

NAACCR

## Unusual Sites and Histologies

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2015-2016 NAACCR Webinar Series

October 1, 2015

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ooooo Q&A

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- 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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ooooo Fabulous prizes

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 Guest speaker

- Donna Hansen, CTR
  - Auditor/Trainer
  - UC Davis Institute of Population Health
  - California Cancer Registry

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 Agenda

- Mesothelioma
- Sinuses
  - Quiz 1
- Merkel Cell Carcinoma
- Melanoma of the Uvea and Conjunctiva
- Gestational Trophoblastic Tumors
  - Quiz 2
- Case Scenarios

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 Mesothelioma

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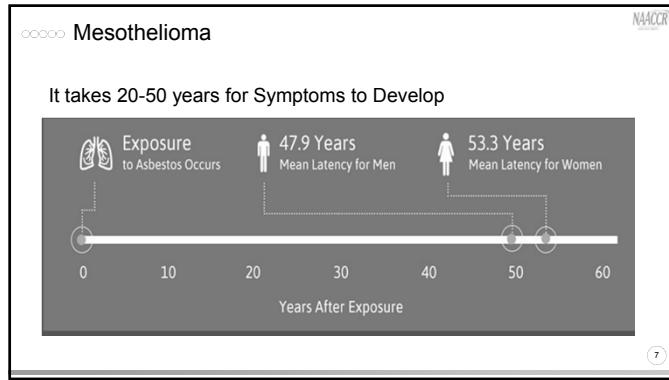
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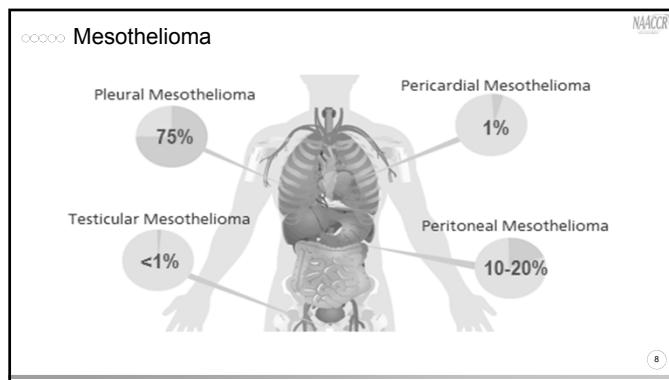
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- NAACCR
- ooooo Mesothelioma
- Mesothelioma has 3 histopathologic subtypes:
    - Epithelioid (ICD-0-3 code 9052)
    - Biphasic (sometimes called mixed) (ICD-0-3 code 9053)
      - Contains at least 10% of both epithelioid & sarcomatoid components
    - Sarcomatoid (ICD-0-3 code 9051)
      - Desmoplastic is a sub group of sarcomatoid
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ooooo Pleural Mesothelioma

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- Primary Site: Pleura, NOS C38.4
- Pleural mesothelium covers the external surface of lungs and the inside of the chest wall
- The pleura is composed of:
  - Parietal pleural - Outside layer
  - Pleural space - Between visceral & parietal pleura
  - Visceral Pleura - Covers the lungs

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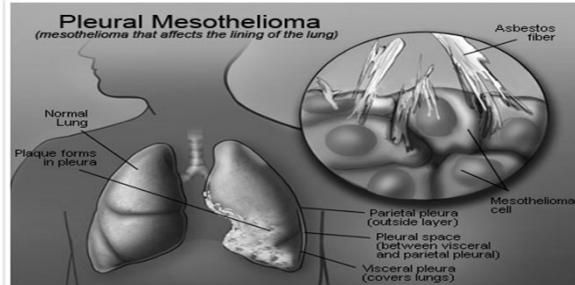
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ooooo Pleural Mesothelioma

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ooooo Pleural Mesothelioma

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- Diagnostic Workup may include:
  - CT Chest
  - Thoracentesis
  - Pleural biopsy, and
  - Possible SMRP (Soluble Mesothelin-Related Peptide) blood test
- Staging Workup may include:
  - Chest MRI or PET
  - Possible VATS and/or laparoscopy

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ooooo Pleural Mesothelioma

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- Prognostic Factors – important to document
  - Pleural effusion
  - Histologic Subtype
  - History or exposure to asbestos
  - Presence or absence of chest pain
- Distant Metastases - Most frequent sites
  - Lung
  - Contralateral pleura
  - Peritoneum

CS SSFs 1-4

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Pleural Mesothelioma

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How is Mesothelioma Treated?

Multi-Modality Therapy

Surgery

Radiation

Chemotherapy

Clinical Trials  
Immunotherapy

Treatment based on

- Histology
- Stage
- Patient Status

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ooooo Treatment

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- Surgery Procedures
  - Debulking
  - Pleurectomy/Decortication
  - Extrapleural Pneumonectomy (EPP)
- Palliative Procedure
  - "Talc" Pleurodesis
  - Chemotherapy agents
  - Pemetrexed, Cisplatin, Avastin, Carboplatin, Gemcitabine, Vinorelbine, Doxorubicin

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 Treatment

- Radiation Therapy
  - Before or after surgery
  - Seeding prevention
  - Palliative pain relief
  - Radiation types include 3D-CRT, IMRT, Proton beam
- Clinical Trials
  - Immunotherapy
  - Vaccine being researched

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 Primary Tumor

- Look for involvement of:
    - Parietal Pleura
    - Visceral Pleura
    - Lung parenchyma
    - Diaphragmatic muscle
    - Endotracheal fascia
    - Mediastinal fat
    - Soft tissues of chest wall
    - Non-transmural involvement of
      - pericardium
    - Multifocal masses in chest wall
- Extension to:
- Peritoneum
  - Contralateral pleura
  - Mediastinal organs
  - Spine
  - Internal surface of pericardium
  - Tumor involving myocardium

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 Regional Lymph Nodes

- Regional LNs of the Pleura
  - Intrathoracic
  - Scalene
  - SuprACLavicular
  - Internal Mammary
  - Peridiaphragmatic
- Pleural mesothelioma often metastasizes to LNs *not* usually involved in lung cancers
  - Most common - Internal mammary & Peridiaphragmatic nodes (N2)

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 Metastasis

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- Most Common
  - Contralateral pleura
  - Lungs
  - Peritoneum
- Advanced disease
  - Retroperitoneal LNs
  - Brain
  - Spine

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 Practice Case 1

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- 78-year-old male with history of asbestos exposure working as automotive brake mechanic for 30 years.
- Presents to ER with severe shortness of breath and chest pain.
- Workup revealed right lung pleural effusion with pleural nodules and thickening along the right upper lobe c/w neoplastic process.  
Enlarged right paratracheal LN.

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 Practice Case 1(cont.)

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- Pt underwent right thoracoscopy w/MDLNND revealing thickened pleura, extensive tumor plaques w/studding encasing superior right lobe involving parietal and visceral pleura. No lung invasion.
- Bx of tumor plaques positive for Epithelioid Mesothelioma. Bx of R2 paratracheal LNs positive for metastatic mesothelioma. Pleural fluid positive for malignant cells. Talc pleurodesis performed.
- Metastatic workup negative for other sites of disease. Patient is not a surgical candidate due to comorbidities. Radiation to LNs + Pemetrexed.

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Practice Case 1(cont)

• How do we stage this case?

Data Items as Coded in Current NAACCR Layout				
	T	N	M	Stage Group
Clin				
Path				
<b>Summary Stage</b>				

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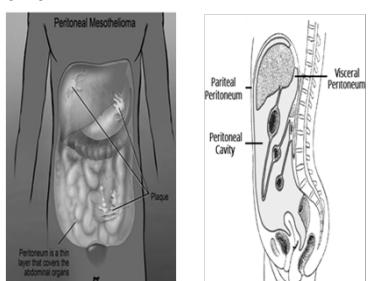
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Peritoneal Mesothelioma

- Primary Site
- C48.2



Peritoneum is a thin layer that covers the abdominal organs.

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Peritoneal Mesothelioma

• Treatment

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graph TD
    Surgery[Surgery] --> Treatment[Treatment]
    Treatment --> Chemotherapy[Chemotherapy]
    Treatment --> Radiation[Radiation]
    Treatment --> PalliativeSurgery["Palliative Surgery  
• Cytoreductive Surgery  
• Peritonectomy  
• w/o intraperitoneal chemo"]
    
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• Systemic Chemotherapy  
• Perioperative intraperitoneal Chemo

Based on Stage and Patient performance status

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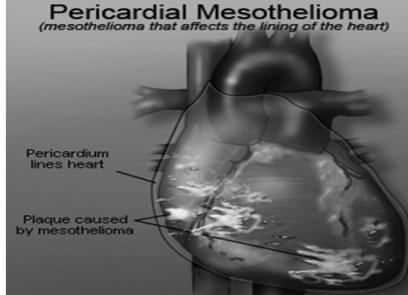
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ooooo Pericardial mesothelioma

**Pericardial Mesothelioma**  
*(mesothelioma that affects the lining of the heart)*



The diagram shows a cross-section of the human heart. The outer layer is labeled 'Pericardium lines heart'. Inside the heart, there are several white, irregular plaques labeled 'Plaque caused by mesothelioma'.

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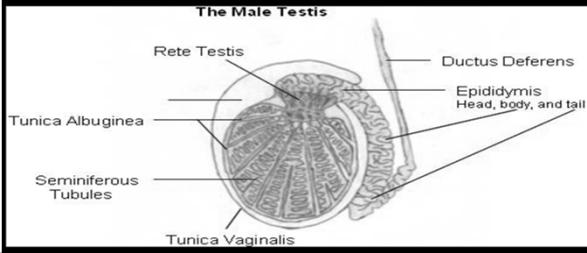
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ooooo Testicular Mesothelioma

- Originates in the Tunica Vaginalis membrane

**The Male Testis**



The diagram shows a cross-section of the male testis. Labels include: Rete Testis, Ductus Deferens, Epididymis Head, body, and tail, Tunica Albuginea, Seminiferous Tubules, and Tunica Vaginalis.

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ooooo Non-Pleural Mesothelioma

- AJCC TNM includes a formal staging schema for Pleura only
- There is no formal TNM schema for Mesothelioma of
  - Peritoneum
  - Pericardium
  - Testis

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**Mesothelioma - Summary Stage**

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- Summary Stage 2000 can be used to assign a stage code for all sites where mesothelioma occurs.
- Select Summary Stage and CS schema per disease location:
  - Pleura
  - Heart, Mediastinum
  - Peritoneum
  - Testis
- Assign the stage which includes the farthest disease extent.

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**Ethmoid and Maxillary Sinus**

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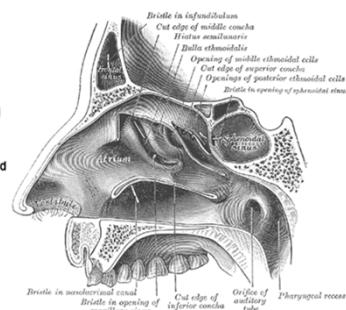
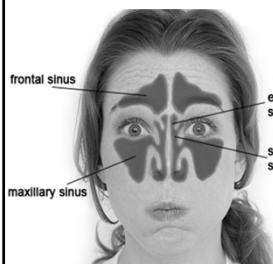
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**Anatomy**

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Primary Site/Histology

- Primary Site**
  - Maxillary C31.0
  - Ethmoid C31.1
  - Frontal C31.2
  - Sphenoid C31.3
  - Accessory, NOS C31.9
- Histology**
  - Squamous cell carcinoma
    - Keratinizing 8070/3
    - Non-keratinizing 8071/3
    - Squamous and spindle cell 8072/3
    - Transitional cell carcinoma 8074/3
    - 8120/3
- MP/H Rules-Head and Neck

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Primary Tumor

- Maxillary Sinus**
  - Things to look for
    - Confined to the mucosa
    - Bone erosion or destruction
    - Involvement of the bone at the back of the sinus
    - Moderately advanced local disease
    - Very advanced localized disease

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Primary Tumor

- Nasal cavity and ethmoid sinus**
  - Involvement of a single cavity or multiple cavities
  - Involvement of the:
    - Eye socket
    - Roof of the mouth (hard palate)
    - Cribiform plate (separates the nose from the brain)
    - Maxillary sinus
  - Moderately advanced localized disease
  - Very advanced localized disease

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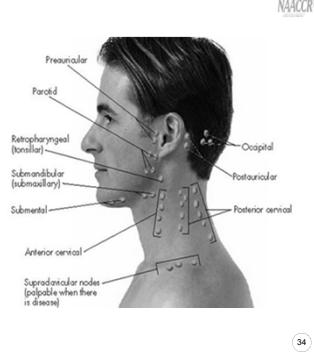
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**Regional Lymph Nodes**

- Lymph nodes are assessed based on how many lymph nodes are involved and the size of the malignancy within the lymph nodes.



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**Practice Case 2**

- An MRI revealed a large tumor in the left maxillary sinus. The tumor invasion is causing bony destruction of the lateral wall of the maxillary sinus. No further extension is identified. An enlarged retropharyngeal lymph node measuring 1.5cm is highly suspicious for malignancy. No further areas of concern were identified. A biopsy of the primary tumor confirmed squamous cell carcinoma.

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**Practice Case 2 (cont.....)**

- The patient went on to have an en bloc resection of the primary tumor and left neck dissection.
  - Final pathology
    - Tumor size-3.4cm
    - Tumor extension-Tumor involves the ethmoid sinus, hard palate, and bone of the posterior wall. Margins were negative.
  - Regional lymph nodes
    - 2 of 2 submandibular lymph nodes positive for malignancy. Metastasis within the lymph nodes was positive and measured 1.5cm's.
    - 1 of 3 retropharyngeal lymph nodes positive for malignancy. Metastasis within the lymph nodes were positive measured .5cm's
    - 0 of 6 level III lymph nodes positive for malignancy

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Practice Case 2 (cont.....)

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- How do we stage this case?

Data Items as Coded in Current NAACCR Layout				
	T	N	M	Stage Group
Clin	2	1	0	III
Path	3	2b		IVA
Summary Stage			4 Regional by Ext and LN	

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### Mucosal Melanoma

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### Mucosal Melanoma

- Rare
- Most common sites are nasal cavity and paranasal sinuses
  - Primaries in the sinuses are the most lethal
  - May also be found in the oral cavity and other mucosal sites of the head and neck
- Highly malignant
  - Even small tumors have a high rate of recurrence and mortality

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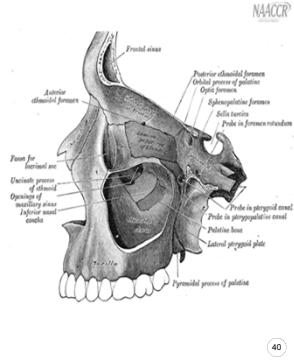
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ooooo Staging

- No T1 or T2
  - T3 would include any tumor that does not meet the criteria of moderately or very advanced
  - Moderately Advanced
    - Deep soft tissue, cartilage, bone, or skin
  - Very advanced
    - Brain, dura, skull base, lower cranial nerves, masticator space, carotid artery, prevertebral space, mediastinal structures



Staging

- Any lymph node involvement makes a mucosal melanoma a stage IV disease.
  - Hematogenous spread is relatively rare
    - Bone
    - Lung
  - Rules for classification
    - Refer to the chapter for the anatomic site where the melanoma occurred for rules for classification

## Treatment

- Primarily surgical treatment
  - Radiation
    - Can be given for local control, but does not improve overall survival
  - Chemotherapy and immunotherapy (BCG)
    - May be used as an adjuvant therapy, but have shown little success

Pop Quiz

- What Summary Stage schema would you use to code a mucosal melanoma of the ethmoid sinus?

**ETHMOID SINUS**  
C31.1  
C31.1 Ethmoid sinus

**SUMMARY STAGE**

0 In situ: Noninvasive, intraspatial  
Localized, NOS

1 Localized only  
Invasive tumor confined to ethmoid with or without bone erosion (cribriform plate)

2 Regional by direct extension only  
Extension to:  
More than one ethmoid sinus  
Antrum of orbit  
Base of skull  
Frontal sinus  
Intraorbital extension  
Maxillary sinus  
Nasal cavity, NOS  
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Practice Case 3

- MRI showed a 5mm mass confined to the left posterior ethmoid sinus. A biopsy of the tumor confirmed mucosal melanoma. No abnormal lymph nodes or indications of distant metastasis. The patient was not a surgical candidate. He opted for radiation treatment.

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Practice Case 3 (cont.....)

- How do we stage this case?

Data Items as Coded in Current NAACCR Layout				
	T	N	M	Stage Group
Clin	3	0	0	3
Path				99
Summary Stage				1-Localized

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Quiz 1

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QUESTIONS?

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Merkel Cell Carcinoma



SKIN

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Merkel Cell Carcinoma (MCC)

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- MCC is a rare and aggressive skin cancer
- The incidence of MCC is approx. 1500 new U.S. cases annually
- MCC will prove fatal in roughly 1 in 3 patients as compared to 1 in 5 patients with melanoma
- Most deaths occur within 3 years of diagnosis.

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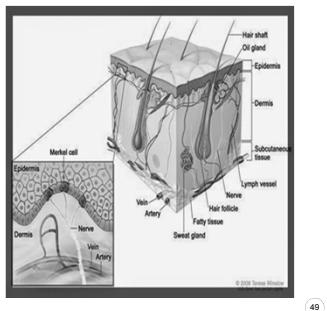
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#### ○○○○○ Anatomy

- MCC are found in the epidermis (outer layer of skin)
- MCC have both sensory and hormonal functions
- Sometimes referred to as neuroendocrine carcinoma of the skin



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#### ○○○○○ Primary Site

##### Primary Site

- Skin including skin of male and female genital sites
- If Merkel cell presents in a nodal or visceral site with primary unknown, code primary site as C44.9

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#### ○○○○○ Histology

**8247/3**

- Merkel Cell
  - Subtypes
    - Intermediate
    - Small Cell
    - Trabecular
  - Code ALL subtypes **8247/3**
- Grade – Code 9
  - Histologic grade is not used in the staging of Merkel cell carcinoma
- Special stains required to confirm - some you may see in path
  - AE1/A3, pan-keratin, CK20, CMA 5.2, synaptophysin, NSE, chromogranin, TTF-1, CK-7

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oooooo Treatment

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- Surgery is primary treatment for MCC
- Adjuvant Radiation therapy after complete resection recommended
  - To primary tumor site in node negative patients
  - To tumor site and nodal basin for patients with positive LNs
- Chemotherapy - usually reserved for Stage IV patients.
  - Sometimes advised for pathologically node positive patient in addition to radiation

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oooooo What to Look For

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Clinical Staging	Pathologic Staging
<ul style="list-style-type: none"><li>• Tumor Size</li><li>• Presence of satellite lesions</li><li>• Biopsy with H&amp;E</li><li>• Assessment of regional lymph nodes by palpation or imaging</li><li>• Bx of clinically detected LNs with appropriate immunopanel</li><li>• If LN bx positive, CT, MRI or PET</li></ul>	<ul style="list-style-type: none"><li>• Tumor Size / Depth of invasion</li><li>• Wide local excision of lesion with 1-2.5 cm negative margins (depending on site)</li><li>• SLNB for patient's who are cN0.</li><li>• Presence of ITCs in LNs</li><li>• Lymphadenectomy for patient's with cN+ nodes</li></ul>

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oooooo Primary Tumor

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- Primary Tumor
  - Based on size
  - < 2 cm
  - > 2cm
  - >5cm
  - Extension to fascia, cartilage, muscle, bone

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### Regional Lymph Nodes

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- The draining regional lymph nodes - most common site of mets
- Regional LN mets occurs frequently & early even with small tumors
  - No regional LN mets
  - Micrometastases
    - Detected on path exam only- clinically negative
    - Lymph nodes with Isolated Tumor cells are considered POSITIVE
  - Macrometastases
    - Clinically evident
  - In transit metastasis
    - Satellite skin lesions are considered "in transit metastasis" and assigned N2

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### Distant Metastasis

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- Distant metastasis is subdivided based on location of metastasis
- Distant skin, subcutaneous tissues or distant LNs
  - Lung
  - All other visceral sites

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### Staging

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Sub stage groups based on whether LNs were microscopically **Proven Negative**.

T	N	M	Clinical Stage Group	Pathologic Stage Group
T1	pN0	M0		Stage IA
T1	cN0	M0	Stage IB	
T2/T3	pN0	M0		Stage IIA
T2/T3	cN0	M0	Stage IIB	
T4 Extracutaneous	c/p N0	M0	Stage IIC	Stage IIC
Any T	<b>pN1a</b> "Micrometastasis" or + ITCs & Clinically occult	M0		Stage IIIA
Any T	<b>cN1/N1b/N2</b> "Macroscopic" Clinically Evident & path confirmed	M0	Stage IIIB	
Any T	Any N	M1	Stage IV	Stage IV

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**Practice Case 4**

- 71-year old female with hx of CLL.
- Presents with painless mass left forearm. States it is growing quickly. Prior history of melanoma.
- PE reveals a 3.1 cm diameter vibrant red nodular lesion area on left arm.
- No palpable axillary, supraclavicular or cervical LAD.
- Per MD lesion is presumed malignant.
- Pt declines bx; prefers to have one definitive surgical procedure to excise lesion.

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**Practice Case 4 (cont....)**

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- Patient had wide local excision and sentinel LN biopsy.
- Final Pathology: 2.9 x 1.9 cm ellipse of erythematous skin within which is a 2.2 x 0.8 cm Merkel Cell Ca confirmed on IHC stains. Deep margin was positive. Tumor extended to a depth of 1.5 cm. Additional 2.5cm margin were submitted with no residual tumor, for clear surgical margins.
- Sentinel lymph node left axilla, biopsy: 1 LN negative for malignancy by routine stain and H&E with No ITCs present.

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**Practice Case 4 (cont....)**

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- How do we stage this?

	Data Items as Coded in Current NAACCR Layout			
	T	N	M	Stage Group
Clin				
Path				
Summary Stage:				

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Practice Case 4 (cont.....)

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CS Site Specific Factors	Practice Case Code
SSF 1 Measured Thickness (Depth)	
SSF 3 Clinical Status of LN Mets	
SSF16 Size of metastasis in Lymph nodes	
SSF17 Extracapsular Extension of Regional LNs (clinically and pathologically)	
SSF18 Isolated Tumor Cells (ITC) in Regional Lymph nodes	
SSF 22 Profound Immune Suppression	

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Schema

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- TNM
  - Merkel Cell Carcinoma
  - Carcinoma of the eyelid for eyelid primary
- CS has separate schemas for Merkel Cell Ca
  - Penis
  - Scrotum
  - Vulva
- Summary Stage
  - Skin (except eyelid)
  - Skin of the eyelid
  - Penis
  - Other and Unspecified Male Genital Organs
  - Vulva

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Ophthalmic Primaries

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## Melanoma of the Eye

ooooo Uvea  
Conjunctiva

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ooooo Primary Site and Histology

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- Primary Site
  - Conjunctiva C69.0
  - Retina C69.2
  - Choroid C69.3
  - Ciliary Body C69.4
  - Iris C69.4
  - Eye C69.9
- Histology
  - Melanoma, nos 8720/3
  - Spindle cell melanoma 8772/3
  - Mixed cell melanoma (epithelioid and spindle cell) 8770/3
  - Epithelioid cell melanoma 8771/3

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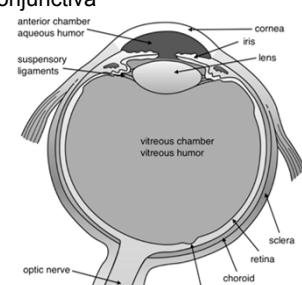
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ooooo Melanoma of the Uvea and Conjunctiva

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- The uvea is the vascular middle layer of the eye
- The uvea consists of three parts
  - Iris
  - Ciliary body
  - Choroid



The diagram illustrates a cross-section of the eye. It shows the anterior chamber containing aqueous humor, the iris, suspensory ligaments, and the lens. The vitreous chamber contains vitreous humor. The posterior part of the eye includes the optic nerve, choroid, retina, fovea, and sclera.

[https://commons.wikimedia.org/wiki/File:Three\\_Internal\\_chambers\\_of\\_the\\_Eye.png](https://commons.wikimedia.org/wiki/File:Three_Internal_chambers_of_the_Eye.png)

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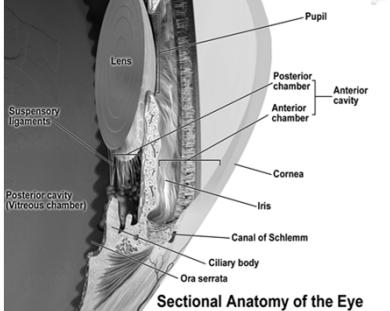
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oooooo Iris

- Consists of two layers
  - Stroma
  - Epithelial
- Epithelial layer includes melanin that gives the eye color



**Sectional Anatomy of the Eye**

[https://commons.wikimedia.org/wiki/File:Blausen\\_0390\\_EyeAnatomy\\_Sectional.png](https://commons.wikimedia.org/wiki/File:Blausen_0390_EyeAnatomy_Sectional.png)

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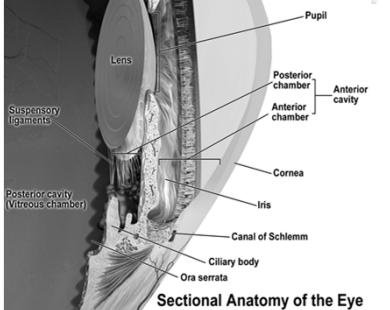
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oooooo Ciliary Body

- Ciliary body epithelium consists of two layer.
  - Outer pigmented layer
  - Inner non-pigmented layer
- Glaucoma may occur if invasion of the iridocorneal angle



**Sectional Anatomy of the Eye**

[https://commons.wikimedia.org/wiki/File:Blausen\\_0390\\_EyeAnatomy\\_Sectional.png](https://commons.wikimedia.org/wiki/File:Blausen_0390_EyeAnatomy_Sectional.png)

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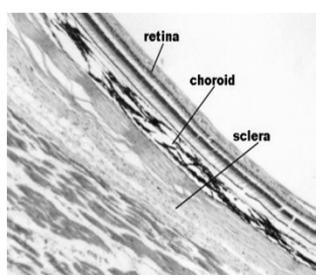
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oooooo Choroid

- Located beneath the retina
- The most common site for melanoma
  - The largest collection of melanocytes in the eye.
  - Receives the most ultraviolet radiation



<http://www.daviddarling.info/encyclopedia/C/choroid.html>

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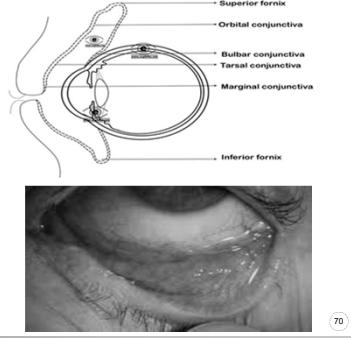
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**Conjunctiva**

- Three subcategories
  - Palpebral conjunctiva
  - Fornicles
  - Bulbar conjunctiva
- Two layers
  - Epithelium
  - Substantia propria



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**Staging**

- Clinical Staging
  - Slit lamp examination
  - High frequency ultrasound
  - Low frequency ultrasound
  - PET/CT
  - Biopsy
  - Palpation of regional lymph nodes
- Pathologic Staging
  - Complete resection of the primary site
    - Iridectomy
    - Enucleation of globe
    - Removal of sentinel or palpable lymph nodes

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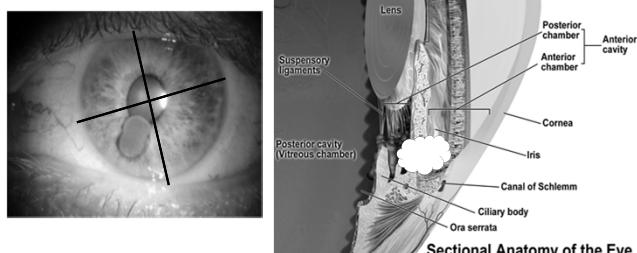


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**Primary Tumor- Iris**



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**Sectional Anatomy of the Eye**

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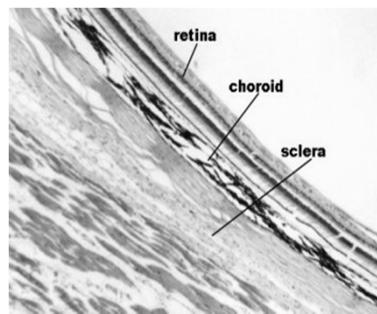
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Primary Tumor Ciliary Body and Choroid

- Primary Tumor is assessed based on...
  - Tumor size categories 1-4
    - Thickness
    - Largest diameter
  - Ciliary body involvement
  - Extraocular extension



SSF's

- Measured Basal Diameter
- Measured Thickness (Depth)
- Size of Largest Mets
- Status of chromosomes 3q, 6q, and 8q
- Mean Diameter Nucleoli (MLN)
- Extravascular Matrix Patterns, Loops
- Extravascular Matrix Patterns, Networks

Treatment

- Surgery
  - Iridectomy
  - Enucleation
  - Sentinel lymph node biopsy
  - Excision of palpable lymph nodes
- Radiation
  - Proton therapy
  - Plaque radiation therapy
- Chemotherapy
  - Topical-mitomycin, 5-fu
  - Interferon alpha 2b

**Practice Case 5**

- A patient presents with complaints of flashing lights and floating specks. Examination showed a medium sized tumor within the eye. Ophthalmoscopy and ultrasound showed the tumor was 9.3 in thickness and 15.5mm in basal diameter and extended into the ciliary body. The tumor was causing partial retinal detachment. No extraocular involvement was identified. Additional work-up did reveal any metastasis. Based on the exam the physician made the diagnosis of choroid melanoma. The patient was treated with plaque radiotherapy.

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**Practice Case 5 (cont....)**

- How do we stage this case?

Data Items as Coded in Current NAACCR Layout

	T	N	M	Stage Group
Clin	3b	0	0	IIIA
Path				99
Summary Stage				1-Localized

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**Retinoblastoma**

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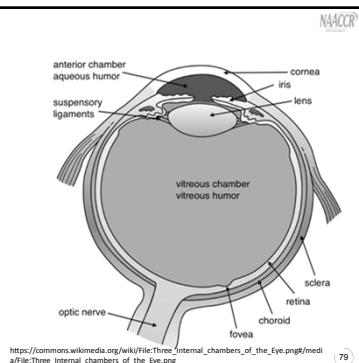
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### Retinoblastoma

- Develops in the lining on the inside of the eye
- Commonly affects young children
- Bilateral involvement is common
- Trilateral retinoblastoma
  - Bilateral retinoblastoma
  - Pinealblastoma of the pineal gland



### Primary Site and Histology

- Primary Site
  - Retina C69.2
- Histology
 

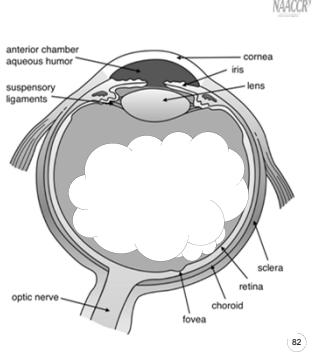
• Retinoblastoma, nos	9510/3
• Differentiated	9511/3
• Diffuse	9513/3
• Spontaneously regressed	9514/3
• Undifferentiated	9512/3
- Multiple Primary Rule
  - Rule M4 (Other) Retinoblastoma is always a single primary (unilateral or bilateral).

### Staging Systems

- International Classification for Intraocular Retinoblastoma
- The Reese-Ellsworth staging system-groups 1-5
  - Group 2 (favorable for saving [or preserving] the eye)**
    - 2A: one tumor, 4 to 10 DD, at or behind the equator
    - 2B: multiple tumors, with at least one 4 to 10 DD, and all at or behind the equator
  - Group 3 (doubtful for saving [or preserving] the eye)**
    - 3A: any tumor in front of the equator
    - 3B: one tumor, larger than 10 DD, behind the equator
- AJCC
- Summary Stage

**ooooo Staging**

- Primary Tumor
  - Volume of the tumor within the globe
  - Absence or presence of seeding
  - Invasion of optic nerve
  - Choroid invasion
- Lymph nodes
  - No lymphatics in globe
- Metastasis



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**ooooo Pop Quiz**

- Since a bilateral retinoblastoma is considered a single primary, do we stage as a single disease?
  - No. Each eye should be staged separately.

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**ooooo Treatment**

- Surgery
  - Enucleation
- Radiation Therapy
  - Beam radiation
  - Brachytherapy (plaque radiotherapy)
- Laser Therapy (photocoagulation)
- Cryotherapy
- Chemotherapy
  - Neoadjuvant
  - adjuvant
- Clinical Trials

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**Practice Case 6**

- A patient presented with a previously diagnosed retinoblastoma in the left eye. On slit lamp exam the tumor was found to involve slightly more than 2/3's of the globe. Seeding was present. No indication that the optic nerve was involved. The patient received neoadjuvant chemotherapy followed by enucleation.
- Pathology from enucleation
  - A single tumor measuring 2mm confined to the retina. No additional tumor identified

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**Practice Case 6 (cont....)**

- How do we stage this case?

Data Items as Coded in Current NAACCR Layout

	T	N	M	Stage Group
Clin	3a	0	0	99
Path	1	X		99
Summary Stage				1-Localized

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Stage Descriptor of Y

**Gestational Trophoblastic Tumors (GTT)**

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### Gestational Trophoblastic Tumors (GTT)

- Rare group of tumors which develop in the uterus after conception.
- These tumors arise from the placental tissue - trophoblast cells.
- Gestational trophoblastic tumors are uncommon and occur in 1 of 1,000 pregnancies in the U.S.
- More common in many Asian and African countries
- Occur as a result of a genetic accident.

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### Gestational Trophoblastic Tumors

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Two main groupings:

- *Premalignant conditions*
  - *Hydatidiform Moles (HM)*
    - Complete & partial moles, or "invasive" mole
    - Slow-growing and *Most Often* benign
    - Can be precursor for GTT
- *Gestational Trophoblastic Neoplasia (GTN)*
  - Several types
  - *Almost always* malignant

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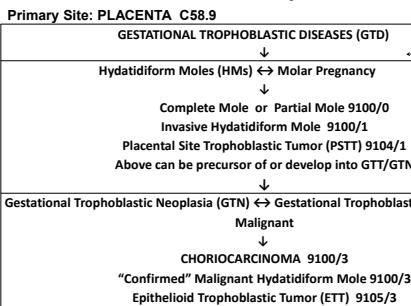
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### Gestational Trophoblastic Tumors

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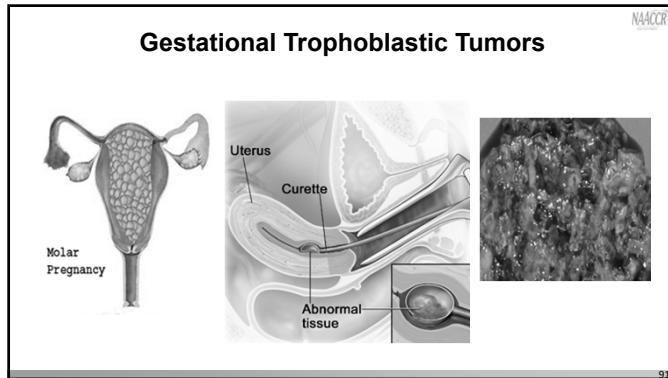
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ooooo Treatment

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- Depends on Histologic type, Stage, Risk grouping of GTT and patient preferences (fertility sparing Rx)
- Surgery (D&C or Complete Hysterectomy)
- Chemotherapy (Methotrexate, Dactinomycin)
- Radiation Therapy - to sites of distant spread
- Any combination of 2 or more above

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ooooo POP QUIZ

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- Is Gestational Trophoblastic Neoplasia (GTN) reportable if there is no mention of metastasis but the patient has been treated with chemotherapy?

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 Primary Tumor

- Tumor confined to uterus
- Tumor extends to other GYN structures
  - Ovary
  - Tubes
  - Vagina
  - Broad Ligaments
  - By metastases or direct extension

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 Regional Lymph Nodes

- There is no regional nodal designation in the staging of these tumors in TNM
- Any nodal metastases should be classified as metastatic (M1) disease

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 Metastasis

- Lungs
  - Most frequent site
- Other Distant Mets
  - Kidney
  - Gastrointestinal tract
  - Spleen
  - Liver
  - Brain

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**Gestational Trophoblastic Tumors**

**Prognostic Risk Score**

Used to assign a sub-stage in TNM & collected in CS Site Specific Factor 1

Prognostic Factor	Risk Score			
	0	1	2	4
Age Previous Pregnancy	<40 Hydatidiform Mole	>40 Abortion	- Full-term Pregnancy	-
Months since last pregnancy	<4	4 to 6	7-12	>12
Pretreatment hCG(IU/ml)	<10 <sup>3</sup>	10 <sup>3</sup> - ≤10 <sup>4</sup>	10 <sup>4</sup> - <10 <sup>5</sup>	>10 <sup>5</sup>
Largest Tumor size	<3cm	3- <5cm	> 5cm	-
Site of Mets	Lung	Spleen, Kidney	GI Tract	Brain, Liver
Number of Mets	0	1-4	5-8	>8
Previous failed chemotherapy	None	None	Single Drug	Two or more drugs
LOW RISK = 6 or less		HIGH RISK = 7 or greater		97

**Gestational trophoblastic Tumors**

**CS Site Specific Factor 1 – Prognostic Risk Score**

- Code the Clinician stated risk score**
  - Stated value, e.g. "Risk score of 7"
  - Low risk (6 or less) or Stage A
  - High risk (7 or greater) or Stage B
- If MD risk score not available**
  - Registrar may assign if ALL information available.
  - If any one of the risk factors is unknown, assign code 999.....

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**Staging**

- TNM
  - Gestational Trophoblastic Tumors Chapter 39
- Collaborative Stage
  - Placenta
- Summary Stage
  - Placenta

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**Practice Case 7**

- 4/1/15: 45 year old female with postmenopausal bleeding and pelvic pain. Abdomen tender. No palpable LAD. H&P otherwise WNL.
- US: 5cm cystic intrauterine mass w/invasion into uterine wall.
- 4/3/15 D&C positive for malignant trophoblastic tumor, likely choriocarcinoma.
- B-hCG 69,000
- 4/10/15 CT C/A/P: No distant mets.
- 4/16/15 TAH/BSO with pathology revealing high grade Choriocarcinoma, Grade 3, involving superficial myometrium, with extension into cervix and upper vagina.
- Per MD Risk Factor, High

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**Practice Case 7 (cont....)**

- How do we stage this case?

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Data Items as Coded in Current NAACCR Layout					
	T	N	M	Risk Factors	Stage Group
Clin					
Path					
Summary Stage					

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**POP QUIZ**

- What is the number one risk factor for development of a Gestational Trophoblastic Disease/Tumor?
  - Age
  - History of previous molar pregnancy
  - Elevated Beta hCG
  - Any previous pregnancy

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Questions?

Quiz 2

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Coming Up...

• Collecting Cancer Data: Pharynx  
• 11/5/15

• Directly Coded Cancer Stage...NOW  
• 12/3/15

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And the Winners Are....



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CE Certificate Quiz/Survey

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- Phrase
  - Mesothelioma
- Link
  - <http://www.surveygizmo.com/s3/2352129/Unusual-Sites-and-Histologies>

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