>
</tr>
<tr>
<td>2</td>
<td>G2: Sum of differentiation score, mitotic count score and necrosis score of 4 or 5</td>
</tr>
<tr>
<td>3</td>
<td>G3: Sum of differentiation score, mitotic count score and necrosis score of 6, 7, or 8</td>
</tr>
<tr>
<td>A</td>
<td>Well differentiated</td>
</tr>
<tr>
<td>B</td>
<td>Moderately differentiated</td>
</tr>
<tr>
<td>C</td>
<td>Poorly differentiated</td>
</tr>
<tr>
<td>D</td>
<td>Undifferentiated, anaplastic</td>
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</table>
CS SSF 3/SSDI-BONE INVASION

- Direct tumor extension from the primary sarcoma into adjacent bone.
- This field does not include distant or discontinuous metastases to the skeletal system.
- Information in this field is based on radiology and other imaging techniques.

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<td>Not documented in medical record</td>
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<td>Bone invasion not assessed or unknown if assessed</td>
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POP QUIZ 2

- A patient presents with a 6cm mass in her left breast.
  - MRI: 5.7cm mass in the left breast. No additional masses identified.
  - A core biopsy confirmed fibrosarcoma. Grade could not be assessed due to inadequate specimen.
  - Modified radical mastectomy:
    - 5.7cm fibrosarcoma confined to the breast.
    - Mitotic Count Score: 11 per 10 HPF
    - Tumor Necrosis: 75% tumor necrosis
    - Differentiation Score: 2
    - FNCLCC grade 3
    - No bone involvement

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<td>Path T</td>
<td>pT2</td>
<td>pT2</td>
</tr>
<tr>
<td>Path N</td>
<td>Blank</td>
<td>cN0</td>
</tr>
<tr>
<td>Path M</td>
<td>cM0</td>
<td>cM0</td>
</tr>
<tr>
<td>Path Grade</td>
<td></td>
<td>3</td>
</tr>
<tr>
<td>Stage</td>
<td>3</td>
<td>3A</td>
</tr>
<tr>
<td>SSF 3 Bone Invasion</td>
<td>000</td>
<td>0</td>
</tr>
</tbody>
</table>
GIST

7th Edition Chapter 28-Soft Tissue Sarcoma
8th Edition Chapter 43-Gastrointestinal Stromal Tumor

• No changes in 8th edition
• Follow Rules for Classification for Soft Tissue Tumors
7TH EDITION CHAPTER 28-SOFT TISSUE SARCOMA
8TH EDITION CHAPTER 43-GASTROINTESTINAL STROMAL TUMOR

• Primary tumor is assessed based on size
  • Is the tumor ≤ 2cm?
  • Is the tumor ≤ 5cm?
  • Is the tumor ≤ 10cm?
  • Is the tumor more than 10cm?
• Regional node metastasis is extremely rare
  • cN values may be used in the pN data item
• Distant metastasis is rare, but may occur arise in intraabdominal soft tissue, liver (parenchyma), bone, soft tissues, and skin

GRADE-CLIN, PATH, P-T

• Codes L and H take priority over A-D.
• Codes A-D are equivalent to a GX when assigning AJCC Stage Group.
• Record the mitotic rate as Low or High as indicated on the pathology report or CAP protocol. Assume the denominator is 5 square mm if not specified.
  • Low: 5 or fewer mitoses per 5 mm² (L)
  • High: Over 5 mitoses per 5 mm² (H)
• SSF for pre-2018 cases

<table>
<thead>
<tr>
<th>Code</th>
<th>Grade Description</th>
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<tbody>
<tr>
<td>L</td>
<td>Low: 5 or fewer mitoses per 5 mm²</td>
</tr>
<tr>
<td>H</td>
<td>High: Over 5 mitoses per 5 mm²</td>
</tr>
<tr>
<td>A</td>
<td>Well differentiated</td>
</tr>
<tr>
<td>B</td>
<td>Moderately differentiated</td>
</tr>
<tr>
<td>C</td>
<td>Poorly differentiated</td>
</tr>
<tr>
<td>D</td>
<td>Undifferentiated, anaplastic</td>
</tr>
<tr>
<td>9</td>
<td>Grade cannot be assessed; Unknown</td>
</tr>
<tr>
<td></td>
<td>Not applicable</td>
</tr>
</tbody>
</table>
GIST STAGE GROUPING

- Mitotic rate strongly influences stage group
- Stage grouping is different for tumors arising in the stomach and tumors arising in the small intestine
  - Primary omental GIST - Gastric Omental table (8th edition)
  - Tumors arising in sites other than stomach/omentum or small intestine should be grouped based on Small Intestine table

SSF/SSDI-KIT GENE IMMUNOHISTOCHEMISTRY

- KIT immunohistochemistry is a special immunofluorescent stain that turns mutated cells brown and confirms a diagnosis of GIST.
- The presence of the KIT gene also indicates that the patient may respond to Gleevec or Sutent.

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>KIT negative/normal; within normal limits</td>
</tr>
<tr>
<td>1</td>
<td>KIT positive</td>
</tr>
<tr>
<td>7</td>
<td>Test ordered, results not in chart</td>
</tr>
<tr>
<td>8</td>
<td>Not applicable: Information not collected for this case</td>
</tr>
<tr>
<td>9</td>
<td>Not documented in medical record; Cannot be determined by pathologist; KIT not assessed or unknown if assessed</td>
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</tbody>
</table>
SSDI-SHEMA DISCRIMINATOR

- Since both omental and peritoneal gastrointestinal stromal tumors (GIST) are coded with the same ICD-O-3 topography code (C48.1), this data item must be used to identify the appropriate AJCC stage table.

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
<th>Stage Table</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Mesentery; Mesoappendix; Mesocolon; Pelvic peritoneum; Rectouterine pouch; Cul de sac; Pouch of Douglas; Other specified peritoneal site</td>
<td>Small Intestinal, Esophageal, Colorectal, Mesenteric and Peritoneal GIST</td>
</tr>
<tr>
<td>2</td>
<td>Omentum</td>
<td>Gastric and Omental GIST</td>
</tr>
<tr>
<td>9</td>
<td>Unknown or no information</td>
<td>Small Intestinal, Esophageal, Colorectal, Mesenteric and Peritoneal GIST</td>
</tr>
<tr>
<td></td>
<td>Not documented in patient record</td>
<td></td>
</tr>
</tbody>
</table>

**POP QUIZ 3**

- A patient presents with severe flank pain.
  - CT showed a 12.1 x 5.9cm hypodense mass mesenteric mass suspicious for a solid mass vs large hematoma. No associated lymphadenopathy. Surgery was recommended.
  - Pathology from Surgery
    - 8.2cm, cystic, hemorrhagic malignant spindle cell lesion with coagulative necrosis, most consistent with extra-gastrointestinal stromal tumor
    - CKIT, DOG1 and Vimentin were positive.
    - Mitosis rate 8/50hpf.

Primary Site: C48.1 Mesentery
Histology: 8936/3 GIST

<table>
<thead>
<tr>
<th>Data Item</th>
<th>7th ed</th>
<th>8th ed</th>
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<tbody>
<tr>
<td>Schema D 1</td>
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<td>1</td>
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<tr>
<td>Clinical T</td>
<td></td>
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<tr>
<td>Clinical N</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Clinical M</td>
<td></td>
<td></td>
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<tr>
<td>Grade/ Clinical Grade</td>
<td>9</td>
<td>9</td>
</tr>
<tr>
<td>Stage</td>
<td>99</td>
<td>99</td>
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<td>Path T</td>
<td>pT3</td>
<td>pT3</td>
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<td>Path N</td>
<td>Blank or cN0</td>
<td>cN0</td>
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<td>cM0</td>
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<td>SSF 3 Mitotic Count</td>
<td>080</td>
<td>H</td>
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<td>Path Grade</td>
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<td></td>
</tr>
<tr>
<td>Stage</td>
<td>3B</td>
<td>3B</td>
</tr>
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</table>
TREATMENT
SARCOMA/GIST

TREATMENT - OSTEOSARCOMA

- Intramedullary
  - Wide excision
- Periosteal
  - Consider neoadjuvant chemotherapy
  - Wide excision
- High grade Intramedullary
  - Neoadjuvant chemotherapy
  - If resectable – Wide Excision
    - Adjuvant chemotherapy +/- Radiation
    - If unresectable
      - Radiation
      - Chemotherapy
### TREATMENT OSTEOSARCOMA

- **Metastatic disease at diagnosis**
  - **Resectable:**
    - Wide excision local tumor
    - Excision of metastasis
    - Chemotherapy
  - **Unresectable**
    - Chemotherapy
    - Radiation

### SURGERY

- **C40.0-C41.9; C47.0-C47.9; C49.0-C49.9**
  - Local excision – 25
    - Excisional biopsy – tumor itself
  - Partial resection – 26
    - Wide excision – more healthy tissue removed around tumor
  - Radical excision or resection with limb salvage – 30
    - Significant amount of healthy tissue removed
SURGERY

C40.0-C41.9; C47.0-C47.9; C49.0-C49.9

• Amputation of limb – 40
  • Partial amputation of limb – 41
    • Portion of the arm or leg
  • Total amputation of limb – 42
    • Leg and the hip
    • Arm and the shoulder

POP QUIZ

• 48yo with large mass on right 3rd toe
• Incisional biopsy right 3rd toe reveals sarcoma
• Decision to amputate the entire right 3rd toe
• Pathology R 3rd toe, amputation: pleomorphic sarcoma, margins negative within 1cm.
POP QUIZ

• Diagnostic/Staging Procedure:
  • 02

• Surgery Primary Site:
  • 30

• Scope Regional LN Surgery:
  • 0

RADIATION

• Primary Tumor Volume /Phase I Radiation Primary Treatment Volume

<table>
<thead>
<tr>
<th>FORDS-2017</th>
<th>STORE-2018</th>
<th>STORE Description</th>
</tr>
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<tbody>
<tr>
<td>25</td>
<td>80</td>
<td>Skull</td>
</tr>
<tr>
<td>24</td>
<td>81</td>
<td>Spine/Vertebral Bodies</td>
</tr>
<tr>
<td>37</td>
<td>82</td>
<td>Shoulder</td>
</tr>
<tr>
<td>26</td>
<td>83</td>
<td>Ribs</td>
</tr>
<tr>
<td>27</td>
<td>84</td>
<td>Hip</td>
</tr>
<tr>
<td>28</td>
<td>85</td>
<td>Pelvic Bones</td>
</tr>
<tr>
<td>38</td>
<td>88</td>
<td>Extremity bone, NOS</td>
</tr>
<tr>
<td>30</td>
<td>90</td>
<td>Skin</td>
</tr>
<tr>
<td>31</td>
<td>91</td>
<td>Soft Tissue</td>
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## RADIATION

### Regional Modality / Phase I Radiation Treatment Modality

<table>
<thead>
<tr>
<th>FORDS-2017</th>
<th>STORE-2018</th>
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<td>External beam, photons, low energy</td>
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<td>22-27, 31, 41, 42, 43</td>
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</tr>
<tr>
<td>40</td>
<td>03</td>
<td>External beam, protons</td>
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<tr>
<td>28</td>
<td>04</td>
<td>External beam, electrons</td>
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<tr>
<td>30</td>
<td>05</td>
<td>External beam, neutrons</td>
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<tr>
<td>20</td>
<td>06</td>
<td>External beam, carbon ions</td>
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<tr>
<td>29</td>
<td>09</td>
<td>External beam, NOS</td>
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</table>

### RADIATION

### Regional Modality / Phase I External Beam Radiation Planning Technique

<table>
<thead>
<tr>
<th>FORDS Modality-2017</th>
<th>STORE-2018</th>
<th>STORE Description</th>
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<td>20, 22-27, 29, 30, 40</td>
<td>01</td>
<td>External beam, NOS</td>
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<tr>
<td>21</td>
<td>02</td>
<td>Low energy x-ray/photon</td>
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<tr>
<td>28</td>
<td>03</td>
<td>2-D therapy</td>
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<tr>
<td>32</td>
<td>04</td>
<td>Conformal or 3-D conformal therapy</td>
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<tr>
<td>31</td>
<td>05</td>
<td>Intensity Modulated Therapy</td>
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<tr>
<td>41, 42</td>
<td>06</td>
<td>Stereotactic radiotherapy or radiosurgery, NOS</td>
</tr>
<tr>
<td></td>
<td>07</td>
<td>Stereotactic radiotherapy or radiosurgery, robotic</td>
</tr>
<tr>
<td>43</td>
<td>08</td>
<td>Stereotactic radiotherapy or radiosurgery, Gamma Knife</td>
</tr>
<tr>
<td></td>
<td>09</td>
<td>CT guided online adaptive therapy</td>
</tr>
<tr>
<td></td>
<td>10</td>
<td>MR guided online adaptive therapy</td>
</tr>
</tbody>
</table>
POP QUIZ 4

- A 48 year old patient with a recent right 3rd toe amputation for sarcoma presents to discuss radiation treatment.
  - Plan: 6MV photons, IMRT to right foot, surgical bed
  - The patient completed radiation on 8/1/2017, right foot surgical bed, 6MV photons, 1800cGy in 6fx

POP QUIZ

<table>
<thead>
<tr>
<th>Radiation Data Items</th>
<th>2017</th>
<th>2018</th>
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</thead>
<tbody>
<tr>
<td>Radiation Primary Tumor Volume</td>
<td>38</td>
<td>88</td>
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<tr>
<td>Radiation Treatment Modality</td>
<td>31</td>
<td>02</td>
</tr>
<tr>
<td>External Beam Radiation Planning Technique</td>
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<td>05</td>
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</tbody>
</table>
**SYSTEMIC - CHEMOTHERAPY**

- Isolated Limb Infusion
- Ifosfamide and Doxorubicin
  - Mesna – used with Ifosfamide
- MAID – Mesna, Adriamycin, Ifosfamide, Dacarbazine
- Pazopanib (Votrient)
  - Blocks Tyrosine Kinases

**SYSTEMIC – IMMUNOTHERAPY**

- Olaratumab (Lartruvo)
  - FDA approved 10/19/16
  - Radiation or surgery not curative
  - In combination with Doxorubicin
**TREATMENT – GIST**

**Gastric:**
- Surgery
  - Small tumors removed laproscopically

**Other Sites:**
- Surgery
- Targeted Therapy
  - Imatinib (Gleevec) - Chemotherapy

---

**TREATMENT – GASTRIC SURGERY**

- Excisional biopsy (NOS) – 27
  - WITH electrocautery – 22
  - WITH cryosurgery – 23
  - WITH laser ablation – 24
  - Laser excision – 25
- Gastrectomy – 30
  - Antrectomy, lower – 31
  - Lower gastrectomy – 32
  - Upper gastrectomy – 33
TREATMENT – OTHER SITES SURGERY

C15.0-C15.9
- Excisional Biopsy (NOS) – 27
  - WITH electrocautery – 22
  - WITH Cryosurgery – 23
  - WITH Laser ablation – 24
- Partial Esophagectomy - 30

C18.0-C18.9; C19.9; C20.9
- Excisional biopsy – 27
  - WITH electrocautery – 22
  - WITH cryosurgery – 23
  - WITH laser ablation – 24
- Segmental resection - 30

POP QUIZ 5
- A 52 year old male presents with chronic anemia. He has never had a colonoscopy. Plan: screening colonoscopy
- Colonoscopy – Rectal mass, biopsy performed; no other abnormalities noted
- Rectum mass, biopsy: malignant GIST
- Patient returns for excision of rectal GIST
- Rectum, excision mass: malignant GIST, 3cm, mitoses: 6/50HPF, KIT+
POP QUIZ 5 CONT.

• Rectal GIST with high mitotic rate, 3cm, and KIT + is here to discuss adjuvant treatment options. Plan is to begin Gleevec

• Patient here for 3mo follow up for rectal GIST, started Gleevec and is doing well with minimal side effects

---

### Treatment data items

<table>
<thead>
<tr>
<th>Treatment data items</th>
<th>Codes</th>
</tr>
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<tbody>
<tr>
<td>Surgery Primary Site</td>
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<td>Scope Regional LN Surgery</td>
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</tr>
<tr>
<td>Chemotherapy</td>
<td>02</td>
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<tr>
<td>Immunotherapy</td>
<td>00</td>
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<tr>
<td>Systemic/Surgery Sequence</td>
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</tbody>
</table>
QUESTIONS?

QUIZ 2
CASE SCENARIOS

COMING UP....

• Collecting Cancer Data: Stomach and Esophagus
  • 02/01/2018

• Boot Camp!
  • 3/1/18
**Fabulous Prizes Winners**

- Image of a pie with a slice taken out, showing the inside filling.
- Image of a coffee cup with a saucer and a sugar bowl.
- Image of sausage rolls or pastries on a plate.
- Image of a snowman on a beach.
- Image of Christmas cookies with candy canes and ornaments.

---

**CE CERTIFICATE QUIZ/SURVEY**

- Phrase
  - Necrosis
- Link
JIM HOFFERKAMP jhofferkamp@naaccr.org
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RECINDA SHERMAN rsherman@naaccr.org
ANGELA MARTIN amartin@naaccr.org