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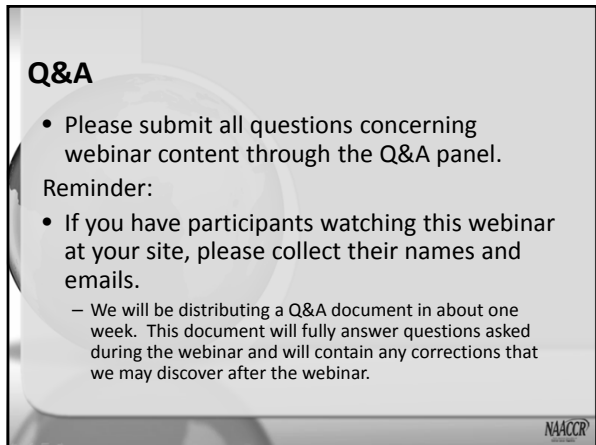
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**Agenda**

- Overview/Treatment
  - Quiz 1
- Collaborative Stage Data Collection System
  - Quiz 2
- Case Scenarios



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**OVERVIEW**



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**Key Statistics**

- Estimated new cases and deaths from brain and other nervous system cancers in the United States in 2012
  - New Cases 22,910
  - Deaths 13,700
- The incidence of primary CNS primaries has risen over the last 30 years, especially in the elderly



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**Tumor Types**

- Anaplastic gliomas and glioblastoma multiforme
- Low grade infiltrative astrocytomas
- Oligodendroglioma
- Ependymomas
- Meningiomas
- Primary spinal cord tumors
- Primitive neuroectodermal tumors (PNET)

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**Case Eligibility for CNS Tumors**

- Include ICD-O-3 malignant (behavior code 2, 3) and ICD-O-3 nonmalignant (behavior code 0, 1) diagnosed on or after 1/1/2004 tumors of the following sites:
  - Meninges (C70.\_)
  - Brain (C71.\_)
  - Spinal cord, cranial nerves, and other parts of CNS (C72.\_)
  - Pituitary gland (C75.1)
  - Craniopharyngeal duct (C75.2)
  - Pineal gland (C75.3)

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**Reportable Terms**

- “Neoplasm” and “Tumor”
  - Beginning with 2004 diagnoses and only for C70.0–C72.9, C75.1–75.3
  - Terms for nonmalignant primary intracranial and central nervous system tumors only
    - Section 1 page 3 FORDS

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**Equivalent Terms**

- 2007 Multiple Primary and Histology Rules, General Instruction Page 9
  - Equivalent Terms
    - Tumor
    - Mass
    - Lesion
    - Neoplasm
- ONLY equivalent when determining the number of primaries or histology
- Should NOT be used to determine reportability



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**Hemangioma**

- 9120/0 Hemangioma, NOS and 9121/0 Cavernous hemangioma are reportable when they arise in the dura or parenchyma of the CNS.
- 9122/0 Venous hemangioma is not reportable



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**Benign and Borderline**

- Benign and borderline intracranial and CNS neoplasms must meet two conditions to be reportable:
  - The histology must be reportable **AND**
  - The primary site must be reportable



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### Cranial Tumors

- Report neoplasms described as intradural or intracranial
- Do not report cranial neoplasms described as extradural

Cross-section of skull and the Meninges

© 2004 American Society of Clinical Oncology

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### Case Eligibility

- Juvenile astrocytoma is listed in the ICD-O 3 manual as 9421/1
  - Record in the registry as 9421/3

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### Sequence Number

- Records sequence of malignant and nonmalignant neoplasms over patient's lifetime
  - 00-59 and 99 for malignant and in situ behavior
    - 00 = solitary malignant neoplasm
    - 01 = first of multiple malignant neoplasms
  - 60-88 for non-malignant behavior
    - 60 = solitary non-malignant neoplasm
    - 61 = first of multiple non-malignant neoplasms

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**Anatomy**

- YouTube video
  - <http://www.youtube.com/watch?v=78kW3vNO2YU>



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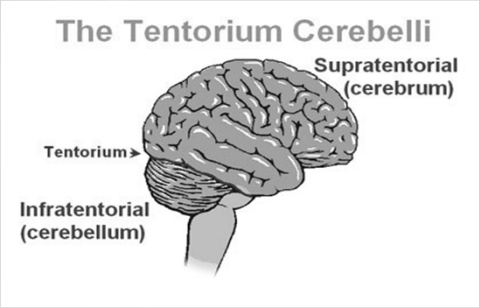
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**Location of Intracranial Tissues**

**The Tentorium Cerebelli**




Supratentorial (cerebrum)

Tentorium

Infratentorial (cerebellum)

Image source: SEER Training Website



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**Location of Intracranial Tissues**

- Supratentorial sites
  - Cerebrum
    - Frontal, temporal, parietal, and occipital lobes
  - Meninges of cerebrum
  - Ventricle, NOS
    - Lateral & 3rd
  - Corpus callosum
  - Tapetum
  - Anterior cranial fossa
  - Middle cranial fossa
  - Suprasellar
- Infratentorial sites
  - Cerebral subsites
    - Hypothalamus
    - Pallium
    - Thalamus
  - Cerebellum
  - Meninges of cerebellum
  - Brain Stem
    - 4<sup>th</sup> ventricle
  - Posterior cranial fossa



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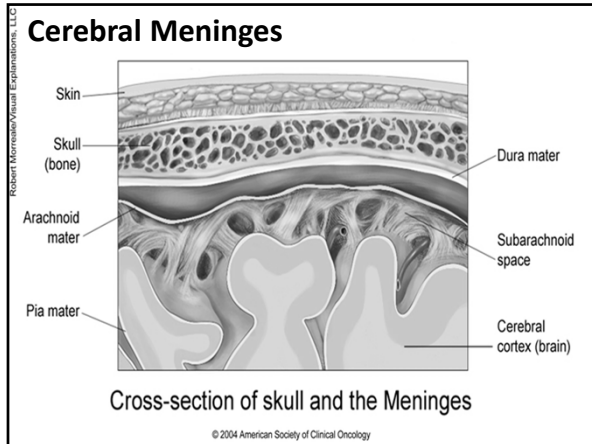
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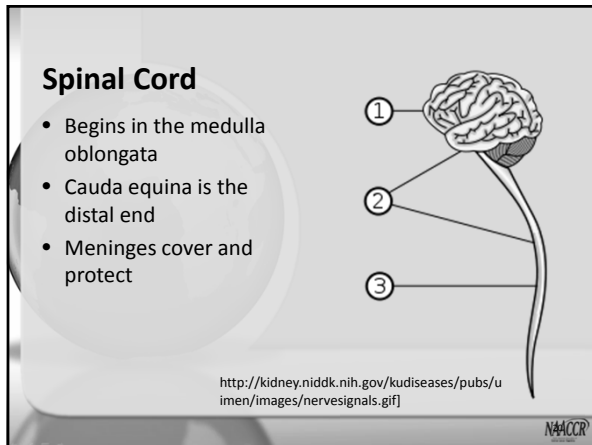
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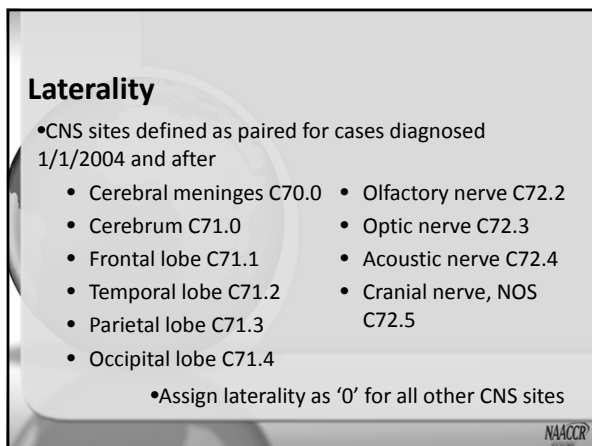
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**Grade/Differentiation**

- Do not record the WHO Grade, Anne/Mayo, or Kernohan grades in the grade field
  - Record the WHO grade in the appropriate CS data item
  - If no grade is given, code 9 (unknown)
- Anaplastic is synonymous with undifferentiated and should be assigned grade 4

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**World Health Organization (WHO) Grading System**

- Grade I
  - Benign = non-cancerous
  - Slow growing
  - Cells look almost normal under a microscope
  - Usually associated with long-term survival

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**WHO Grading System**

- Grade II
  - Relatively slow growing
  - Sometimes spreads to nearby normal tissue and comes back (recurs)
  - Cells look slightly abnormal under a microscope
  - Sometimes comes back as a higher grade tumor

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**WHO Grading System**

- Grade III
  - Malignant = cancerous
  - Actively reproduces abnormal cells
  - Tumor spreads into nearby normal parts of the brain
  - Cells look abnormal under a microscope
  - Tends to come back, often as a higher grade tumor

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**WHO Grading System**

- Grade IV
  - Most malignant
  - Grows fast
  - Easily spreads into nearby normal parts of the brain
  - Actively reproduces abnormal cells
  - Cells look very abnormal under a microscope
  - Tumor forms new blood vessels to maintain rapid growth
  - Tumors have areas of dead cells in their center (called necrosis)

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**Tumor Types**

Gliomas  
↓

- Anaplastic gliomas and glioblastoma multiforme
- Low grade infiltrative astrocytomas
- Oligodendroglioma
- Ependymomas

- Meningiomas
- Primary spinal cord tumors
- Primitive neuroectodermal tumors (PNET)

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### Glioma Classification

- Cell type
  - Ependymal cells
  - Astrocytes
  - Oligodendrocytes
  - Mixed glioma
    - Oligoastrocytoma
- WHO Grade
  - Low grade-WHO II
  - High grade-WHO III-IV
- Location
  - Supratentorial
  - Infratentorial

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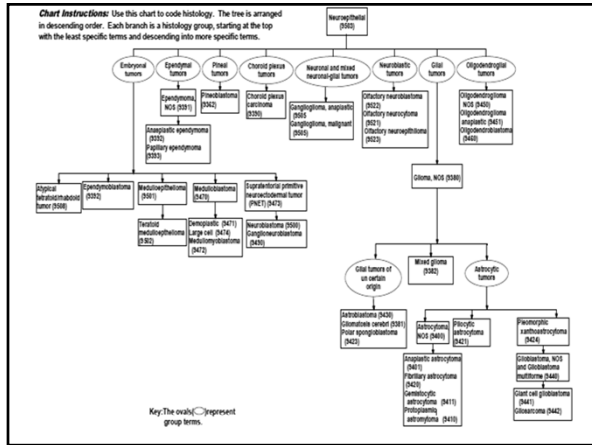
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### Low Grade Astrocytoma and Oligodendroglioma

- Astrocytoma
  - Pilocytic astrocytoma (9421/1)
  - Pleomorphic xanthoastrocytoma (9424/3)
  - Diffuse astrocytoma (9400/3)
- Oligodendroglioma (9450/3)

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**Treatment**

- Surgery
  - Total gross resection
  - Stereotactic biopsy
  - Open biopsy
  - Subtotal resection
- Radiation
- Watchful Waiting



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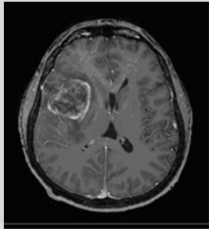
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**Anaplastic Gliomas and Glioblastoma**

- Anaplastic astrocytoma
  - WHO Grade III
  - 7% of all gliomas
  - 27% 5-year survival
- Anaplastic Oligodendroglioma
  - WHO Grade III
  - 4% of all glioma
  - Primarily occur in adults 50-60 years old



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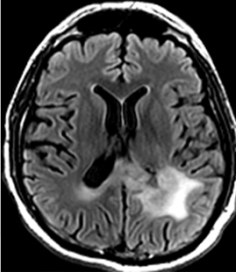
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**Anaplastic Gliomas and Glioblastoma**

- Glioblastoma
  - WHO Grade IV
  - 54% of all gliomas
  - 5% 5-year survival



Belden C J et al. Radiographics 2011;31:1717-1740



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**Treatment**

- Surgery
  - Total gross resection of the tumor
  - Subtotal resection
  - Stereotactic or open biopsy
- Radiation Therapy
  - Standard adjuvant treatment after surgery
- Chemotherapy
  - Temozolomide
  - PCV
  - Carmustine wafers (intraoperative)



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**Ependymoma**

- Grade I
  - Subependymoma (9383/1)
  - Myxopapillary ependymoma (9394/1)
- Grade II
  - Ependymoma, nos (9391/3)
- Grade III
  - Anaplastic ependymoma (9392/3)



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**Treatment**

- Grade II Ependymoma
  - Gross total resection
  - If subtotal resection, then adjuvant radiation
- Grade III Ependymoma
  - Gross total resection
  - Adjuvant radiation



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### Treatment

- Grade III
  - Gross total resection followed by radiation
  - If not a surgical candidate, radiation alone.
- Grade II
  - Observation if asymptomatic and tumor is less than 30mm
  - Gross total resection
  - Subtotal resection with adjuvant radiation
- Grade I
  - Observation if asymptomatic and tumor is less than 30mm
  - Gross total resection
  - Subtotal resection with a radiation if the tumor is more than 30mm




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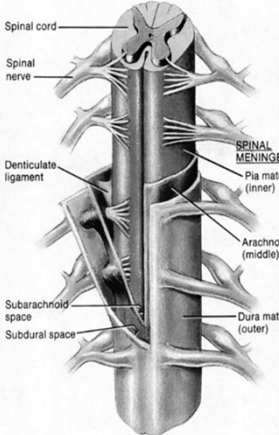
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### Primary Spinal Cord Tumors

- Extradural
  - Usually mets
- Intradural-extramedullary
  - Usually meningiomas
- Intradural-intramedullary
  - Usually astrocytomas in children
  - Usually ependymomas in adults




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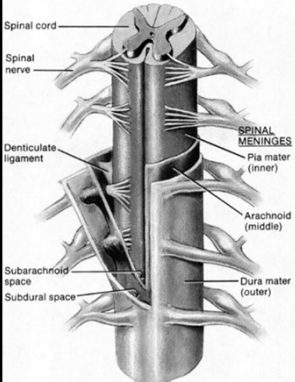
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### Spinal Nerve Tumors



- Neoplasms arising from the dura covering the spinal cord roots are meningiomas.
- Neoplasms arising in the spinal nerve roots are primarily Schwannomas and neurofibromas.
- The peripheral nerves are the portion of nerve extending beyond the spinal dura.
  - Benign /0 or borderline /1 neoplasms of the peripheral nerves are not reportable.




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**Question**

- Are Schwannomas reportable?



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**Answer**

- Reportability depends on the primary site:  
When they originate in the intracranial (intradural) or intraspinal space they are reportable.
  - Data Collection Answers from the CoC, NPCR, SEER Technical Workgroup,  
<http://www.seer.cancer.gov/registrars/data-collection.html#reportability>



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**Treatment**

- WHO Grade I meningiomas and peripheral nerve sheath tumors
  - Observation if asymptomatic
  - Surgery if symptomatic
  - Radiation if symptoms persist after treatment
- WHO Grade I astrocytoma and ependymoma
  - Gross total resection
- WHO Grade II and higher
  - Partial resection



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**Meningioma**

- Grade I
  - Meningioma, nos (9530/0)
  - 92% of all meningiomas
- Grade II
  - Atypical meningioma (9539/1)
  - 6% of all meningiomas
- Grade III
  - Malignant melanoma (9530/3)
  - 2% of all meningiomas

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**Treatment**

<p><b>WHO Grade I or Grade II</b></p> <ul style="list-style-type: none"> <li>• Observation           <ul style="list-style-type: none"> <li>– Asymptomatic</li> <li>– Tumor &lt;30 mm</li> </ul> </li> <li>• Surgery           <ul style="list-style-type: none"> <li>– Symptomatic</li> <li>– Surgical candidate</li> <li>– Tumor &gt;30mm</li> </ul> </li> <li>• Radiation           <ul style="list-style-type: none"> <li>– Tumor &gt;30mm</li> <li>– Non-surgical candidate</li> </ul> </li> </ul>	<p><b>WHO Grade III</b></p> <ul style="list-style-type: none"> <li>• Surgery</li> <li>• Adjuvant radiation</li> </ul>
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**Primitive Neuroectodermal Tumors (PNET)**

- Medulloblastoma (Infratentorial) or Supratentorial
- WHO Grade IV
- Frequently metastasized to the cerebral spinal fluid
- Rare disease
  - More common in children than adults

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**Treatment**

- Surgery
  - Gross total resection whenever possible
- Adjuvant radiation
- Adjuvant systemic treatment



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**TREATMENT TIPS**



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**Surgery Codes**

- 20 Local excision of tumor, lesion or mass; excisional biopsy
  - Used when the surgeon describes the procedure “biopsy,” or “excisional biopsy”, or when there are no details about the procedure
  - Unknown whether total or partial tumor resected
- 21 Subtotal resection of tumor, lesion or mass in brain
  - Near total, partial, subtotal, debulking, open biopsy (if residual tissue)
- 22 Resection of tumor of spinal cord nerve



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**Surgery Codes**

- 30 Radical, total, gross resection of tumor, lesion or mass in brain
  - The resection of the brain tissue surrounding the tumor is limited to ensure clean margins.
  - New code can be used with all cases regardless of diagnosis year.



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**Surgery Codes**

- 40 Partial resection of lobe of brain, when the surgery can not be coded as 20-30.
  - Less than lobectomy, but more than it would be necessary to ensure clean margins (when you can not code to 20 or 30)
- 55 Gross total resection
  - Lobectomy



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**Radiation**

- External beam radiation
  - Codes 20 – 30: Orthovoltage, cobalt, photons, electrons, or neutrons
  - Code 31: Intensity modulated radiation therapy
    - IMRT
  - Code 32: Conformal radiation
    - 3D conformal radiation



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**Treatment Modality**

- Radiosurgery
  - Code 40: Particle or proton beam
  - Code 41: Stereotactic radiosurgery NOS
  - Code 42: Linac radiosurgery
    - Cyberknife
  - Code 43: Gamma knife



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**Multiple Primary and Histologies**

- Rules are based on behavior of the tumor
  - Benign and Borderline Intracranial and CNS Tumors
  - Malignant Meninges, Brain, Spinal Cord, Cranial Nerves, Pituitary gland, Craniopharyngeal duct and Pineal gland



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**MULTIPLE PRIMARY RULES**



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### Multiple Tumors

- Rule M6
  - A glioblastoma or glioblastoma multiforme (9440) following a glial tumor is a single primary (See Chart 1).
  - Glioblastoma:
    - A malignant rapidly growing Astrocytoma of the central nervous system. These neoplasms grow rapidly, invade extensively, and occur most frequently in the cerebrum of adults. Any glial tumor can recur as a glioblastoma or a glioblastoma multiforme

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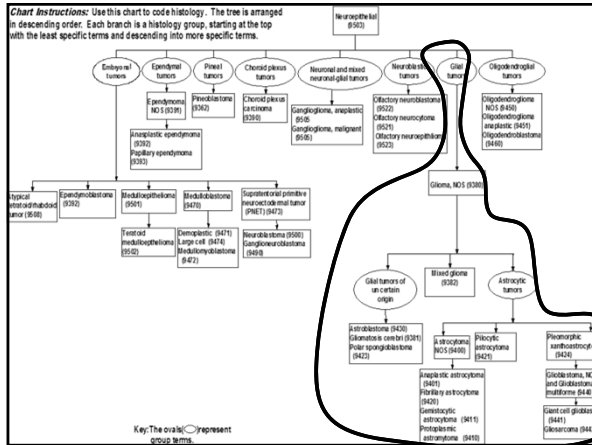
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### Multiple Tumors

- Rule M7
  - Tumors with ICD-O-3 histology codes on the same branch in Chart 1 or Chart 2 are a single primary.
- Rule M8
  - Tumors with ICD-O-3 histology codes on different branches in Chart 1 or Chart 2 are multiple primaries.

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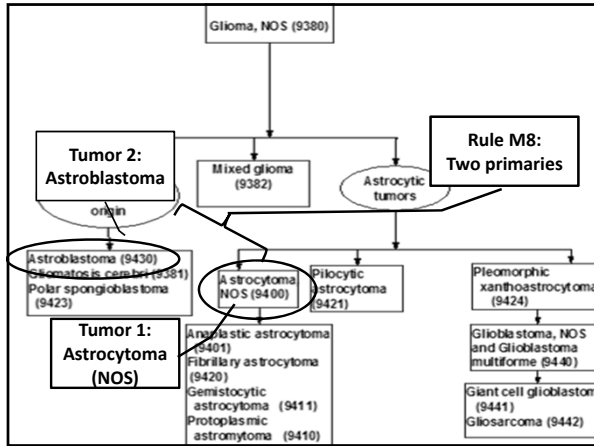
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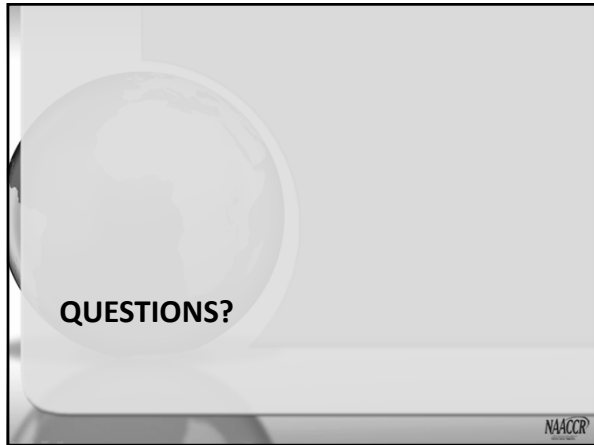
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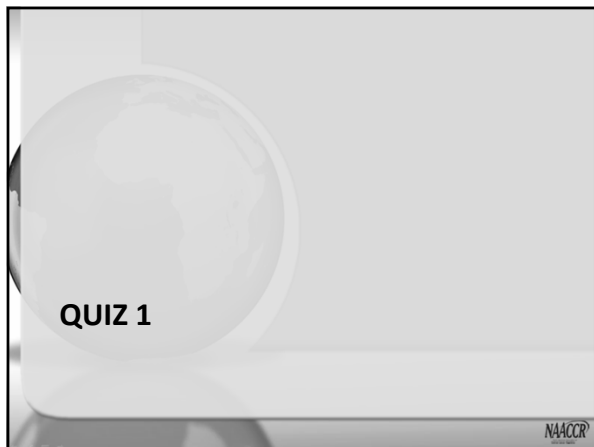
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Bone & Soft Tissue  
**COLLABORATIVE STAGE DATA  
 COLLECTION SYSTEM (CSV02.04)**

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**CNS Schemas**

Schema Name	Site Codes
Brain	C70.0, C71.0-C71.9
CNSOther	C70.1, C70.9, C72.0-C72.5, C72.8-C72.9
IntracranialGland	C75.1, C75.2, C75.3

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**Brain Schema**

- Cerebrum
- Frontal lobe
- Temporal lobe
- Parietal lobe
- Occipital lobe
- Ventricle
- Cerebellum
- Brain Stem
- Overlapping lesion of brain
- Brain, NOS
- Cerebral meninges

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**CS Extension: Brain**

<b>Supratentorial Sites</b>	<b>Infratentorial Sites</b>
<ul style="list-style-type: none"><li>• C71.0 except hypothalamus, pallium, thalamus</li><li>• C71.1-C71.5</li><li>• C71.8: Corpus callosum, tapetum</li><li>• C71.9: Anterior cranial fossa, middle cranial fossa, suprasellar</li></ul>	<ul style="list-style-type: none"><li>• C71.0: Hypothalamus, pallium, thalamus</li><li>• C71.6-C71.7</li><li>• C71.9: Posterior cranial fossa</li></ul>

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**CS Extension: Brain**

- Code 050
  - Benign or borderline
- Codes 100-510
  - Confined to brain or cerebral meninges
    - Supratentorial tumor
    - Infratentorial tumor
    - Crosses midline
    - Crosses tentorium cerebelli
- Codes 600-800
  - Extension beyond brain or cerebral meninges
  - 710: Circulating cells in CSF

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**CS Mets at DX: Brain**

- 00: No distant metastasis
- 20: Drop metastasis
- 30: Metastasis outside the CNS (extra-neural)
- 50: 20 + 30
- 99: Unknown

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**Question?**

- If a patient has a brain tumor with effacement of the lateral ventricles, is the CS Extension code 300 (invades or encroaches upon ventricular system)?
- Effacement of ventricles is due to mass effect on the ventricles, but the mass effect may not necessarily be caused directly by tumor in the ventricles. Use code 300 when there is involvement of the tumor in the ventricular system.

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**CNSOther Schema**

- Spinal meninges
- Meninges, NOS
- Spinal cord
- Cauda equina
- Olfactory nerve
- Optic nerve
- Acoustic nerve
- Cranial nerve, NOS
- Overlapping lesion of brain and central nervous system
- Nervous system, NOS

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**CS Extension: CNSOther**

- Code 050
  - Benign or borderline
- Codes 100-300
  - Confined to CNS site of origin
- Codes 400-600
  - Extension to adjacent structures or tissues
- Codes 700-800
  - Further contiguous extension

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**CS Mets at DX: OtherCNS**

- 00: No distant metastasis
- 10: Distant lymph nodes
- 40: Distant metastasis except distant lymph nodes; carcinomatosis
- 50: 10 + 40
- 60: Distant metastasis, NOS
- 99: Unknown



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**Pop Quiz**

- Final diagnosis: Myxopapillary ependymoma of the cauda equina with a drop metastasis in cauda equina.
- How is the drop metastasis coded?
  - CS Extension = 800 (further contiguous extension)
  - CS Mets at DX = 40 (distant metastasis except distant lymph nodes)



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**IntracranialGland Schema**

- Pituitary gland
- Craniopharyngeal duct
- Pineal Gland



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**CS Extension: Intracranial Gland**

- Code 000
  - In situ
- Code 050
  - Benign or borderline
- Codes 100-300
  - Confined to intracranial gland
- Codes 400-600
  - Extension to adjacent structures or tissues
- Codes 800
  - Further contiguous extension

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**CS Mets at DX: Intracranial Gland**

- 00: No distant metastasis
- 10: Distant lymph nodes
- 40: Distant metastasis except distant lymph nodes; carcinomatosis
- 50: 10 + 40
- 60: Distant metastasis, NOS
- 99: Unknown

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**Pop Quiz**

- Final diagnosis: Pineal gland with pineoblastoma with 2<sup>nd</sup> tumor deposit in anterior horn of lateral ventricle of brain.
- How is the drop 2<sup>nd</sup> tumor deposit coded?
  - CS Extension = 800 (further contiguous extension)
  - CS Mets at DX = 40 (distant metastasis except distant lymph nodes)

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**Other CS Data Items for CNS Schemas**

- CS Tumor Size/Ext Eval = 9
- CS Lymph Nodes = 988
- CS Lymph Nodes Eval = 9
- Regional Nodes Positive = 99
- Regional Nodes Examined = 99
- CS Mets Eval = 9

NAACCR

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**SSF1: WHO Grade Classification**

- Histologic grading classification for CNS tumors
- Important prognostic factor for response to treatment & outcomes for CNS tumors
- Not the same as ICD-O-3 grade/differentiation
  - Do NOT code WHO grade in grade data item
  - Do NOT code terminology like well, moderately, or poorly differentiated in SSF1

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**SSF1: WHO Grade Classification**

- Code WHO grade as documented in health record
  - If WHO grade is not documented see Table 56.3 in AJCC 7<sup>th</sup> Ed. (page 596) for specific histologies with assigned WHO grade
  - *Examples:*
    - Anaplastic astocytoma – grade III
    - Glioblastoma – grade IV
    - Meningioma – grade I

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**SSF1: WHO Grade Classification**

- Grade I: Code 010
  - Slow-growing, nonmalignant
- Grade II: Code 020
  - Slow-growing; can be nonmalignant or malignant
- Grade III: Code 030
  - Malignant
- Grade IV: Code 040
  - Very aggressive malignant tumors



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**SSF2: Ki-67/MIB-1 Labeling Index (LI)**

- Ki-67 is a nuclear protein
- Labeling index (LI)
  - Record percentage of carcinoma cells in the tissue sample with positive IHC staining for Ki-67 protein
  - Staining may be done with MIB-1 monoclonal antibody
  - May correlate with patient’s clinical course



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**SSF3: Functional Neurologic Status - Karnofsky Performance Scale (KPS)**

- 0: Dead
- 10: Moribund
- 20: Very sick
- 30: Severely disabled
- 40: Disabled
- 50: Requires considerable assistance
- 60: Requires occasional assistance
- 70: Cares for self but unable to carry on normal activity
- 80: Normal activity with effort
- 90: Normal activity with minor signs disease
- 100: Normal with no evidence of disease



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**SSF3: Functional Neurologic Status - Karnofsky Performance Scale (KPS)**

- Record the KPS as documented by physician in patient's record
- Do NOT infer KPS from information in record
- Used to compare treatment effectiveness and to assess prognosis



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**SSF4: Methylation of O6-Methylguanine-Methyltransferase (MGMT)**

- MGMT is DNA repair enzyme
- Methylation shuts down DNA repair
- Increased methylation may allow specific drugs to be effective on CNS tumors



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**SSF5 & SSF6: Loss of Heterozygosity (LOH)**

- LOH
  - Chromosome damage that results in failure of tumor suppression
- SSF5
  - Record results of test for LOH in chromosome 1p
- SSF6
  - Record results of test for LOH in chromosome 19q
- Tests may be performed at same time and on single report



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**SSF7: Surgical Resection**

- Code extent of surgical resection as described in operative and pathology reports
  - Correlated to outcome
  - May be determinant in treatment



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**SSF8: Unifocal vs. Multifocal Tumor**

- Record whether tumor is solitary or multifocal at time of diagnosis
  - Multifocal tumors have a worse prognosis
  - Affect treatment decisions



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**SSF Requirements by Standard Setters**

- Commission on Cancer and NCI/SEER
  - Brain & CNSOther: SSF1, SSF4, SSF5, SSF6
  - IntracranialGland: SSF1
- CDC/NPCR
  - Brain, CNSOther, IntracranialGland: SSF1



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**SSF Requirements by Standard Setters**

- Canadian Council of Cancer Registries
  - Brain & CNSOther:
    - SSF1
    - SSF2, SSF5, SSF7, SSF8 if info is available in path report
    - SSF3, SSF4, SSF6 if info is available in clinical chart
  - IntracranialGland
    - SSF1
    - SSF2 if info is available in path report



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**Questions?**

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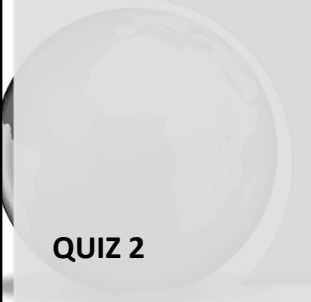
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**QUIZ 2**



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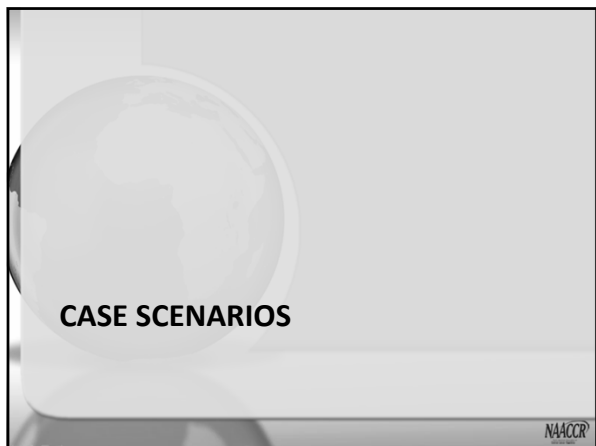
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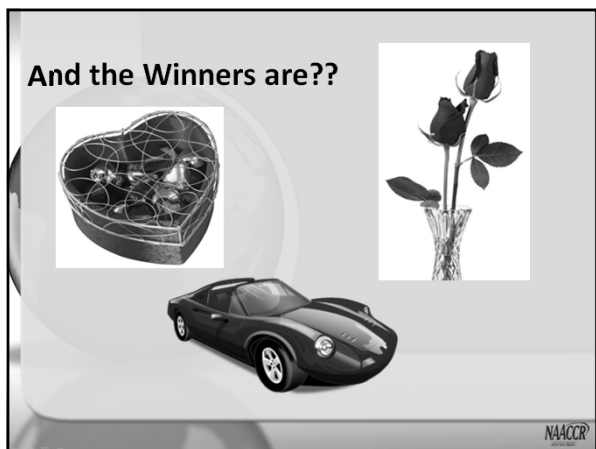
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**Coming up!**

- 3/7/13
  - Abstracting & Coding Boot Camp: Case Scenarios
- 4/4/13
  - Collecting Cancer Data: Breast
- **Certificate phrase: Brain Cancer**
  - <http://www.survevgizmo.com/s3/1125707/CNS-Certificate>



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**THANK YOU!**



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