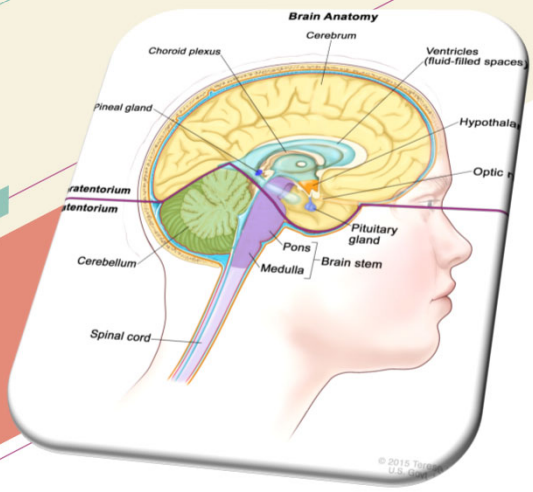

CNS 2022

June 2, 2022

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Q&A

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

If you have participants watching this webinar at your site, please collect their names and emails.

We will be distributing a Q&A document in about one week. This document will fully answer questions asked during the webinar and will contain any corrections that we may discover after the webinar.

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Fabulous Prizes

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Guest Presenter

- Angela Costantini, CTR
  - Senior CTR at Cincinnati Children's Hospital and Senior Clinical Data Specialist at Q-Centrix

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Agenda

- CNS and Epi Moment
- Anatomy and SSDI's
- Solid Tumor Rules
- Pediatric CNS
- Review of Case Scenarios

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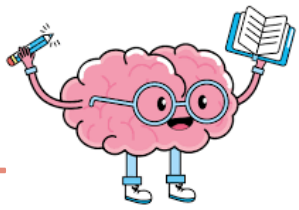
EPI MOMENT
Brain and CNS



Dr. Recinda Sherman

Theme Song:
If I only had a brain

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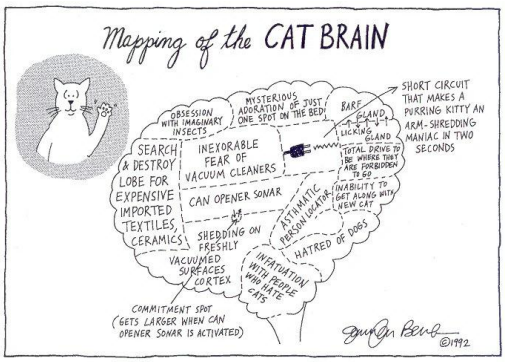
Brain & CNS

- Complex and large category
  - CBTRUS recode
  - Rare Cancers recode
  - Behavior recode
    - Coding changes over time
- Case ascertainment
  - Radiologic diagnosis
  - Malignant vs Non-Malignant
    - Non-malignant underutilized in statistics & research
    - Non-malignant approx 73% of total Brain & CNS
    - Completeness of reporting
    - Completeness metric for central cancer registries



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Anatomy and SSDIs

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Meninges C70.0 – C70.9

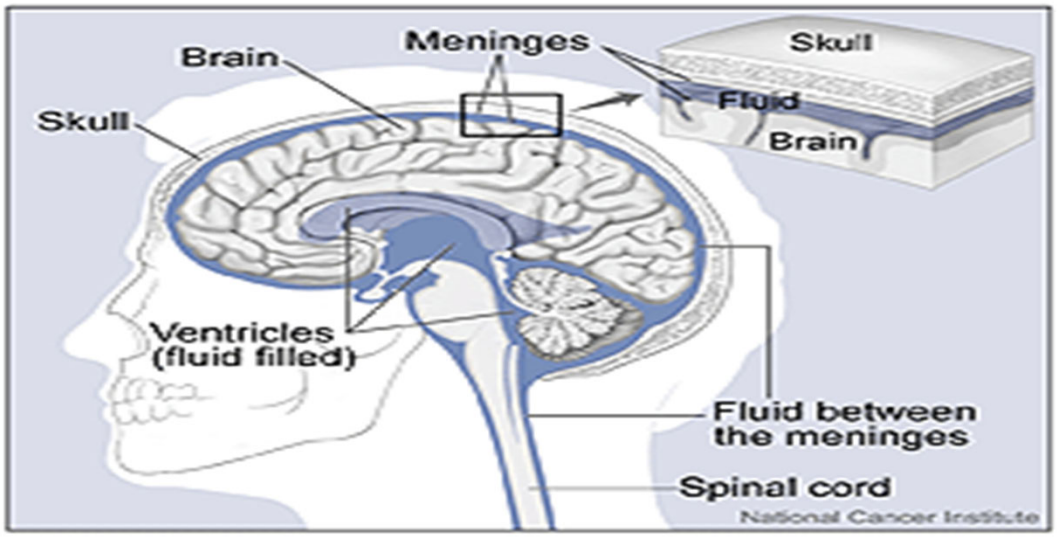

Image source: NCI VisualOnline; Artist – Alan Hoofring

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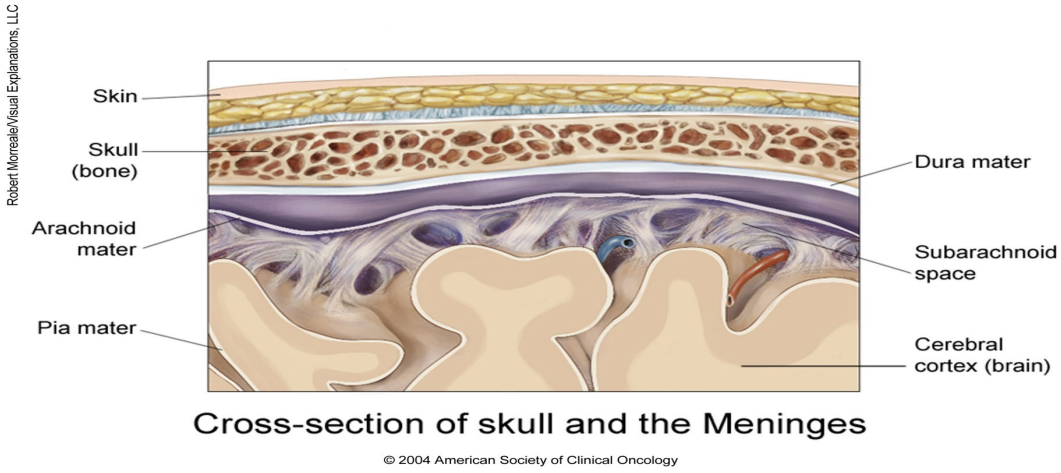
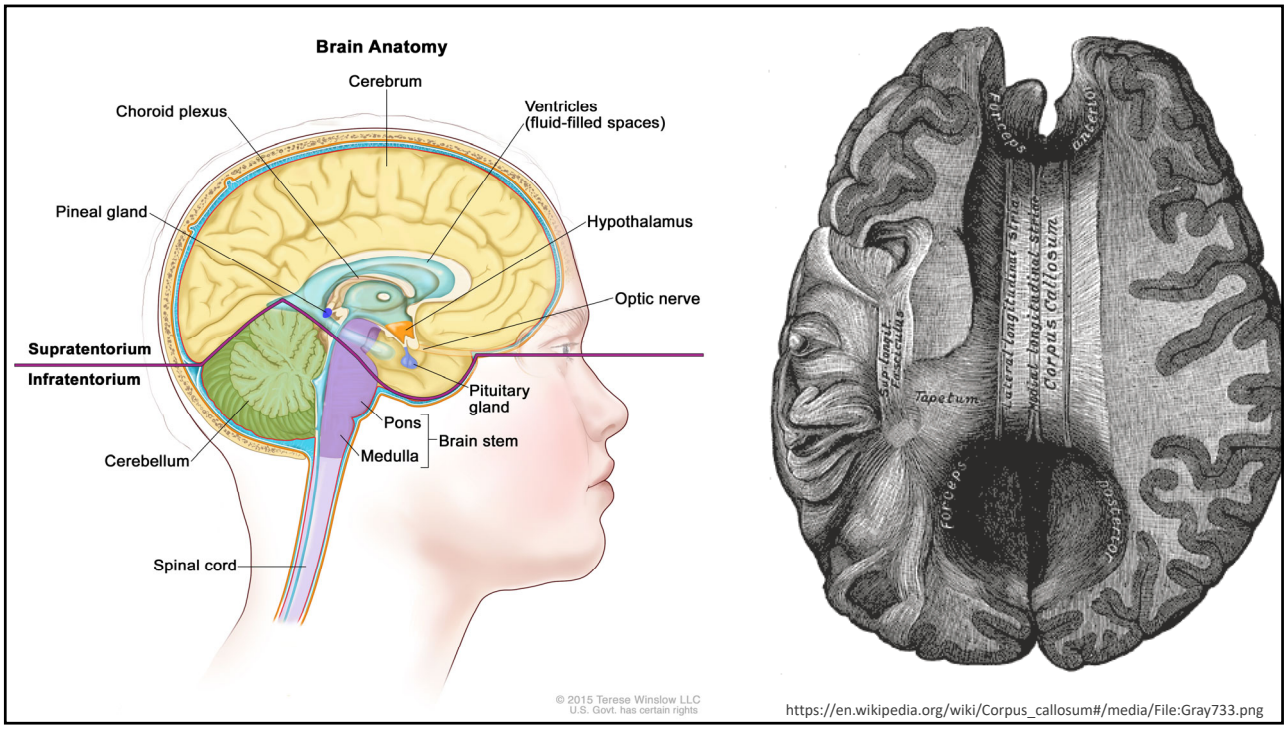
Cerebral Meninges

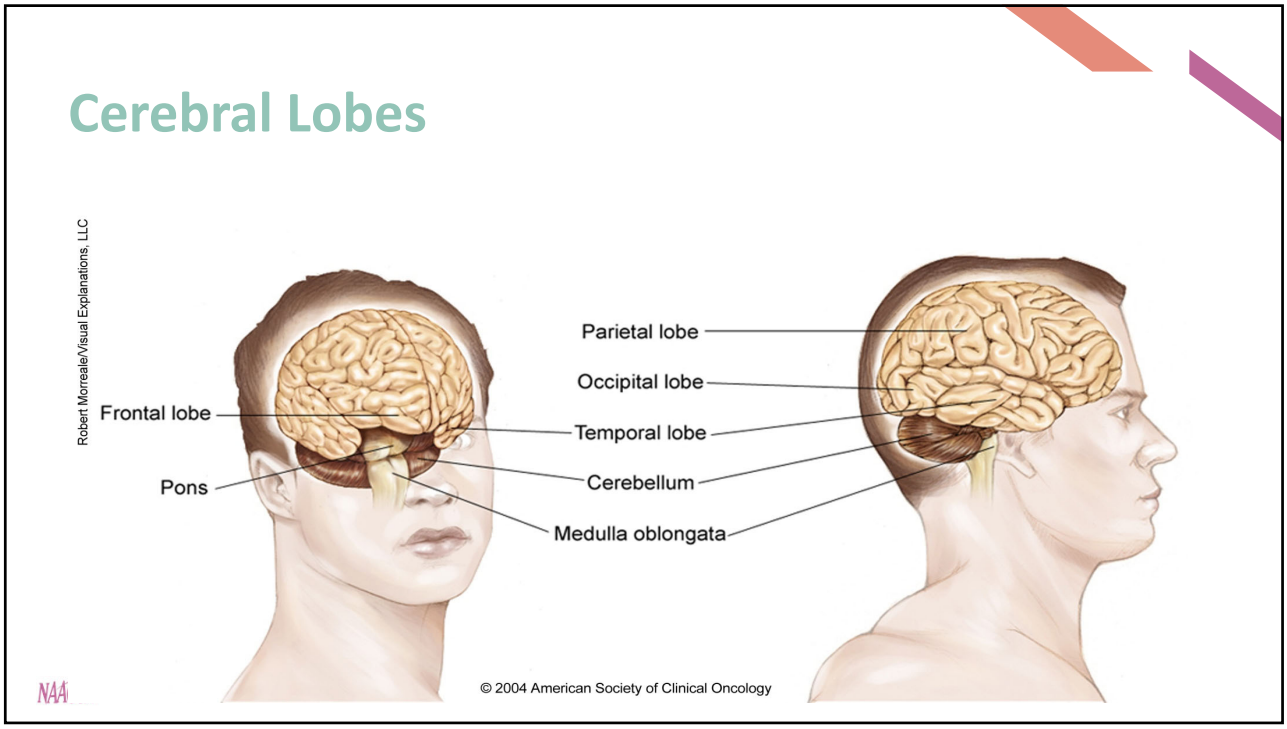


Cross-section of skull and the Meninges

10


Brain Anatomy
Cerebrum
Choroid plexus
Ventricles
(fluid-filled spaces)
Pineal gland
Hypothalamus
Optic nerve
Supratentorium
Infratentorium
Pituitary
gland
Pons
Brain stem
Medulla
Cerebellum
Spinal cord
© 2015 Terese Winslow LLC
U.S. Govt. has certain rights
Forceps anterior
Sup. longit. fasciculus
Tapetum
Corpus Callosum
Forceps posterior
https://en.wikipedia.org/wiki/Corpus_callosum#/media/File:Gray733.png

11


Cerebral Lobes
Robert Morreale/Visual Explanations, LLC
Frontal lobe
Pons
Parietal lobe
Occipital lobe
Temporal lobe
Cerebellum
Medulla oblongata
NAA
© 2004 American Society of Clinical Oncology

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

•Assign laterality as '0' for all other CNS sites

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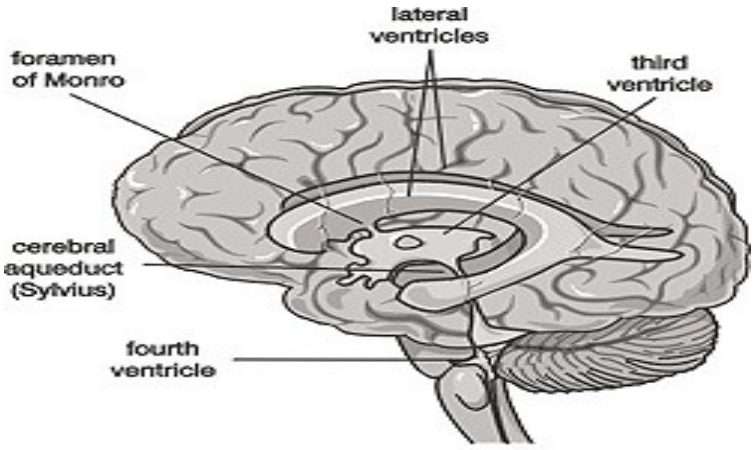
The Ventricular System



The Ventricles

Image source: CDC-NPCR's Data Collection of Primary Central Nervous System Tumors, 2004.

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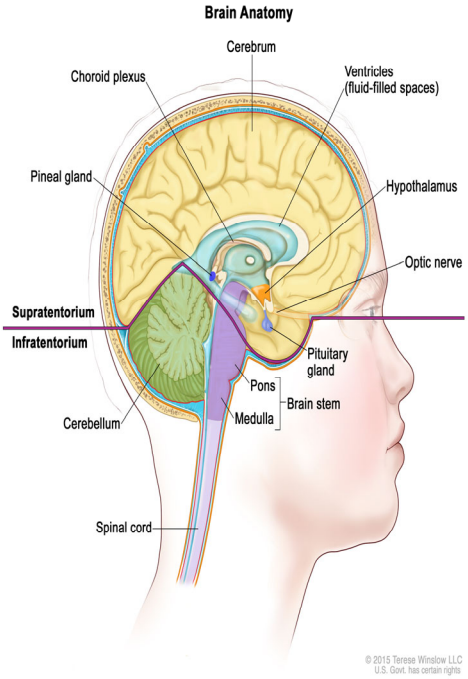
Brain Stem

- Pons: portion of brain stem superior to medulla oblongata
- Medulla oblongata: lower potion of brain stem
  - Olive: pair of oval structures in medulla oblongata
  - Pyramid: anterior or ventral portion of medulla oblongata
- Midbrain: mesencephalon; front of brain stem
  - Cerebral peduncle: ventral portion of midbrain



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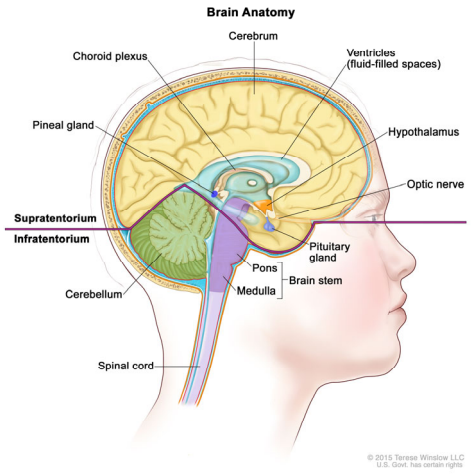
Summary Stage Localized



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SS2018	Description
1	Localized only (localized, NOS) › Confined to brain, NOS › Confined to meninges, NOS › Confined to ventricles › Tumor invades or encroaches upon ventricular system › Infratentorial tumor confined to › Brain stem or meninges of brain stem (one side) › Medulla oblongata › Midbrain (mesencephalon) › Pons › Cerebellum or meninges of cerebellum (one side or midline) › Lateral lobes › Median lobe of cerebellum › Vermis › Hypothalamus › Thalamus › Infratentorial tumor › Both cerebellum and brain stem involved with tumor on one side › Supratentorial tumor confined to › Cerebral hemisphere (cerebrum) or meninges of cerebral hemisphere (one side) › Frontal lobe › Occipital lobe › Parietal lobe › Temporal lobe

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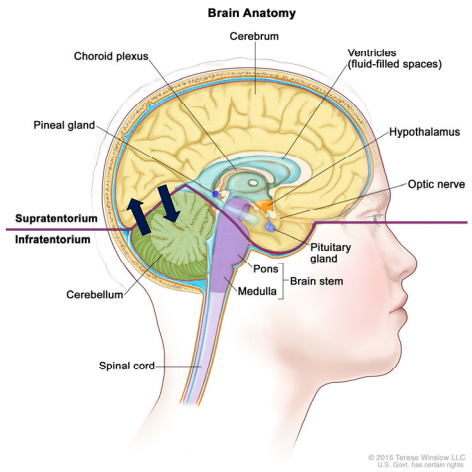
Summary Stage Regional



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Regional by direct extension only

- Bone (skull)
- Contralateral hemisphere
- Corpus callosum (including splenium)
- Major blood vessel(s)
- Meninges (e.g., dura)
- Nerves (cranial, NOS)
- Spinal cord/canal
- Supratentorial tumor extends infratentorially to involve
  - Brain stem
  - Cerebellum
  - Hypothalamus
  - Pallium
  - Posterior cranial fossa
  - Thalamus
- Infratentorial tumor extends supratentorially to involve
  - Anterior cranial fossa
  - Cerebrum (cerebral hemisphere) (excluding hypothalamus, pallium, thalamus)
  - Corpus callosum
  - Middle cranial fossa
  - Suprasellar brain
  - Tapetum
- Tumor crosses the midline

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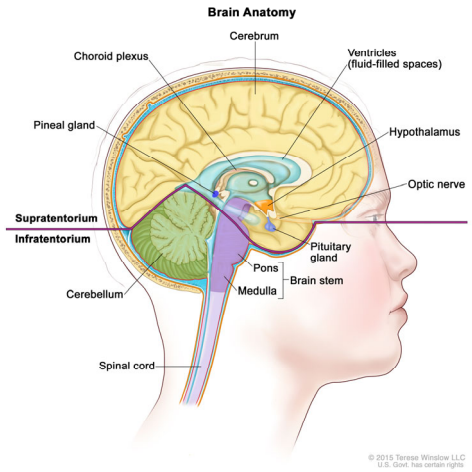
Summary Stage Distant



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Distant site(s)/lymph node(s) involved

- Distant site(s) (including further contiguous extension)
  - Circulating cells in cerebral spinal fluid (CSF)
  - Nasal cavity
  - Nasopharynx
  - Other direct extension outside CNS
  - Posterior pharynx
- Distant lymph node(s), NOS
- Distant metastasis, NOS
  - Carcinomatosis
  - Distant metastasis WITH or WITHOUT distant lymph node(s)
  - Metastasis within CNS and CSF pathways
    - "Drop" metastasis
    - Extra-neural metastasis
    - Metastasis outside the CNS

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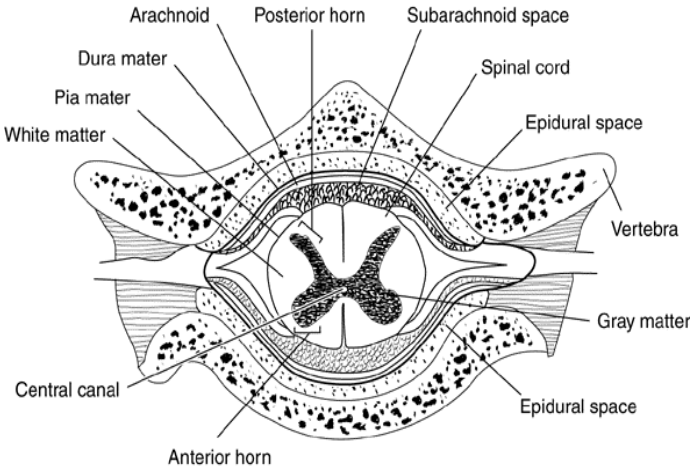
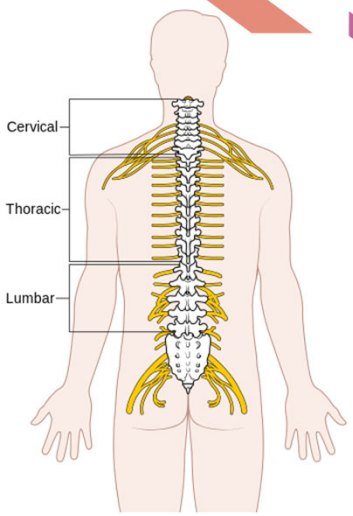
Spinal Cord





https://en.wikipedia.org/wiki/Spinal_cord#/media/File:Diagram_of_the_spinal_cord_CRUK_046.svg

Image source: CDC-NPCR's Data Collection of Primary Central Nervous System Tumors, 2004.

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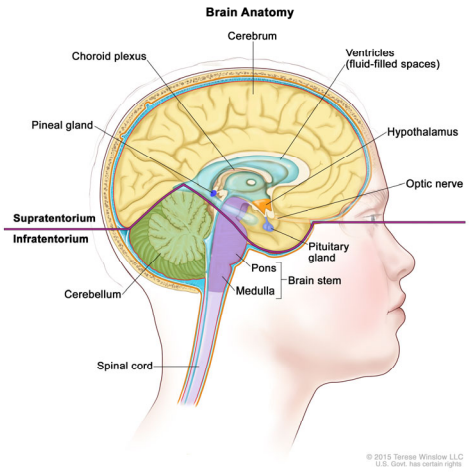
Intracranial Endocrine Glands and Related Structures



- Endocrine glands
  - Pituitary gland
  - Pineal gland
- Craniopharyngeal duct

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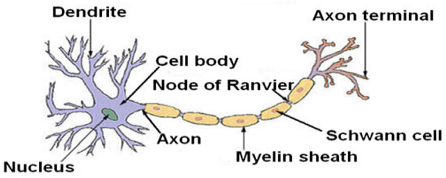
Neurons and Glial Cells

Neurons

- Neurons are the conducting cells of the nervous system.

Structure of a Typical Neuron



Glial Cells

- Do not conduct nerve impulses
- Support, nourish, and protect the neurons
- Glial cells are far more numerous than neurons and, unlike neurons, are capable of mitosis.

http://training.seer.cancer.gov/brain/tumors/anatomy/neurons.html

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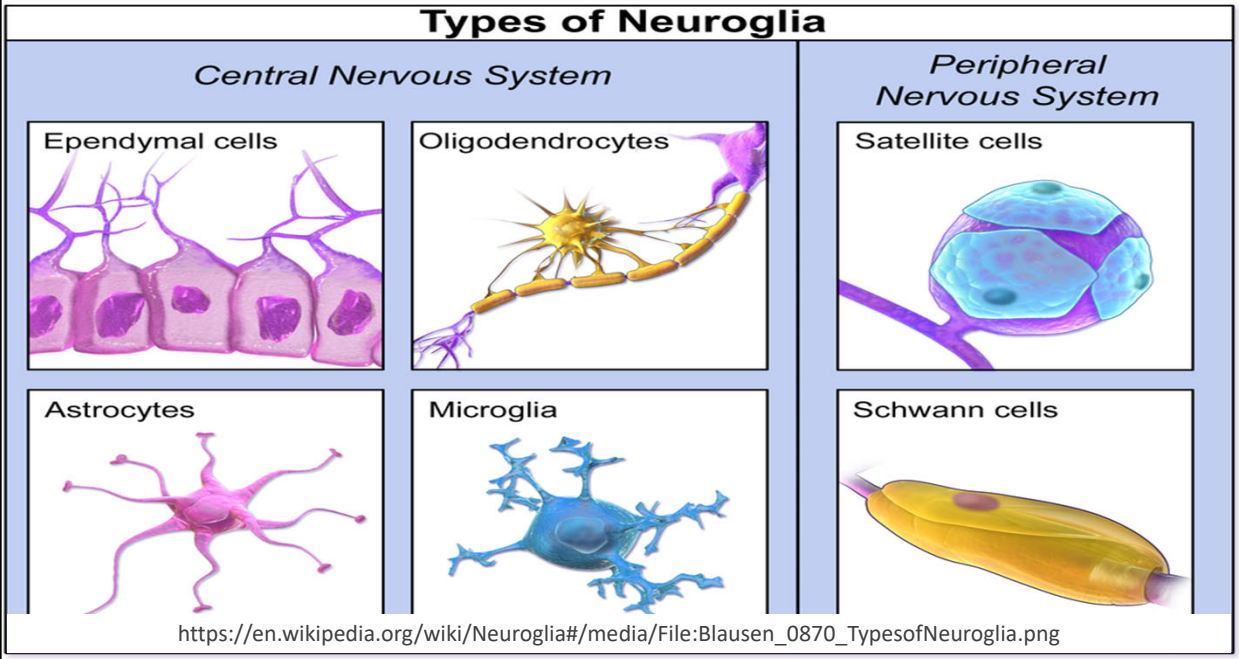

https://en.wikipedia.org/wiki/Neuroglia#/media/File:Blausen_0870_TypesofNeuroglia.png

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WHO Grade

- CNS tumor histologies are based on WHO grade as well as standard nomenclature.
- See Solid Tumor Rules Table 1
- See page 596 of the AJCC Staging manual

Code	Description
1	WHO Grade I : Circumscribed tumors of low proliferative potential associated with the possibility of cure following resection
2	WHO Grade II: Infiltrative tumors with low proliferative potential with increased risk of recurrence
3	WHO Grade III: Tumors with histologic evidence of malignancy, including nuclear atypia and mitotic activity, associated with an aggressive clinical course
4	WHO Grade IV: Tumors that are cytologically malignant, mitotically active, and associated with rapid clinical progression and potential for dissemination
L	Stated as "low grade" NOS

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Brain Molecular Markers

IDH-Mutant have a better prognosis

Code	Description
01	Diffuse astrocytoma, IDH-mutant (9400/3)
02	Diffuse astrocytoma, IDH-wildtype (9400/3)
03	Anaplastic astrocytoma, IDH-mutant (9401/3)
04	Anaplastic astrocytoma, IDH-wildtype (9401/3)
05	Glioblastoma, IDH-wildtype (9440/3)
06	Oligodendroglioma, IDH-mutant and 1 p/19q co-deleted (9450/3)
07	Anaplastic oligodendroglioma, IDH-mutant and 1p/19q co-deleted (9451/3)
08	Medulloblastoma, SHH-activated and TP53-wildtype (9471/3)
09	Embryonal tumor with multilayered rosettes, C19MC-altered (9478/3)
85	Not applicable: Histology not 9400/3, 9401/3, 9440/3, 9450/3, 9451/3, 9471/3, 9478/3

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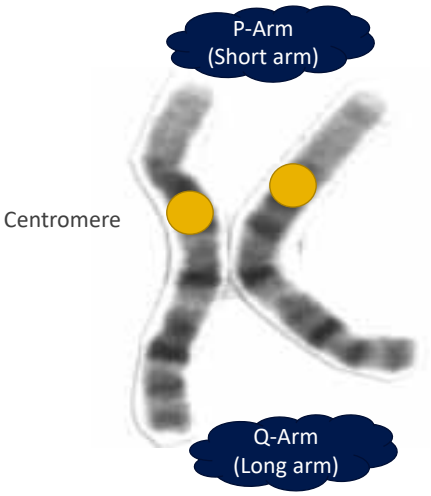
Chromosome 1p: Loss of Heterozygosity (LOH)

Code	Description
0	Chromosome 1p deletion/LOH not identified/not present
1	Chromosome 1p deletion/LOH identified/present
6	Benign or borderline tumor
7	Test ordered, results not in chart
8	Not applicable: Information not collected for this case (If this item is required by your standard setter, use of code 8 will result in an edit error.)
9	Not documented in medical record Cannot be determined by the pathologist Chromosome 1p deletion/LOH not assessed or unknown if assessed

```
RESULTS:          ABNORMAL MICROARRAY RESULT (See Tables below)

INTERPRETATION:
Microarray analysis was performed on this formalin-fixed, paraffin-embedded
(FFPE) primary brain tumor specimen. The following genomic imbalances were
noted:

(1) 483.5 kb 1p loss, including CDKN2C,
```



By National Human Genome Research Institute - Cropped from File:Human male karyotpe high resolution.jpg, Public Domain, https://commons.wikimedia.org/w/index.php?curid=41175399

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Solid Tumor Rules

Malignant CNS and Peripheral Nerves

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General information

- Separate chapters for malignant and non-malignant tumors
- WHO grade is not equivalent to tumor grade
- Equivalent terms
  - WHO Grade III and WHO Grade IV; malignant; invasive
  - WHO Grade I; non-malignant

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Clarification and Changes

Clarifications

- Intraosseous, cavernous sinus, and sphenoid wing meningiomas are reportable
- Multiple cerebral meningiomas are a single primary
  - Same histology or NOS and subtype/variant
- Multiple brain tumors (same histology) are a single primary
- Laterality and timing are NOT used to determine multiple primaries
- The brain (C710-C719) is a single primary site

Changes

- GBM after an astrocytic or glial tumor is now a multiple primary (M6)
- M4 is a new rule that clarifies that a single tumor is **always** a single primary, and the malignant behavior is reported
- Bilateral optic nerve gliomas/pilocytic astrocytomas are a single primary

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Priority Order for Using Documentation to Identify Histology (/3 only)

1. Pathology/tissue from resection of primary tumor
   A. Biomarkers
   B. The addendum(s) and/or comment(s)
   C. Final diagnosis/synoptic report
   D. CAP protocol
2. Pathology/tissue from biopsy of primary tumor
   Same A, B, C, D as #1.
3. Cytology (Usually CSF)
4. Tissue/pathology from metastatic site

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Priority Order for Using Documentation to Identify Histology (/3 only)

5. Scan: In priority order:
   A. MRI
   B. CT
   C. PET
   D. Angiogram
6. Code histology documented by physician when none of the above are available-in the following priority order:
   A. Treatment plan
   B. Documentation from Tumor Board
   C. Documentation in record that refers to original path, cytology, or scans
   D. Physician's reference to type of cancer (histology) in record

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Priority Order for Using Documentation to Identify Histology (/1 only)

1. Pathology/tissue from resection of primary tumor
   - A. The addendum(s) and/or comment(s)
   - B. Final diagnosis/synoptic report
   - C. CAP protocol
   - D. Biomarkers
2. Pathology/tissue from biopsy of primary tumor
   Same A, B, C, D as #1.
3. Cytology (Usually CSF)

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Priority Order for Using Documentation to Identify Histology (/1 only)

4. Scan: In priority order:
   - A. MRI
   - B. CT
   - C. PET
   - D. Angiogram

6. Code histology documented by physician when none of the above are available-in the following priority order:
   - A. Treatment plan
   - B. Documentation from Tumor Board
   - C. References to pathology diagnosis
   - D. Physician's reference to type of cancer (histology) in record

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Conflicting Info on Path Reports

Follow these steps for coding histology and/or behavior when there are discrepancies:

When possible, get advice from pathologist

When possible, contact attending physician

When possible, consult with a registry advisor

If none of those options are available, code from the most dependable source (See priority list for coding histology)

Review of STR

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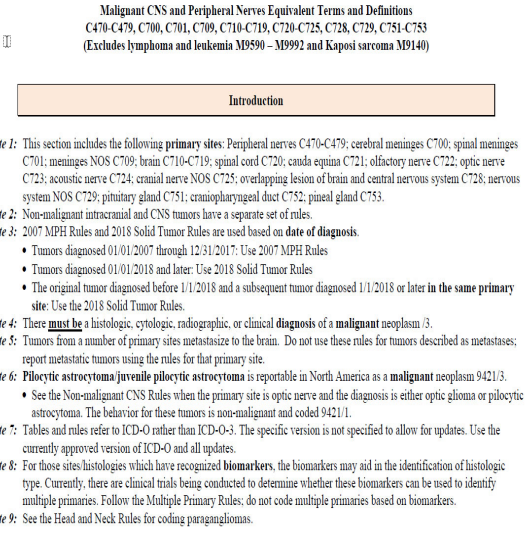
Review of Solid Tumor Manuals

- Malignant CNS Solid Tumor Rules
- Non-malignant CNS Solid Tumor Rules

https://seer.cancer.gov/tools/solidtumor/



Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Introduction

Note 1: This section includes the following **primary sites**: Peripheral nerves C470-C479; cerebral meninges C700; spinal meninges C701; meninges NOS C709; brain C710-C719; spinal cord C720; cauda equina C721; olfactory nerve C722; optic nerve C723; acoustic nerve C724; cranial nerve NOS C725; overlapping lesion of brain and central nervous system C728; nervous system NOS C729; pituitary gland C751; craniopharyngeal duct C752; pineal gland C753.

Note 2: Non-malignant intracranial and CNS tumors have a separate set of rules.

Note 3: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

- Tumors diagnosed 01/01/2007 through 12/31/2017: Use 2007 MPH Rules
- Tumors diagnosed 01/01/2018 and later: Use 2018 Solid Tumor Rules
- The original tumor diagnosed before 1/1/2018 and a subsequent tumor diagnosed 1/1/2018 or later **in the same primary site**: Use the 2018 Solid Tumor Rules.

Note 4: There **must be** a histologic, cytologic, radiographic, or clinical **diagnosis** of a **malignant** neoplasm /3.

Note 5: Tumors from a number of primary sites metastasize to the brain. Do not use these rules for tumors described as metastases; report metastatic tumors using the rules for that primary site.

Note 6: **Pilocytic astrocytoma/juvenile pilocytic astrocytoma** is reportable in North America as a **malignant** neoplasm 9421/3.

- See the Non-malignant CNS Rules when the primary site is optic nerve and the diagnosis is either optic glioma or pilocytic astrocytoma. The behavior for these tumors is non-malignant and coded 9421/1.

Note 7: Tables and rules refer to ICD-O rather than ICD-O-3. The specific version is not specified to allow for updates. Use the currently approved version of ICD-O and all updates.

Note 8: For those sites/histologies which have recognized **biomarkers**, the biomarkers may aid in the identification of histologic type. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

Note 9: See the Head and Neck Rules for coding paragangliomas.

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Pop Quiz

Pop Quiz 1

- 4/16/20 brain MRI: 1.2 cm meningioma (9530/0) rt temporal lobe
- Active surveillance started
- Tumor increases in size, pt becomes symptomatic
- 5/4/21 gross total resection: Anaplastic meningioma (9530/3)

How many primaries?

Pop Quiz 2

- 4/16/20 brain MRI: 1.2 cm meningioma (9530/0) rt temporal lobe
- 4/20/20 Gross total resection: Transitional meningioma (9537/0)
- 5/1/21 brain MRI: 2.3 cm lesion rt temporal lobe
- 5/7/21 resection: anaplastic meningioma (9530/3)

How many primaries?

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Acknowledgment

Special thank you to Mindy Young!
Senior Cancer Registrar
Dana-Farber Cancer Institute

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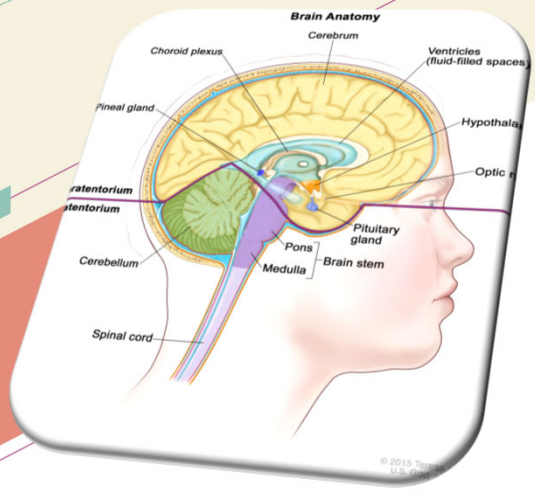

Pediatric Brain Tumors

With Angela Costantini, BA, CTR

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Causes of Pediatric Cancer

For the most part, pediatric cancer is a random occurrence. Sometimes, it can be associated with specific congenital abnormalities.

Diseases and Syndromes

- Beckwith-Wiedemann syndrome
- Li-Fraumeni syndrome
- Down syndrome
- Neurofibromatosis
- Denys-Drash syndrome
- Klinefelter syndrome

Causes of Increased Risk

- Prematurity
- Biliary atresia
- Familial adenomatous polyposis
- RB1 gene mutation
- Tuberous sclerosis complex
- History of solid organ transplant

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GENERAL INFO ABOUT PEDIATRIC BRAIN TUMORS

- Brain tumors are the most common solid tumor in children and the second most common type of cancer in kids overall.
- They account for 15% of pediatric cancers.
- They are often categorized into low grade (WHO grade 1/2) or high grade (WHO grade 3/4) and supratentorial (cerebrum) or infratentorial (cerebellum/brainstem)
- The most common types in children are: medulloblastomas, astrocytomas, brain stem gliomas, ependymomas and optic nerve gliomas
- This includes benign, borderline AND malignant tumors. All brain/CNS tumors have been reportable since 2003 regardless of behavior
- Brain tumors do not have their own staging system. They are assigned WHO grades and are given a SEER Summary Stage, but that's it.

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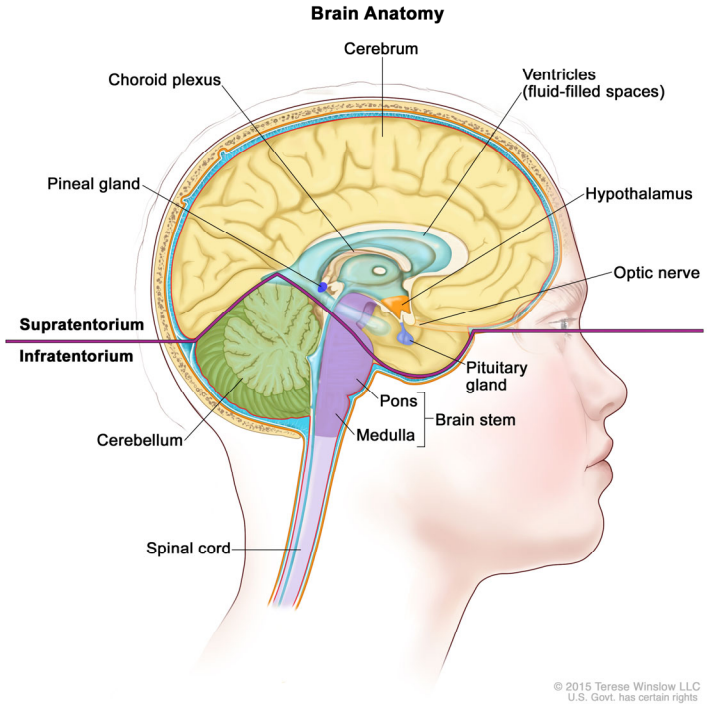

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WHAT WE WILL COVER

- Neurofibromatosis and related tumors
- Optic Pathway Gliomas (OPGs) and Juvenile Pilocytic Astrocytomas (JPAs)
- Diffuse Intrinsic Pontine Gliomas (DIPG)
- Other Treatment considerations

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NEUROFIBROMATOSIS AND RELATED TUMORS

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Neurofibromatosis

Is it reportable? No.

NF is NOT a reportable disease.

WHAT IS IT? It is a genetic disorder that causes tumors to form on nerve tissue. There are three different types – NF1, NF2 and Schwannomatosis. The tumors can form anywhere in the nervous system, including the brain, spinal cord and nerves.

What IS reportable are the tumors associated with the disease.

- There are three different types – NF1, NF2 and Schwannomatosis. The tumors can form anywhere in the nervous system, including the brain, spinal cord and nerves.
- **Type 1** (1 in 3500 births) can cause bone deformities, learning disabilities, and high blood pressure.
- **Type 2** (1 in 25,000 births) can cause hearing loss, vision loss, and difficulty with balance.
- **Type 3** (1 in 40,000 births) can cause chronic pain throughout the body.
- NF1 usually appears in childhood while NF2 and schwannomatosis usually appear in early adulthood.

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NF-Associated Tumors

- **COMMON BENIGN TUMORS**: Optic Nerve Gliomas (OPG), Schwannomas, Plexiform Neurofibromas, Meningiomas, Ganglioneuromas, Acoustic Neuromas
- **COMMON MALIGNANT TUMORS**: MPNSTs, High Grade Gliomas, Undifferentiated Pleomorphic Sarcomas, Leiomyosarcomas, Astrocytomas

- Tumors associated with NF1 and NF2 are reportable when they meet the behavior, site and histology reportability requirements. Use of the Solid Tumor Rules is important in assigning sequence numbers and new primaries.
- Some of these benign tumors also have the potential to transform into malignant tumors.
- The malignant tumors usually arise from neurofibromas under the skin or from plexiform neurofibromas. People who have NF1 also have a higher risk of other forms of cancer, such as breast cancer, leukemia, colorectal cancer, brain tumors and some types of soft tissue cancer. Women who have NF1 should start screening for breast cancer at an earlier age than the general population.

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Notes on NF from the Solid Tumor Rules Manual

G. Neurofibromatosis NOS, Neurofibromatosis 1 (NF1), Neurofibromatosis 2 (NF2), and schwannomatosis are genetic syndromes and not reportable neoplasms. People with this genetic syndrome do have a high risk of developing:
   i. Non-reportable non-malignant tumors occurring in skin and sites other than CNS AND
   ii. Reportable malignant tumors

Neurofibromatosis, NOS	**9540/1**	Genetic disease that produces non-malignant tumors in the skin, brain, CNS, and other sites. The brain and CNS tumors spawned by NF, NOS are reportable, the genetic disease is not.
Neurofibromatosis, type 1 (NF1)	**No code***	Genetic disease that produces non-malignant tumors in the skin, brain, CNS, and other sites. The brain and CNS tumors spawned by NF1 are reportable, the genetic disease is not.
Neurofibromatosis, type 2 (NF2)	**No code***	Genetic disease that produces non-malignant tumors in the skin, brain, CNS, and other sites. The brain and CNS tumors produced by NF2 are reportable, the genetic disease is not.

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Notes on NF from the Solid Tumor Rules Manual

Rule H1 Code the **reportable CNS tumor** (Table 3 in the Equivalent Terms and Definitions) when a patient has any of the following:
- Neurofibromatosis type 1 (**NF1**)
- Neurofibromatosis type 2 (**NF2**)
- **Schwannomatosis**

Note 1: **Do not code** NF1 or NF2 as neurofibromatosis. NF1, NF2, and schwannomatosis are genetic syndromes which have a high risk of developing reportable and non-reportable tumors. **ONLY** abstract **reportable tumors** such as malignant peripheral nerve sheath tumors.
Note 2: Tumors are reportable when they meet the behavior (/3) and histology requirements (see **Reportability Criteria**).
Note 3: Schwannomatosis is a newer term for a distinct subtype/variant of the genetic diseases NF1 and NF2.
Example: Patient presents with vestibular schwannoma (acoustic neuroma). Genetic testing proves the patient has NF2. Report the acoustic nerve neuroma.

Rule H7 Code the **reportable CNS tumor** (Table 6 in the Equivalent Terms and Definitions) when a patient has any of the following:
- Neurofibromatosis type 1 (**NF1**)
- Neurofibromatosis type 2 (**NF2**)
- **Schwannomatosis**

Note 1: Only report tumors such as:
- Plexiform neurofibroma (usually NF1)
- Tumors of brain and spinal cord, meningiomas of cranial or spinal dura, glioma (usually NF2)

Note 2: Tumors are reportable when they meet the behavior code, site, and histology reportability requirements (see **Reportability Criteria**). Do not code neurofibromatosis.
Note 3: NF1 is a genetic disorder causing lesions in the skin, nervous system and skeleton.

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Abstracting NF Tumors: Site and Histology

- First: assign primary site.
  - Often, these patients have numerous benign and malignant tumors, sometimes within the same primary site. Focus on one at a time.
- Second: choose appropriate histology
  - Make sure you CONFIRM it is for the tumor you are abstracting. Just like when you are assigning primary site, there may be multiple path reports on multiple tumors. Include only the path reports that relate to the tumor being abstracted, if applicable.
- Third: determine sequence number
  - Because of the possibility of multiple tumors both benign and malignant, it is important for you to appropriately assign the sequence number.

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Abstracting NF Tumors: Treatment Info

- Treatment depends on the tumor type. For some, like OPGs, active surveillance is all that is required. Many require surgical intervention, especially the benign ones that are causing secondary issues to mobility, hearing or pain.
- Malignant tumors may be treated with chemo and/or radiation and surgery
- Selumetinib, imatinib and everolimus (chemo) are current treatments for many of the tumors of NF. They've been FDA approved in certain capacities but are also being trialed in other capacities.
- Selumetinib is the first drug approved by the FDA specifically for NF. It's an MEK-inhibitor.
- Immunotherapy is emerging as a treatment option as well. Right now, we see a lot of beviacizumab (a monoclonal antibody), specifically for NF2 tumors.

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Monoclonal Antibodies

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Optic Pathway Gliomas and Juvenile Pilocytic Astrocytomas

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What is a Pilocytic Astrocytoma?

- It's a slow-growing brain tumor (sometimes spinal) that arises from astrocytes, the supportive cells in the nervous system.
- Usually occurs in children and are now considered to be the most benign type of astrocytoma.
- Often called "juvenile pilocytic astrocytoma". When positioned in the optic nerve, they are called Optic Pathway Gliomas or "OPGs".
- They usually have well-defined boundaries, so they can often be fully removed and have a low likelihood of recurrence.
- Survival rates at 10 years after dx are 92.2%

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How is Pilocytic Astrocytoma diagnosed?

- Symptoms to look for: headaches, fatigue, nausea, vomiting, balance and walking difficulties, weight gain or loss, premature puberty or endocrinopathies, loss of vision or vision issues
- In young children, there may also be regression of milestones like walking or talking
- MRIs of the brain and/or orbits, CTs of the head, and MRs of the spine to look for drop mets (or even a primary site of the spine)
  - OPGs are diagnosed through imaging
  - JPAs are diagnosed through imaging and biopsies/resection
  - OPGs can also be diagnosed in children without NF
- Bilateral OPGs in NF patients are a single primary
- The morphology code is always 9421.

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Behavior Code Changes and Staging

BEHAVIOR CODES

2017 and older diagnoses of OPG are to be coded with a behavior code of **3**.

2018+ diagnoses of OPG are to be coded with a behavior code of **1**.

ALL non-optic nerve JPAs are currently coded with behavior code of **3**.

STAGING

For behavior 3 tumors, SEER Summary Stage can be 1 or 2 (rarely a 7 or 9).

For behavior 1 tumors, SEER Summary Stage will be SS 8 (2018+ diagnoses).

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What is the treatment for Pilocytic Astrocytoma?

- For OPGs, active surveillance is usually the first course of treatment. If it begins to grow and cause issues, chemo or radiation is administered.
- For non-optic nerve JPAs, they may be under active surveillance OR if they are causing more issues, they may be treated in multiple ways:
  - Gross total or subtotal resection of tumor
  - XRT to primary tumor or spine; proton or photon
  - Chemo is very common, often multiple drugs and often as an established regimen
- After surgical resection of brain tumors, decadron or another hormone is often given to reduce swelling. This is NOT counted as treatment of the tumor.

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CNS Surgical Codes

- 02 = a non-stereotactic biopsy of a CNS tumor (i.e.. An endoscopic biopsy of an intraventricular tumor)
- 20 = stereotactic biopsy of a CNS tumor; local excision of tumor, lesion or mass; excisional biopsy
- 21 = subtotal/near total resection of **tumor, lesion or mass in brain**
- 22 = resection of **tumor of spinal cord or nerve** (subtotal or gross total)
- 30 = radical, total, gross resection of **TUMOR, LESION or MASS in the brain**
- 40 = partial resection of **LOBE of brain**, when the surgery cannot be coded as 20-30
- 55 = gross total resection of **LOBE of brain** (lobectomy)

Codes 30-55 are not applicable for spinal cord or spinal nerve primary sites

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Diffuse Intrinsic Pontine Gliomas (DIPG)

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A Video Intro to DIPG

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What is DIPG?

- DIPG is the most common brainstem tumor in children, comprising 75-80% of all pediatric brainstem tumors
- Leading cause of brain tumor-related death in children
- 150-300 diagnoses in USA per year; average age at diagnosis is 6-7yrs
- 30% survival rate <1year (mean range 8-11mos); 90% of patients die within 2yrs of diagnosis
- There are a few long-term survivors, but there is no cure.
- Research has shown some factors that indicate surviving >2yrs: patient age <3yrs or >10yrs, fewer symptoms at diagnosis, smaller tumors w/ less evidence of extension beyond pons, HIST1H3B mutation
- No evidence that DIPG is caused by any environmental factor or inherited gene variation

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DIPG: How Does it Present?

- Neurologic symptoms may appear 6mos prior to diagnosis and then become more evident ~1 month prior.
- DIPGs cause pressure on cranial nerves
  - Affecting vison, eye & eyelid movement, facial expressions, chewing & swallowing, bladder functions, balance & weakness issues, and headaches
  - May also cause weakness in arms and legs and difficulty speaking and walking
- Rapid growth in tumor size increases pressure inside the skull which then causes fluid build-up resulting in hydrocephalus. This in turn causes headaches, nausea, vomiting, and fatigue.

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DIPG: Diagnosis

- Often, it is first diagnosed or suspected during an MRI of the brain. “DIPG” is a clinical diagnosis. It’s a glial tumor originating in the pons (between the midbrain and medulla) that currently unresectable due to its location.
- Historically, a biopsy was thought to be too dangerous due to risk of damaging the brainstem however, new stereotactic biopsy routes are being studied
  - Transcortical: samples lesions at all brainstem levels
  - Transcerebellar: samples upper medullary and pontine masses
  - Complications include worsening of preexisting ataxia, nerve VI & VII palsy, hemorrhage, brainstem syndromes, and inability to move eyes; deeper bxs can affect motor pathways
  - Autopsies have revealed local infiltration of the medulla, thalamus and midbrain, as even as far reaching as the frontal lobe
- Studies of biopsied tissue evaluate for tumor sequencing which can individualize patient immunotherapy treatment plans

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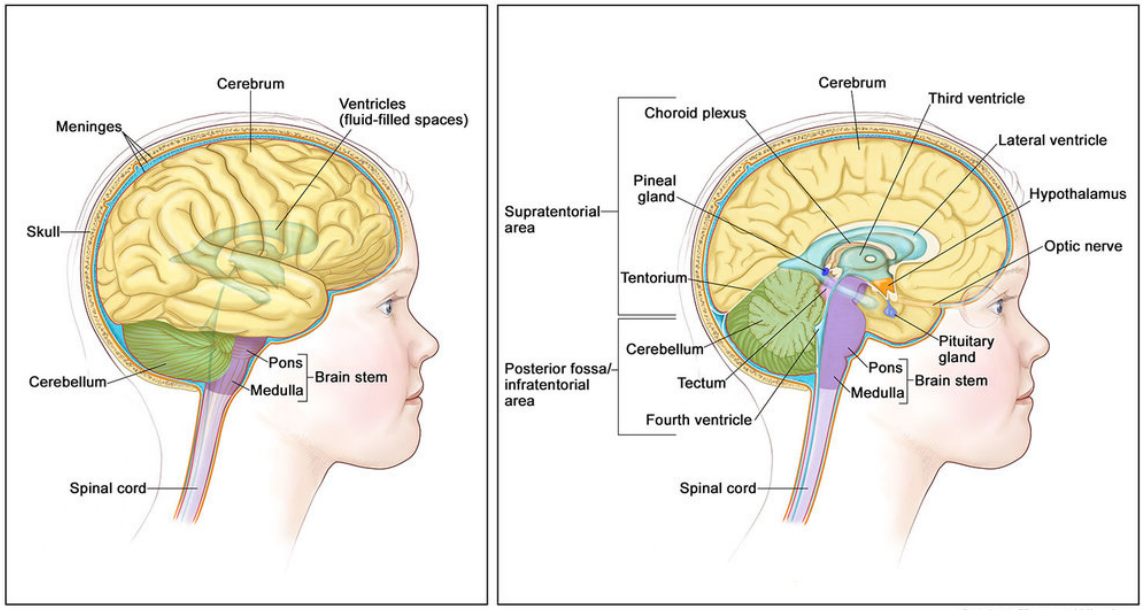

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DIPG: Site and Histology

- Site is always C71.7 – Brainstem, Pons
  - The site and behavior of the tumor is what makes it a DIPG
- Morphology can be a variety of codes
  - Ranges from WHO grade II – IV astrocytic gliomas, glioblastoma or oligodendroglioma, though biopsies are often rare
  - histone 3.3, gene H3F3A: carries a worse prognosis, found to be less responsive to radiotherapy, tends to relapse earlier, and typically has a higher rate of metastatic recurrence; WHO classifies this set of tumors as "diffuse midline glioma, H3K27M mutant" (code 9385/3)
  - histone 3.2, gene HIST2H3C
  - histone 3.1, genes HIST1H3B/C: specific to pontine gliomas, occurs more frequently in females and at a younger age
  - An unbiopsied DIPG that is not pathologically confirmed as anything else will most likely get the code of 9380/3 with a clinical grade of 4.

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DIPG: Treatment

- Steroids: usually limited to short burst use, sometimes used indefinitely.
  - Pros: alleviate/ stabilize neurological symptoms
  - Cons: impaired sleep, impaired wound healing, behavior issues, endocrine & metabolic function effects
- Most tumors will begin to grow again about 6-12mos after diagnosis which again causes neurologic symptoms. Steroids may be restarted, but there is no further tx shown to improve survival
- Radiation has shown to increase overall survival 2-4mos
  - Photon radiotherapy directed to the tumor for total 5400cGy at daily dose of 180cGy
- Palliative care in conjunction with Neuro-oncology can maximize patient function during treatment and identify progression ahead of standard clinical assessments
- Chemo has shown to have no survival benefit, but there are trials available with new immunotherapy agents called monoclonal antibodies

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PEDIATRIC TREATMENT CONSIDERATIONS

And at what age is it no longer "pediatric cancer"?

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Approaches to Treatment

While the treatment for pediatric cancer follows standard guidelines, there are more variables to be considered.

- Treatment is individualized rather than purely site-specific. What can the patient handle? Will it affect growth and development? Will the potential toxicities of a treatment make it too big of a risk?
- What's best for the family? Religious beliefs and social structures/supports are weighed heavily. Palliative care helps both the patient and their family members.
- The ultimate goal is long-term survivorship. Pediatric patients have their entire adult lives left to live and many treatments can cause secondary issues. These issues are discussed.
- When to intervene? Is a guardian refusing treatment necessary for the survival of the child?

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Adolescent/Young Adults (AYA)

- Typically, pediatric cancer "ages" are 0-19, but there is an increasing population of young adults being treated at pediatric hospitals. Adolescents are ages 15-19. Young adults are ages 20-39.
- AYAs have different considerations than young children that are now being addressed and tracked within cancer programs:
  - Oncofertility
  - Sexual health and safety during treatment
  - Mental health
  - Survivorship and transitioning to adult care
  - Job and career disruption
  - Being a parent during treatment

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Resources

- The DIPG/DMG Resource Network
- Toronto Childhood Cancer Stage Guidelines
- ICCC Pediatric Groups
- Neurofibromatosis Network
- Ohio Department of Health's Childhood Cancer Website
- Pediatric Hospital CTRs Facebook Group

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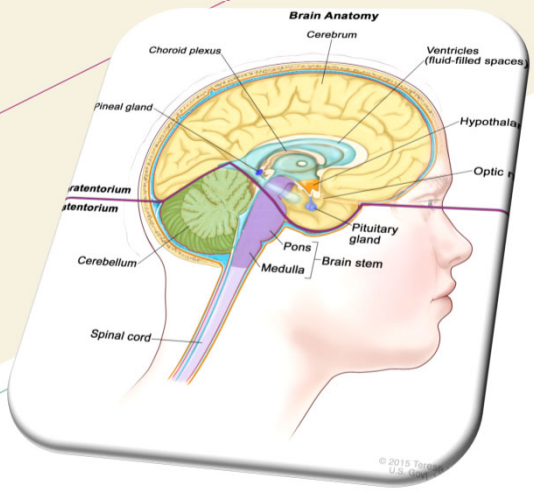

Thank you!

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Coming UP…

- Back to The Future: What year is it and What did I miss?
  - Guest Host: Nancy Etzold, CTR; Lisa Landvogt, CTR
  - 7/07/2022
- Solid Tumor Rules 2022
  - Guest Host: Denise Harrison, CTR; Louanne Currence, RHIT, CTR
  - 08/04/2022

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Fabulous Prizes

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CE Certificate Quiz/Survey

CE Phrase

Hypothalamus

Link

https://survey.alchemer.com/s3/6563881/Central-Nervous-System-2022

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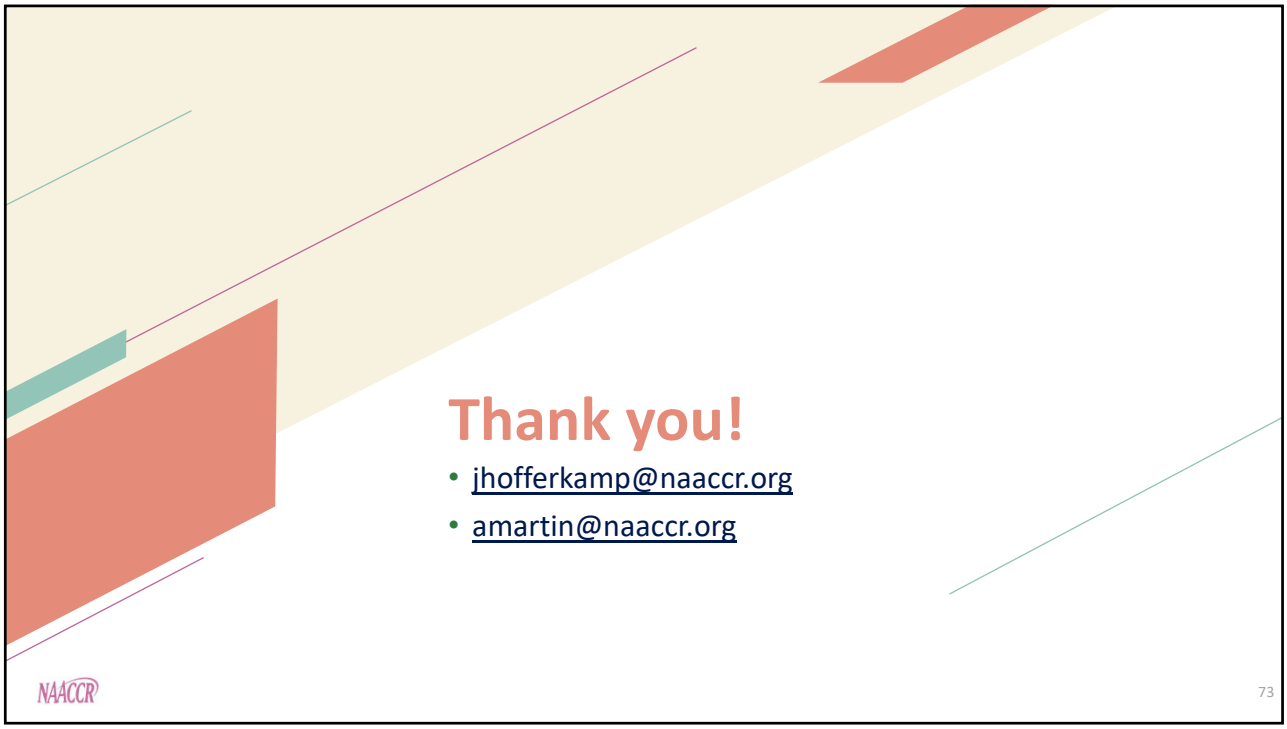
Thank you!
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