

••• Q&A

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

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••• Agenda

- Overview
- Epi Moment
- MP/H
 - Benign
 - Malignant
- Stage

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••• Case eligibility

- Includes malignant & non-malignant tumors diagnosed on or after 1/1/2004 of the following sites:
 - Meninges (C70._)
 - Brain (C71._)
 - Spinal cord, cranial nerves, & other CNS (C72._)
 - Pituitary gland (C75.1)
 - Craniopharyngeal duct (C75.2)
 - Pineal gland (C75.3)

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••• Juvenile Astrocytoma

- Record Juvenile Astrocytoma as 9421/3 in the registry.
 - ICD O 3 Manual lists Juvenile Astrocytoma as 9421/1
 - 9421/3 should also be used for pilocytic astrocytoma

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••• Sequence 00 vs 60

- 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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••• Laterality

CNS sites defined as paired for cases diagnosed 1/1/2004 and after

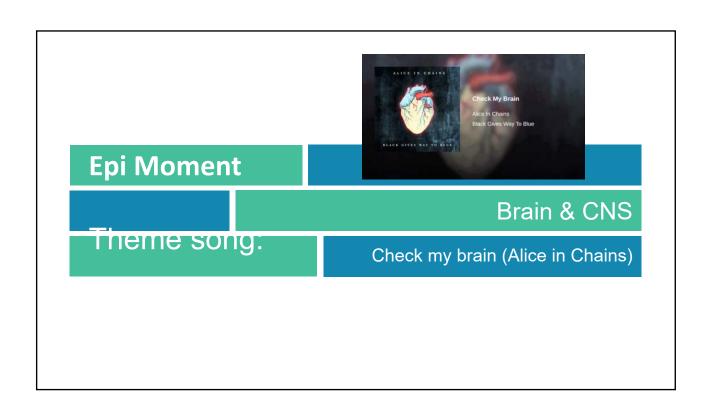
- Cerebral meninges C70.0
- Cerebrum C71.0
- Frontal lobe C71.1
- Temporal lobe C71.2
- Parietal lobe C71.3
- Occipital lobe C71.4

Assign laterality as '0' for all other CNS sites

- Olfactory nerve C72.2
- Optic nerve C72.3
- Acoustic nerve C72.4
- Cranial nerve, NOS C72.5

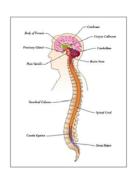


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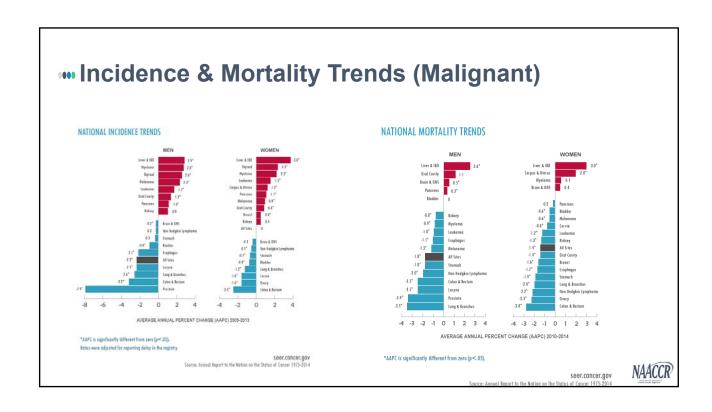


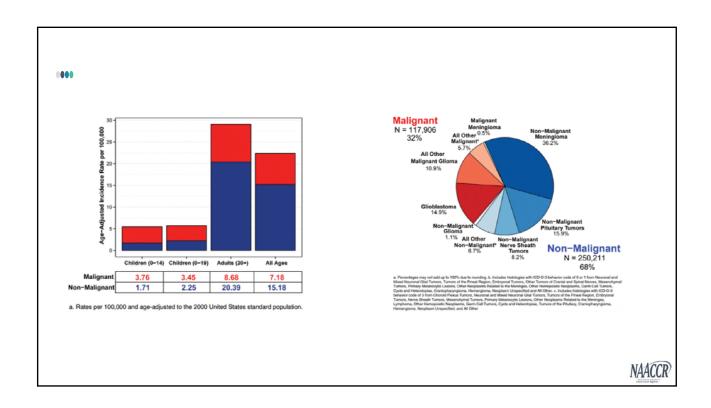
Epidemiology: Brain & CNS Tumors

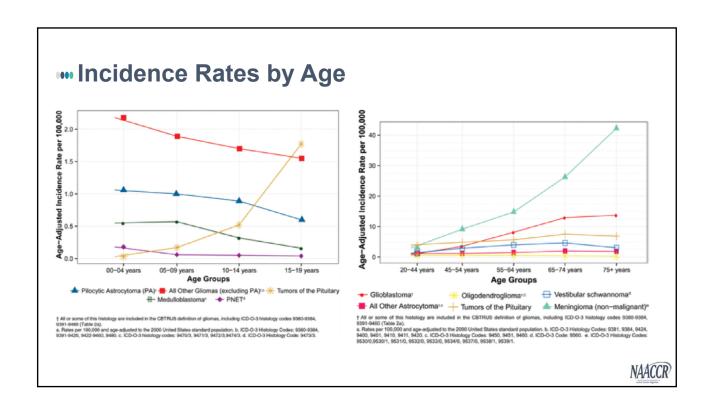
- Non-malignant rates higher (11.0 per 100,000 versus 6.6)
 - Rates higher in women (13.8 per 100,000 versus 7.9)
- Malignant rates higher in developed countries
 - Rates higher in men(7.8 per 100,000 versus 5.6)
- Common site among children
 - #1 0-14
- Survival varies significantly by age, behavior, & histology
 - Pediatric survival a success story
 - 0-19 73% 5-year survival; 20-44 59%; 45-54 31%; 55-64 18%, 65-74 11%; 75+ 6% (malignant)
 - Non-malignant survival higher in US than Europe
 - 96% US 69-77 % Europe (adults)
 - Glioblastoma lowest survival rates
 - 4-17% 5-year survival dependent upon age

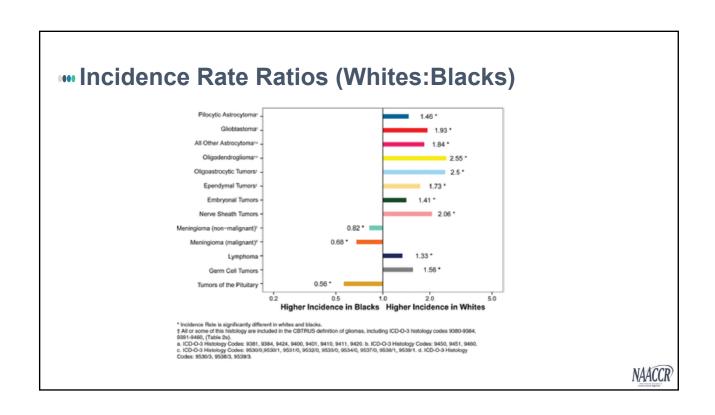


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... Brain & CNS Survival

- Largely dependent upon age
 - decreases with older age at diagnosis
 - children and young adults generally have better survival outcomes for most histologies.
- Malignant
 - 5-year survival 34.9%
 - Large variations in survival estimates depending upon histology
 - 94.2% for pilocytic astrocytoma but 5.5% for glioblastoma.
- Non-malignant
 - 5-year survival 90.4%
 - Less variation in survival estimates
 - · Lowest in craniopharyngioma and meningioma; 5-year relative survival of 83.9% and 86.4%, respectively.
 - Highest in nerve sheath tumors and tumors of the pituitary, 5-relative survival of 99.3% and 96.4%, respectively.
 - Highest in adolescents and young adults is highest (97.7%) versus children (96.4%) and older adults (88.8%).



••• Risk Factors

- Established risks
 - Radiation exposure (Radiation therapy)
 - Genetic disorders: Neurofibromatosis type 1 & 2, tuberous sclerosis, Von Hippel-Lindau disease, Li-Fraumeni
- Suspected risks
 - Cell phone use (radiofrequency rays not ionizing radiation)
 - Occupation exposures (vinyl chloride, petroleum products)
- Popular myths
 - Sugar substitutes (aspartame)
 - EMF
 - Some viruses





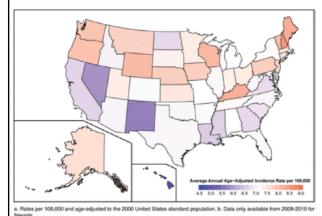
··· CiNA Research

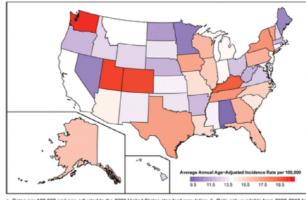
- CBTRUS
 - Descriptive epidemiology
 - Documented under reporting
- Annual Report to the Nation 2010
- Pediatric
 - Appalachian versus non-Appalachian
 - Potential genetic component
 - Pilot project to develop hypotheses about why risk in Appalachian children is higher





Wariability of Brain & CNS Tumors by State



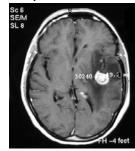


 a. Rates per 100,000 and age-adjusted to the 2000 United States standard population.
 b. Data only available from 2009-2010 for Nevada.

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... Underreporting

- Non-malignant tumors historically have high degree of interregistry variability in rates
- Does the variability have public health importance or is it spurious?
- · Non-malignant variability largely driven by case completeness
- Associated with underreporting
 - Non-microscopically confirmed, non-surgery
 - Younger age, Specific subsites

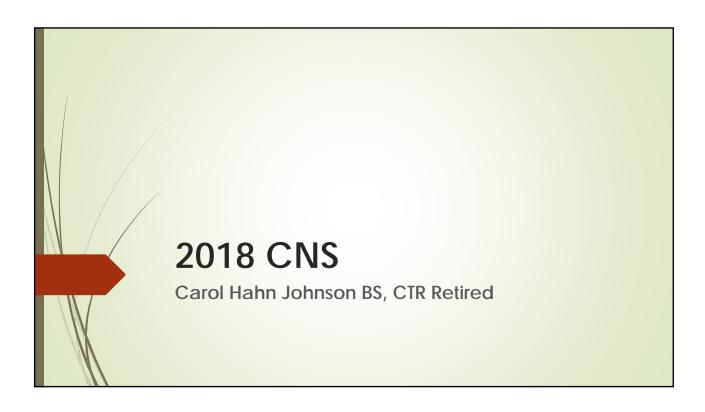


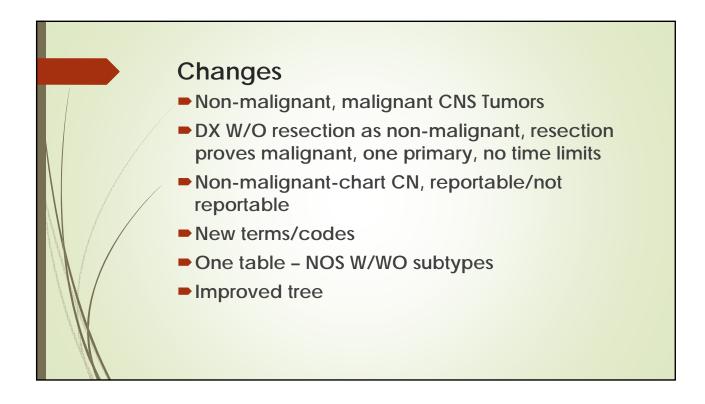
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--- Survey conducted in 2015

- Objectives
 - Assess benign/borderline brain tumors variability by registry
 - Survey high and low incidence registries
 - · Mutable differences in case ascertainment
 - training, operations
 - Assess correlations with rates
 - · Registry capacity, reporting facilities capacity, demographics, geography
- Conclusions
 - Active case finding
 - Linkage, use electronic sources
 - Site Specific, code specific, patient discharge
 - Non-Hospital sources
 - Radiology
 - Active radiology case finding/hospital discharge
 - Site specific
 - AIM software

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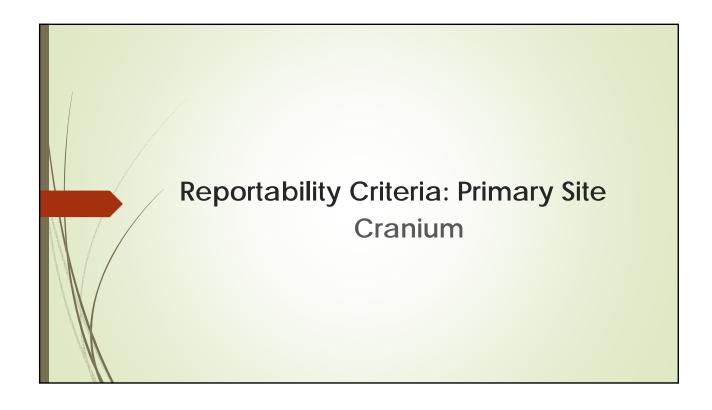




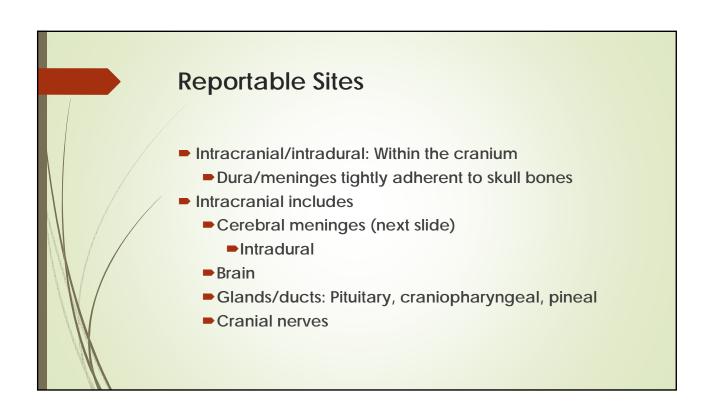
MPH 2007 Benign and Borderline Intracranial and CNS Carol Hahn Johnson BS, CTR (Retired)

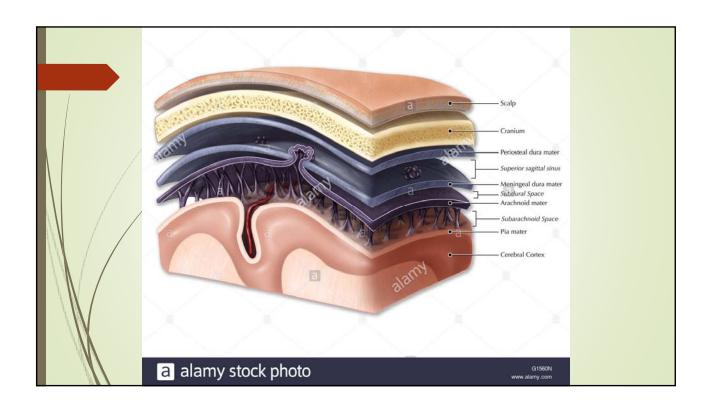
General "Rules" and Information M1 Single tumor ALWAYS single primary No timing rules for CNS neoplasms Laterality not used to determine multiple primaries Multiple cerebral meningiomas – single primary

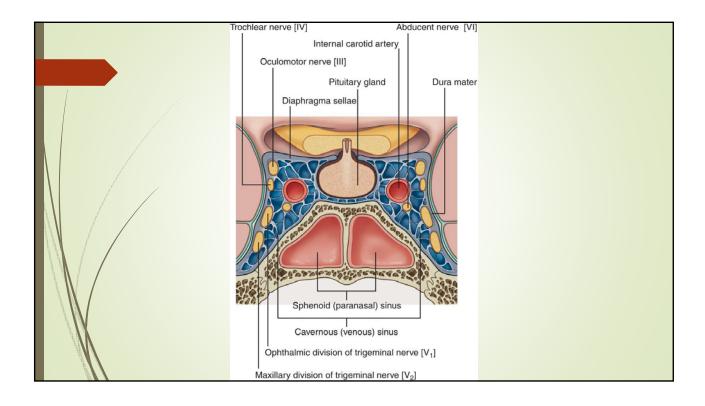




Ouestion How many primaries? Patient has Neurofibromatosis type II and presents with Schwannoma on left facial nerve Schwannoma on right acoustic nerve Multiple meningiomas falx cerebelli





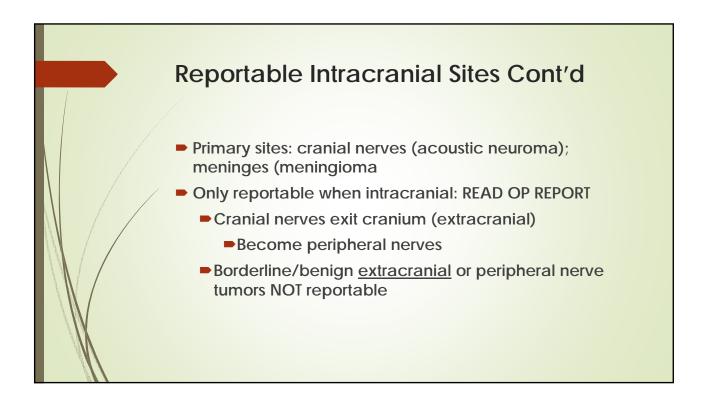


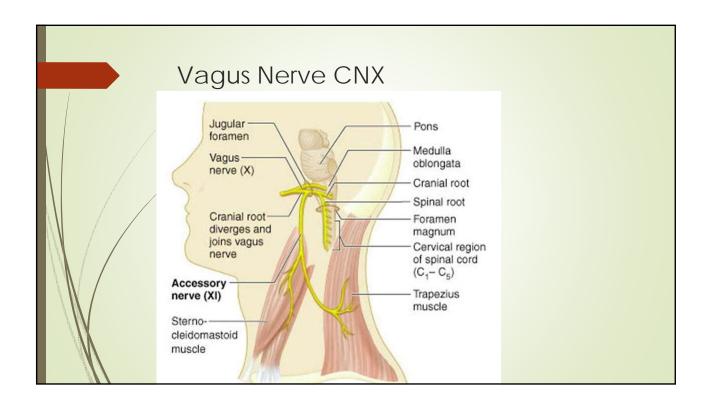
Intradural Sites (Between Layers of Meninges)

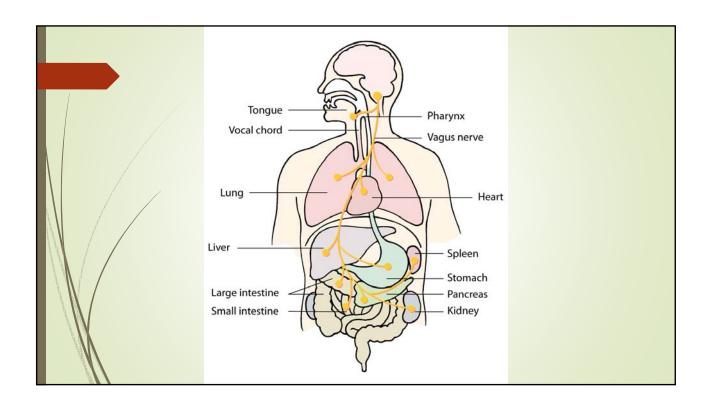
- Sagittal sinus
- Cavernous sinus
 - No ICD-O site/topography code
 - Tumors originate in
 - Cranial nerves passing through sinus
 - Dura/meninges of cranial nerves
 - When histology not compatible with nerve or meningeal tumor, may be extension from another site

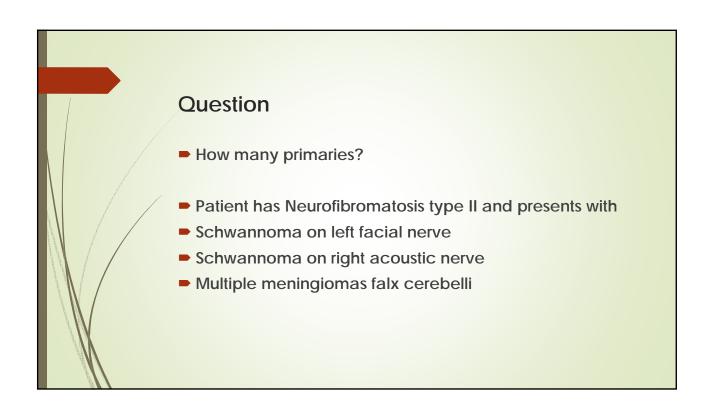
Reportable Cranial Meninges Tumors

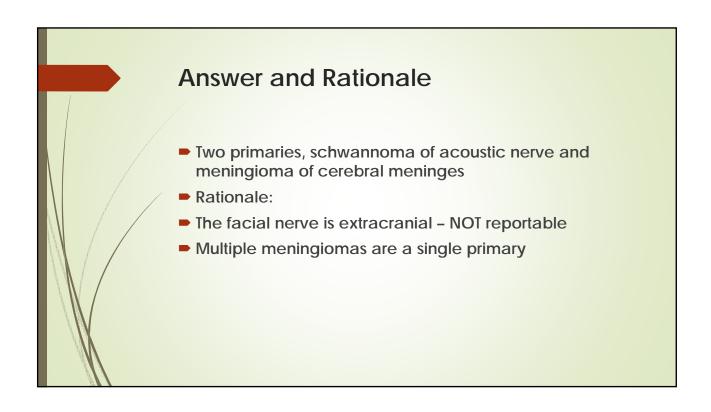
- Sphenoid wing meningioma
 - Originates in cranial meninges overlaying sphenoid wing
- Intraosseous meningioma
 - Originates inner layer of meninges
 - Grows "outward" toward bone forming periosteum

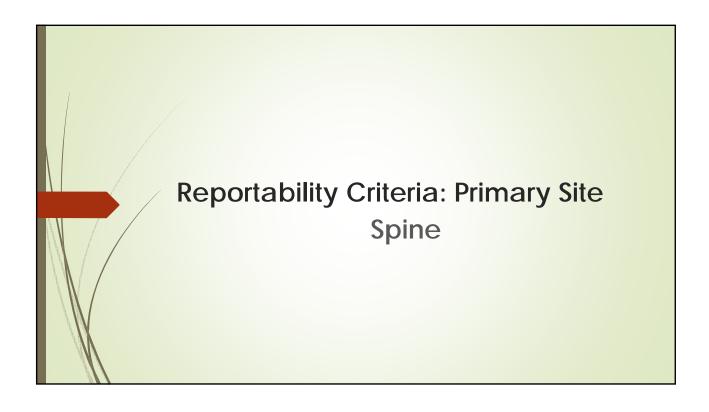


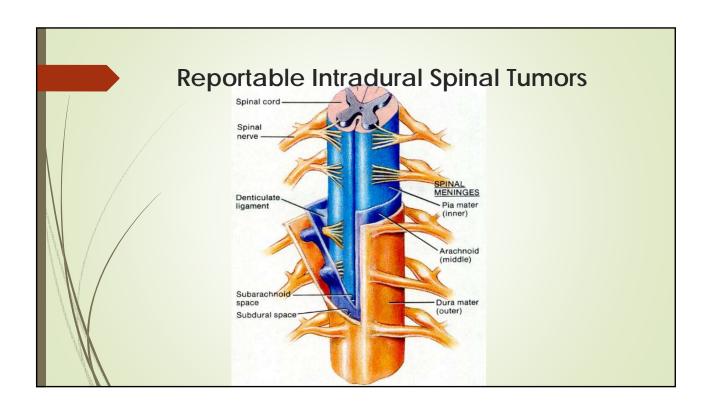


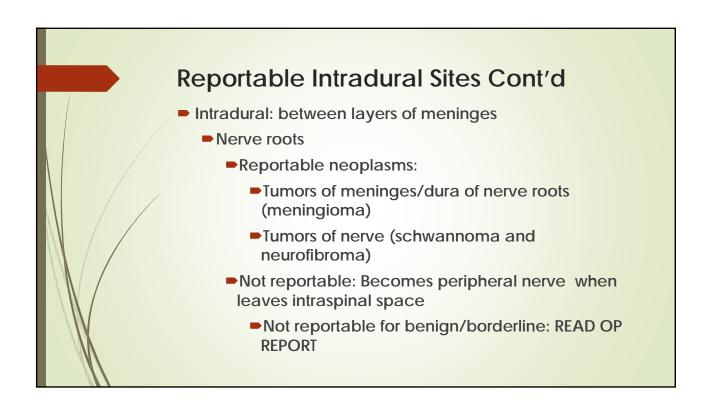


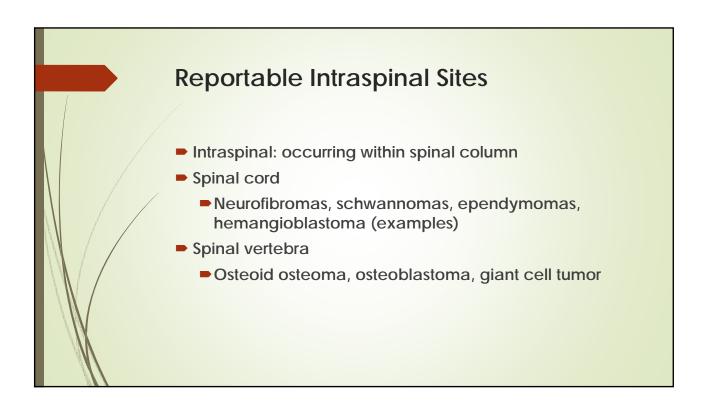


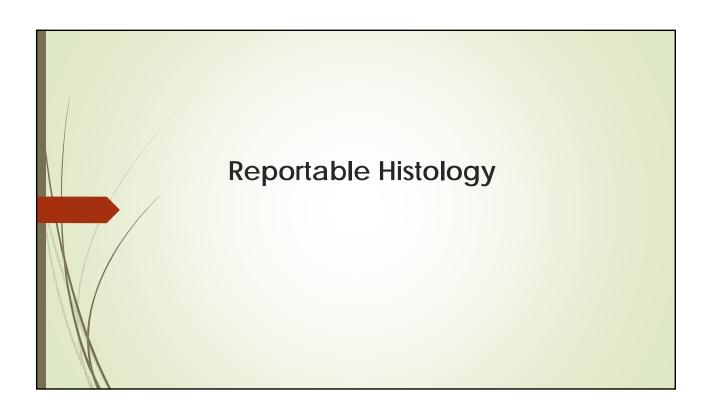




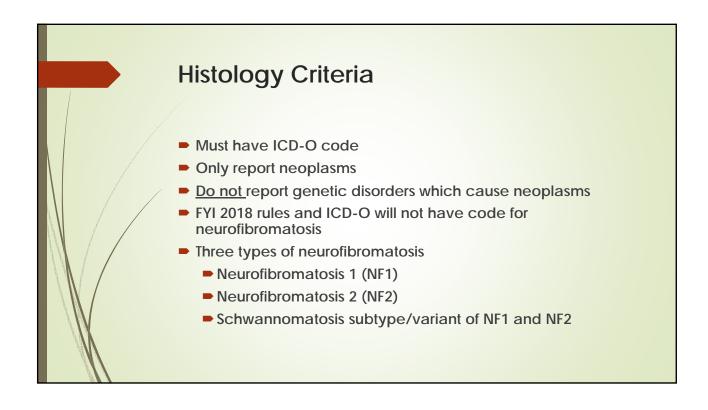


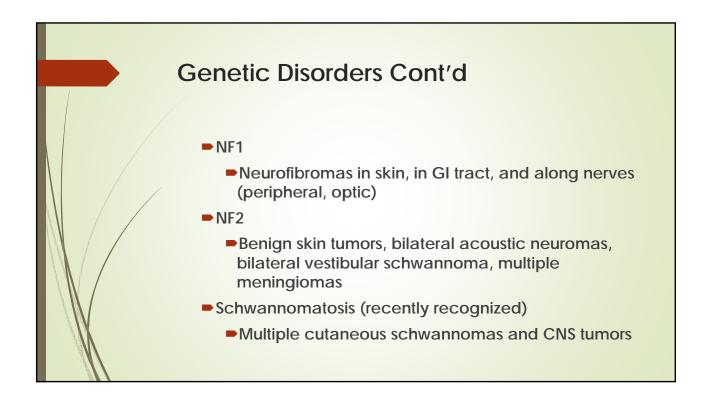




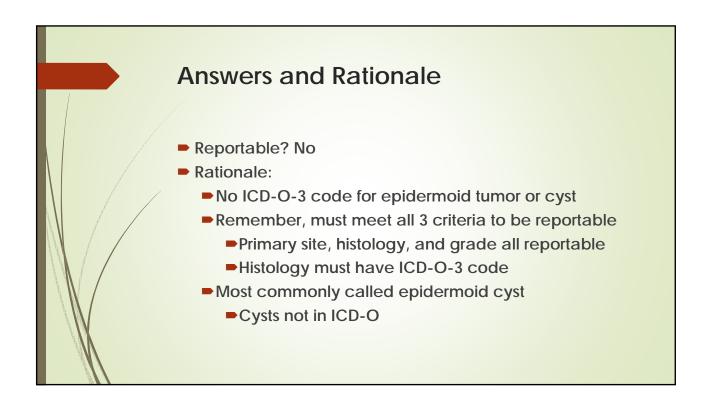


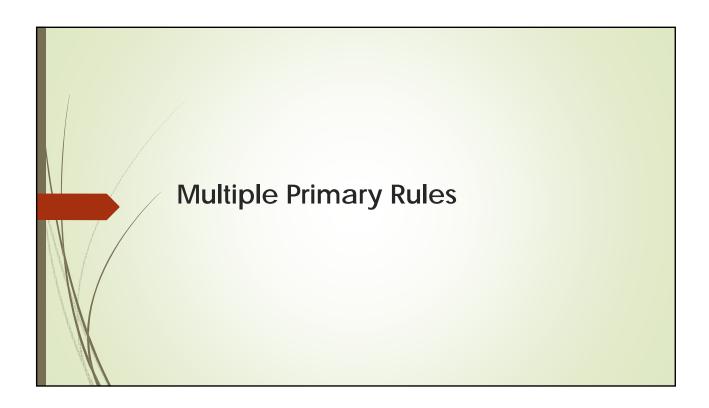
Pop Quiz Epidermoid tumor of the cerebellopontine angle (CPA) and trigeminal vesicle nerve. Patient presented to hospital ED and had brain MRI that revealed 3.2 cm space occupying lesion in region of the left CPA and trigeminal vesicle nerve compatible with epidermoid tumor. Reportable Y/N Histology code (if reportable) __ _ _ _ _





Pop Quiz • Epidermoid tumor of the cerebellopontine angle (CPA) and trigeminal vesicle nerve. Patient presented to hospital ED and had brain MRI that revealed 3.2 cm space occupying lesion in region of the left CPA and trigeminal vesicle nerve compatible with epidermoid tumor. • Reportable Y/N • Histology code (if reportable) _______







- M6 Chordoid plexus papilloma 9390/0 subsequent atypical chordoid plexus papilloma 9390/1
- M7 Neurofibroma 9540/0 subsequent neurofibromatosis, NOS 9540/1
 - Neurofibromatosis code will become obsolete
- Do not change original abstract
- COC registrars record recurrence
- Counting new primary inflates incidence



MALIGNANT BRAIN SPINAL CORD

CAROL HAHN JOHNSON, CTR RETIRED

CONTENTS

- Common problem
- Equivalent Terms and Definitions: Histology tree
- MP Rules
- Histology Rules

COMMON PROBLEM

DISCREPANT INFORMATION ON PATHOLOGY REPORT(S)

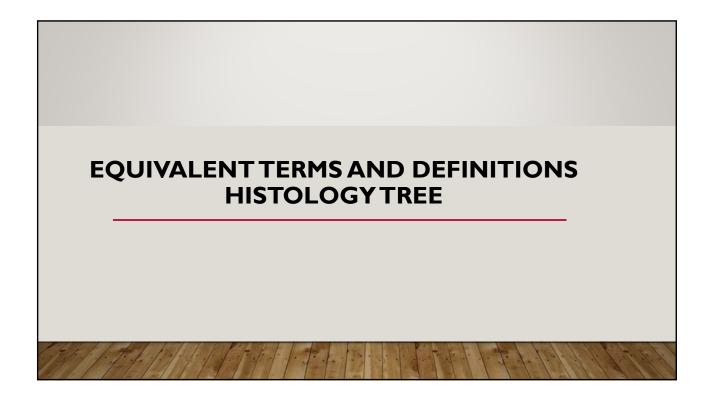
- Classification CNS tumors subjective
 - Grade
 - Histology
- No objective criteria for pathologists

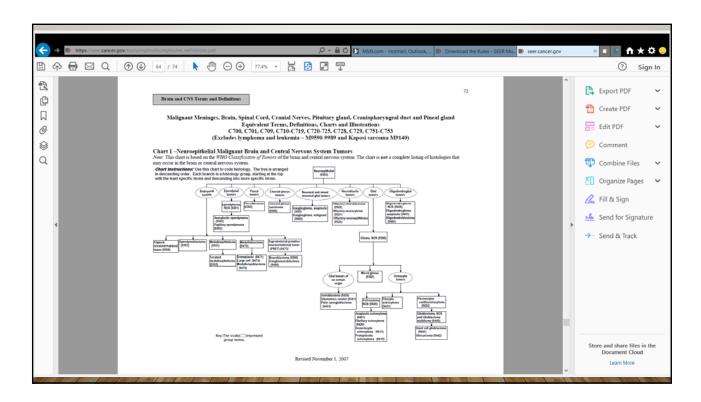
PATHOLOGY MAY BE DISCREPANT WHEN

- Single pathology report
 - · Slides sent for outside review
 - Multiple pathologists within institution disagree
- Multiple pathology report
 - One from biopsy; one from resection
 - First DX is clinical or radiography (no path) AND
 - First course RX watchful waiting AND
 - Subsequent resection shows different histology and/or grade

RESOLUTION

- I. When possible, get advice from pathologist or attending
- 2. When option one is not available, code from most dependable source
 - a. Resection pathology
 - b. Biopsy pathology
 - c. Physician's documentation of grade and histology
 - d. MRI
 - e. CT
 - f. PET
 - g. Angiogram

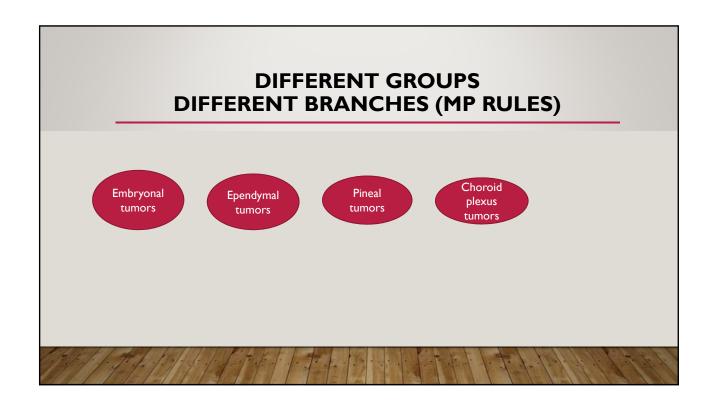


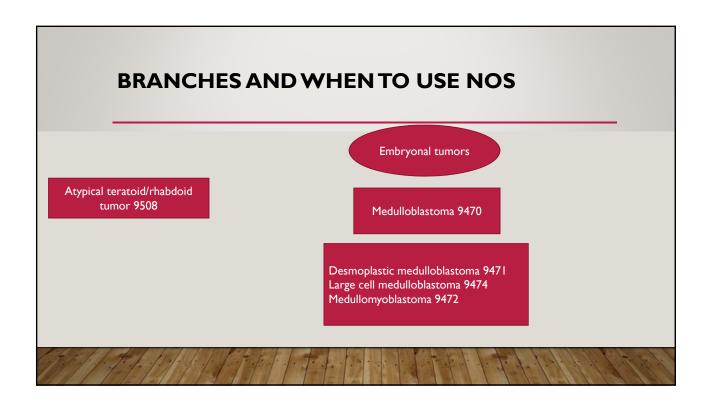


SHAPES HAVE MEANINGS

- Ovals
 - "Group" names
 - No ICD-O code
 - Used on scans and clinical diagnosis
 - Seldom on FNA
- Rectangles
 - NOS
 - Subtypes/variants of NOS

PIRST PART OF TREE - ERROR Neuroepithelial (9503) Neuroepithelial cells are the "stem cells" of the nervous system





MULTIPLE PRIMARY RULES

POP QUIZ

- MRI: Multicentric disease
 - RT frontal lobe 4.5 M
 - RT parietal lobe 3.6 CM
 - RT frontal lobe two tumors; 2.4 CM and 3.1 CM
- Biopsy: Glioblastoma
- How many primaries?

POP QUIZ

- 5/1/2005 MRI SX glioma brain stem no resection
- 4/14/2010 Partial resection same tumor
 - Pathology: astrocytoma
- Single or multiple primaries?
- If single, is the original histology of glioma correct?

M6

 A glioblastoma or glioblastoma multiforme (9440) following a glial tumor is a single primary* (See Chart I)

M7

 Tumors with ICD-O-3 histology codes on the same branch in Chart I or Chart 2 are a single primary.*

Note: Recurrence, progression, or any reappearance of histologies on the same branch in Chart I or Chart 2 is always the same disease process.

Physicians will stage both tumors

M8

 Tumors with ICD-O-3 histology codes on different branches in Chart I or Chart 2 are multiple primaries

M9

- Tumors with ICD-O-3 **histology** codes that are **different** at the first $(\underline{\mathbf{x}}\mathbf{x}\mathbf{x}\mathbf{x})$, second $(\mathbf{x}\underline{\mathbf{x}}\mathbf{x}\mathbf{x})$ or third $(\mathbf{x}\mathbf{x}\underline{\mathbf{x}}\mathbf{x})$ number are multiple primaries. **
- · Remember rules are hierarchical
- Cannot rely on rule
- ONLY use when rules MI-M8 do not apply

POP QUIZ

- Biopsy: Glioblastoma
- MRI: Multicentric disease
 - RT frontal lobe 4.5 M
 - RT parietal lobe 3.6 CM
 - RT frontal lobe two tumors; 2.4 CM and 3.1 CM
- How many primaries?

ANSWER AND RATIONALE

- Single primary
- RATIONALE:
- Rule M7 Tumors with ICD-O-3 histology codes on the same branch in Chart 1 or Chart 2 are a single primary OR
- MIO Tumors that do not meet any of the above criteria are a single primary
- ONLY "SITE" RULE IS M5 topography codes differ at second CXxx or third CxXx character (sites other than brain)
- Glioblastomas spread along the dendrites and nerves which results in multifocal/multicentric tumors

POP QUIZ

- 5/1/2005 MRI SX glioma brain stem no resection
- 4/14/2010 Partial resection same tumor
 - Pathology: astrocytoma
- Single or multiple primaries?
- If single, is the original histology of glioma correct?

ANSWER AND RATIONALE

- Single primary M2
 - A single tumor is a single primary
- · Pathology takes precedence over radiology diagnosis
- Changes to abstract
 - · Change histology from glioma to astrocytoma
 - Change diagnostic confirmation code (SEER and FORDS)
- · Note: astrocytoma is a subtype/variant of glioma
- REPORT CHANGES TO CENTRAL REGISTRY

HISTOLOGY RULES

POP QUIZ

- Pathology:WHO Grade IV astrocytoma with features consistent with secondary glioblastoma
- What histology code should be used?
 - Astrocytoma
 - Glioblastoma
 - Mixed glioma

H5 AND H10

- Code the specific type when the diagnosis includes a **non-specific** term and a **specific** term or type **on** the **same branch** in Chart 1 or Chart 2
- NOS and subtype/variant of same NOS

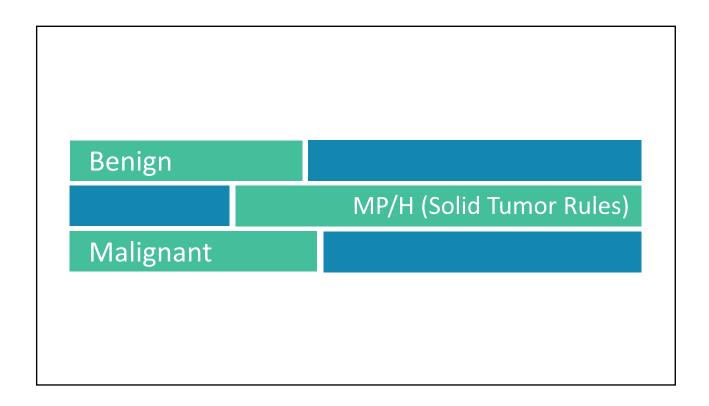
POP QUIZ

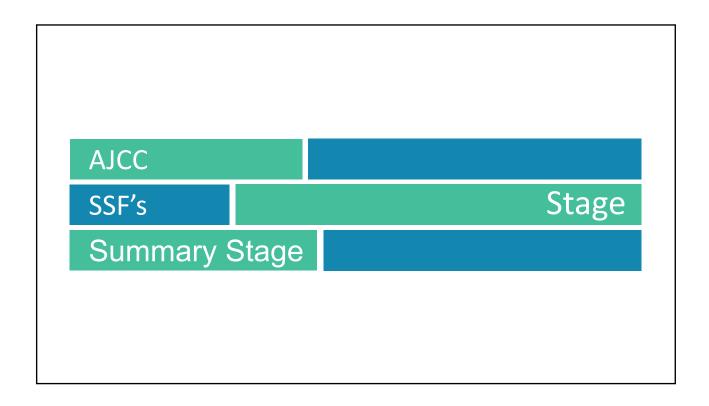
- Pathology:WHO Grade IV astrocytoma with features consistent with secondary glioblastoma
- What histology code should be used?
 - Astrocytoma
 - Glioblastoma
 - Mixed glioma

ANSWER AND RATIONALE

- Code glioblastoma
- · Glioblastoma is a subtype/variant of astrocytoma
- Use rule H5 and code the most specific histology, glioblastoma







••• AJCC Stage

- Chapter 56 page 593
 - No stage grouping
 - Excellent background information
 - Table 56.2 WHO classification of tumors of the central nervous system
 - Table 56.3 WHO grades of CNS Tumors
 - Brain Tumor Survival Data

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AJCC Stage

Clinical Stage
 cT 88 cN 88 cM 88 Stage 88

Pathologic Stage
 pT 88 pN 88 pM 88 Stage 88

Clinical Staged by 88

Pathologic Staged by 88

• TNM Edition 88

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••• Summary Stage 2000

- 1 Local
 - Confined to: one hemisphere in one part of brain (infra/supratentorial); meninges; invading/encroaching on ventricular system
- 5 Regional
 - crossing midline or tentorium invades bone, blood vessel, nerves, spinal cord
- 7 Distant
 - Circulating cells in CSF; extension to nasal cavity, nasopharynx, posterior pharynx; outside CNS
- 8 Benign
- Codes 0, 2, 3, 4 are not applicable https://seer.cancer.gov/tools/ssm/brain.pdf

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••• Pop Quiz

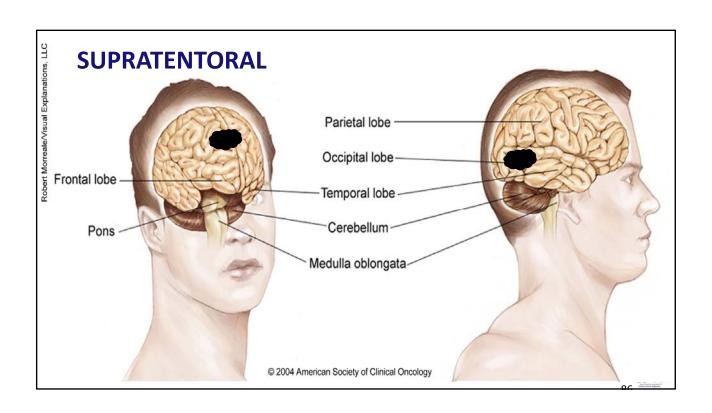
• On what page of the Summary Stage Manual is code 8 defined?

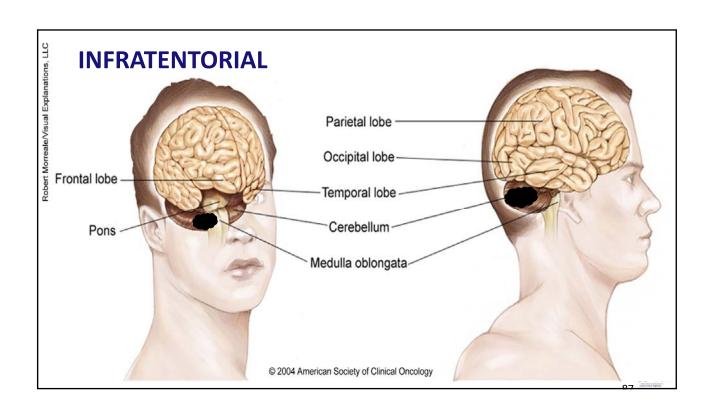
Code Definition

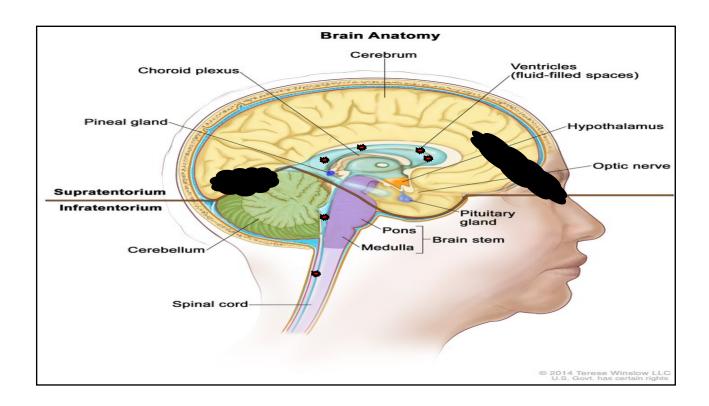
- 0 In situ
- 1 Localized only
- 2 Regional by direct extension only
- 3 Regional lymph nodes involved only
- 4 Regional by BOTH direct extension AND lymph node involvement
- 5 Regional, NOS (Not Otherwise Specified)
- 7 Distant site(s)/node(s) involved
- 9 Unknown if extension or metastasis (unstaged, unknown, or unspecified) Death certificate only case

Page 2 Summary Staging Manual 2000

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••• SSF1: WHO Grade Classification

- Histologic grading classification for CNS tumors by the WHO
- Important prognostic factor for response to treatment & outcomes for CNS tumors
- Not the same as ICD-O-3 grade/differentiation
- Coded in the 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 7th
 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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••• Grade/Differentiation

- Do not record the WHO Grade, Anne/Mayo, or Kernohan grades in the grade field
 - Record the WHO grade in Site Specific Factor 1
 - If no grade is given, code 9 (unknown)
- Anaplastic is synonymous with undifferentiated and should be assigned grade 4

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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
- This can typically be found in the path report as the testing will be completed on tumor tissue.



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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

 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

••• 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

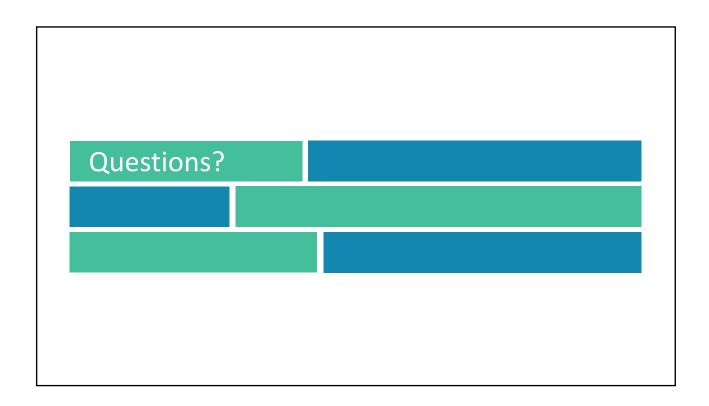
Typically listed on an addendum to a pathology report.



••• 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
- Typically listed on an addendum to a pathology report. Tests may be performed at same time and on single report

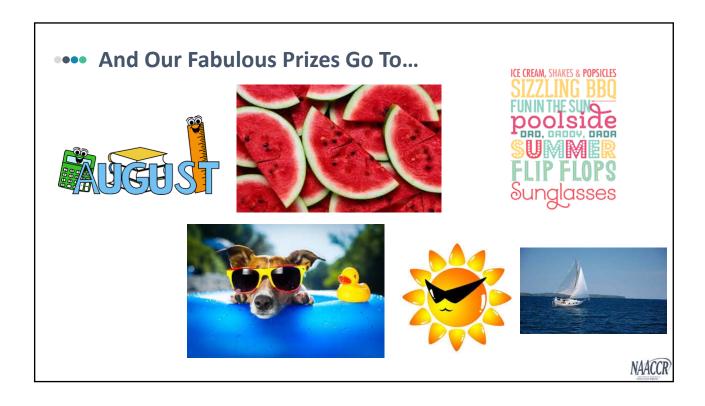
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••• Coming Up....

- 9/7/13 Coding Pitfalls
 - Special Guest: Steve Peace
- 10/5/17 Collecting Cancer Data: Prostate (new season)

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••• CE Certificate Quiz Survey

- Phrase
- Link
 - http://www.surveygizmo.com/s3/3744013/CNS-2017

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