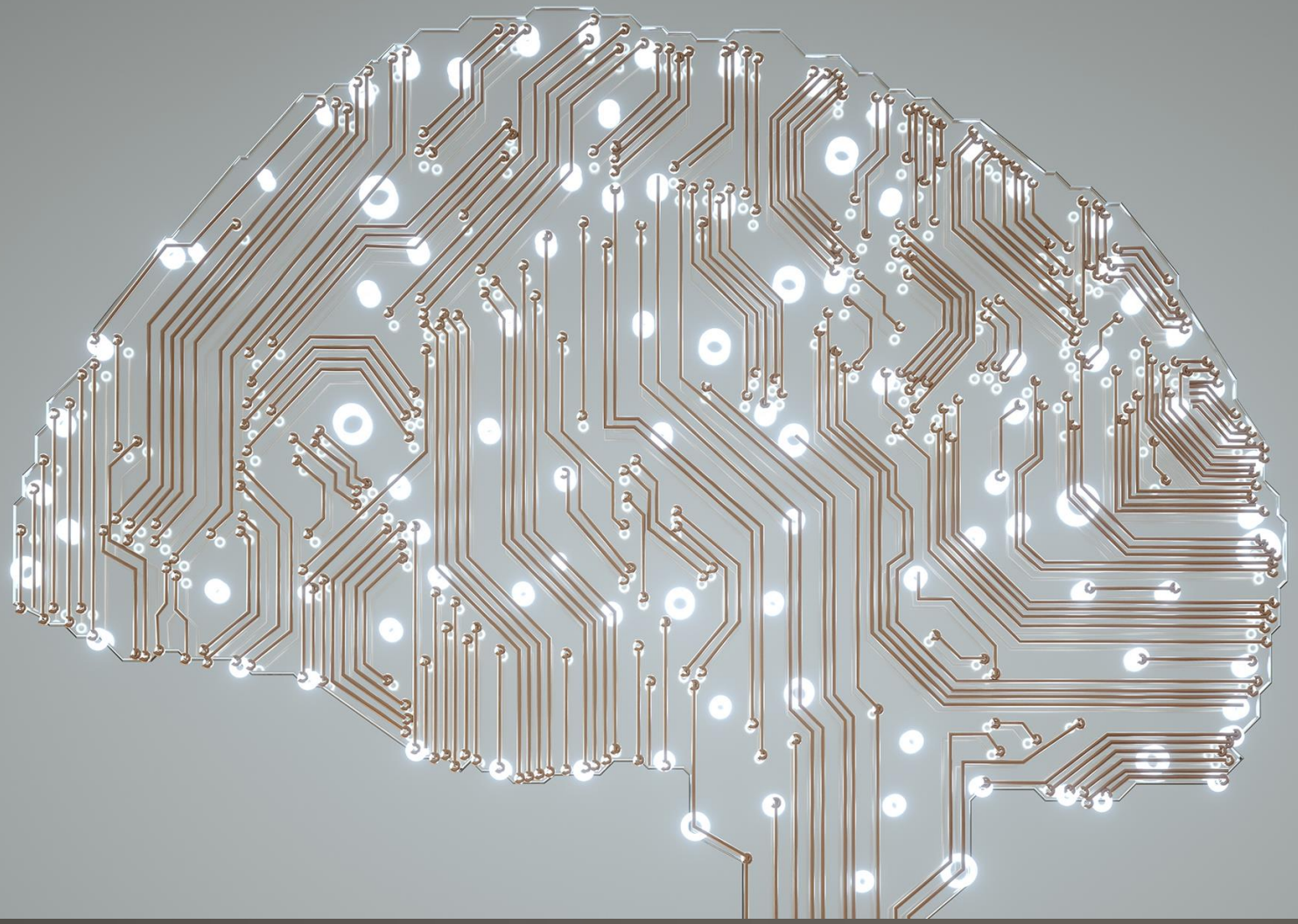


Little Patients, Big Data

Abstracting Brain Tumors in the Younger Population

Objectives

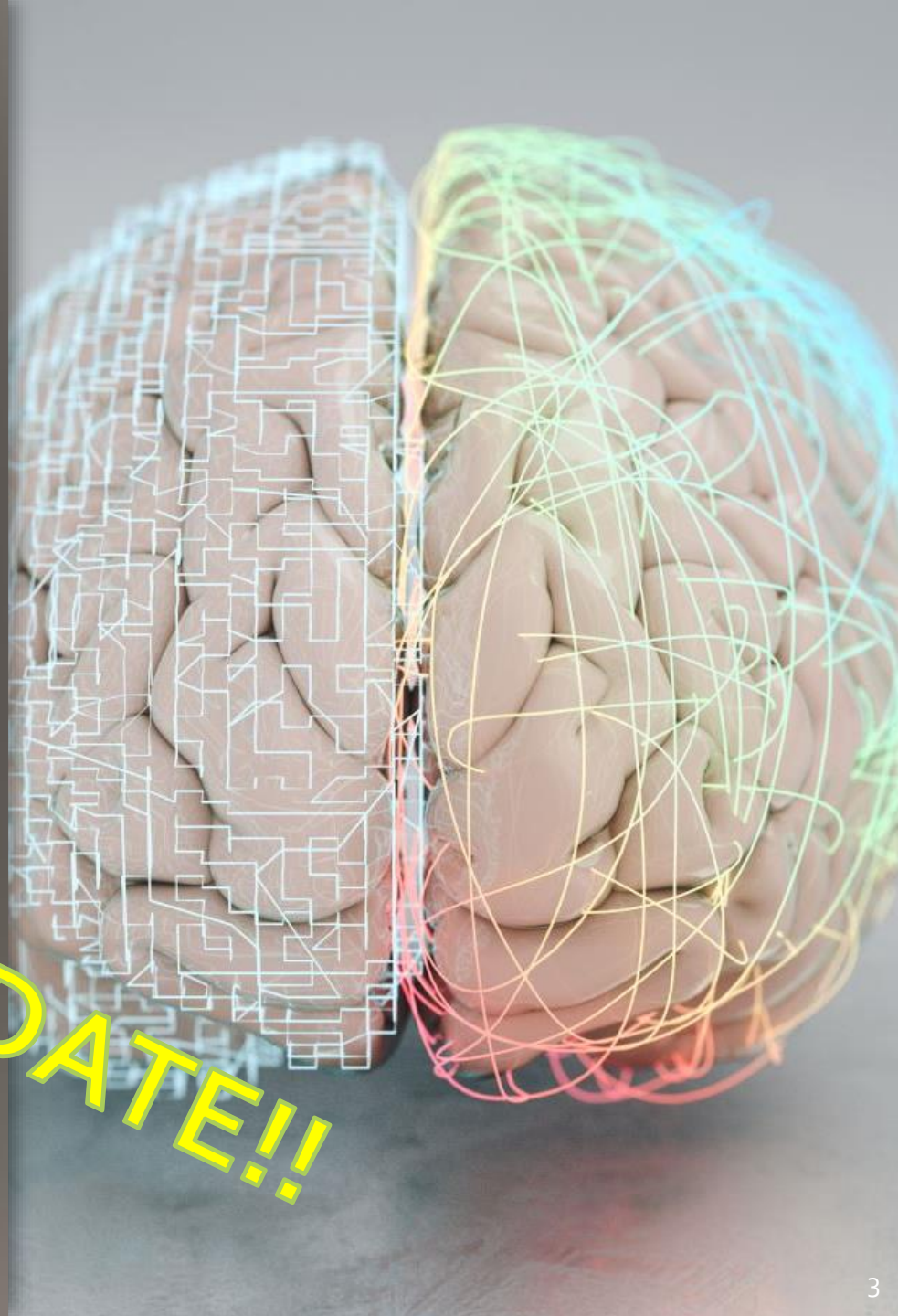
Learn about pediatric brain tumors in general

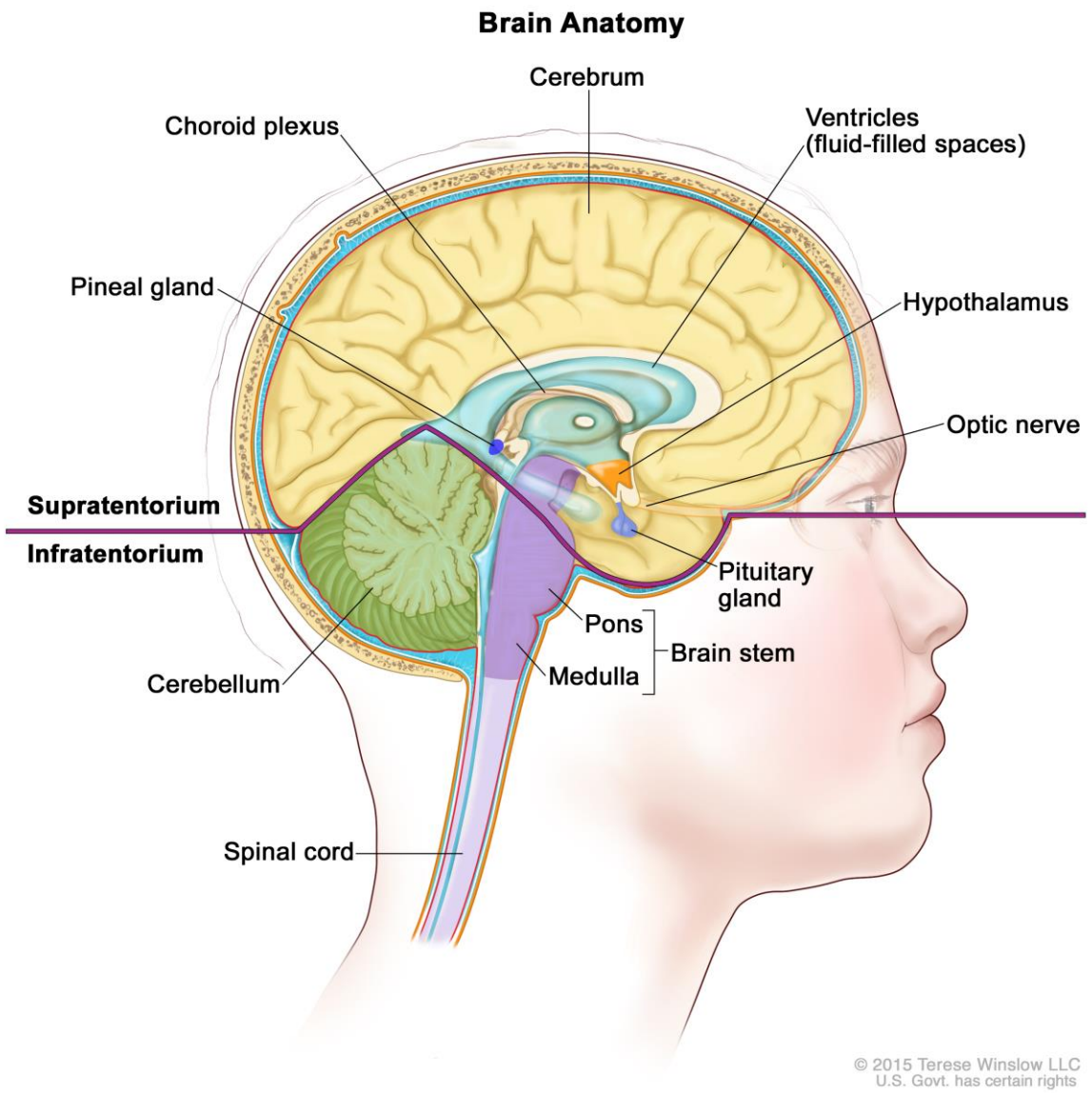
Know different types of pediatric brain tumors and review case scenarios

Get an overview of the PDCS data items for CNS tumors and review other common coding quirks

Pediatric Brain Tumors

- Brain tumors are the most common solid tumor in children and the second most common type of cancer in kids overall
- Approximately 4% of all brain tumor cases diagnosed each year occur in children ages 0-14.
- They account for 15% of pediatric cancers
- Among all childhood cancers, brain tumors are the leading cause of mortality
- Like adult brain tumors, they are classified into supratentorial and infratentorial lesions and are given WHO grades 1-4 which are further categorized as low-grade (1-2) and high grade (3-4)
- Multiple predisposition syndromes show associations with brain tumors (DICER1, Li-Fraumeni, neurofibromatosis, tuberous sclerosis, and Von-Hippel Lindau), although we have been unable to capture this in a standardized way.
- **Starting with 2027 diagnoses, we will be able to capture these codes in the secondary diagnosis data fields. Thank you, CoC!**




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
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Brain Sites

Supratentorial

- C71.0* (see exceptions on infratentorial list)
- C71.1
- C71.2
- C71.3
- C71.4
- C71.5
- C71.8
  - Corpus Callosum
  - Tapetum
- C71.9
  - Anterior Cranial Fossa
  - Middle Cranial Fossa
  - Suprasellar

Infratentorial

- C71.0
  - Hypothalamus
  - Pallium
  - Thalamus
- C71.6
- C71.7
  - Tectal Plate, NOS
- C71.9
  - Cranial Fossa, NOS
  - Posterior Cranial Fossa

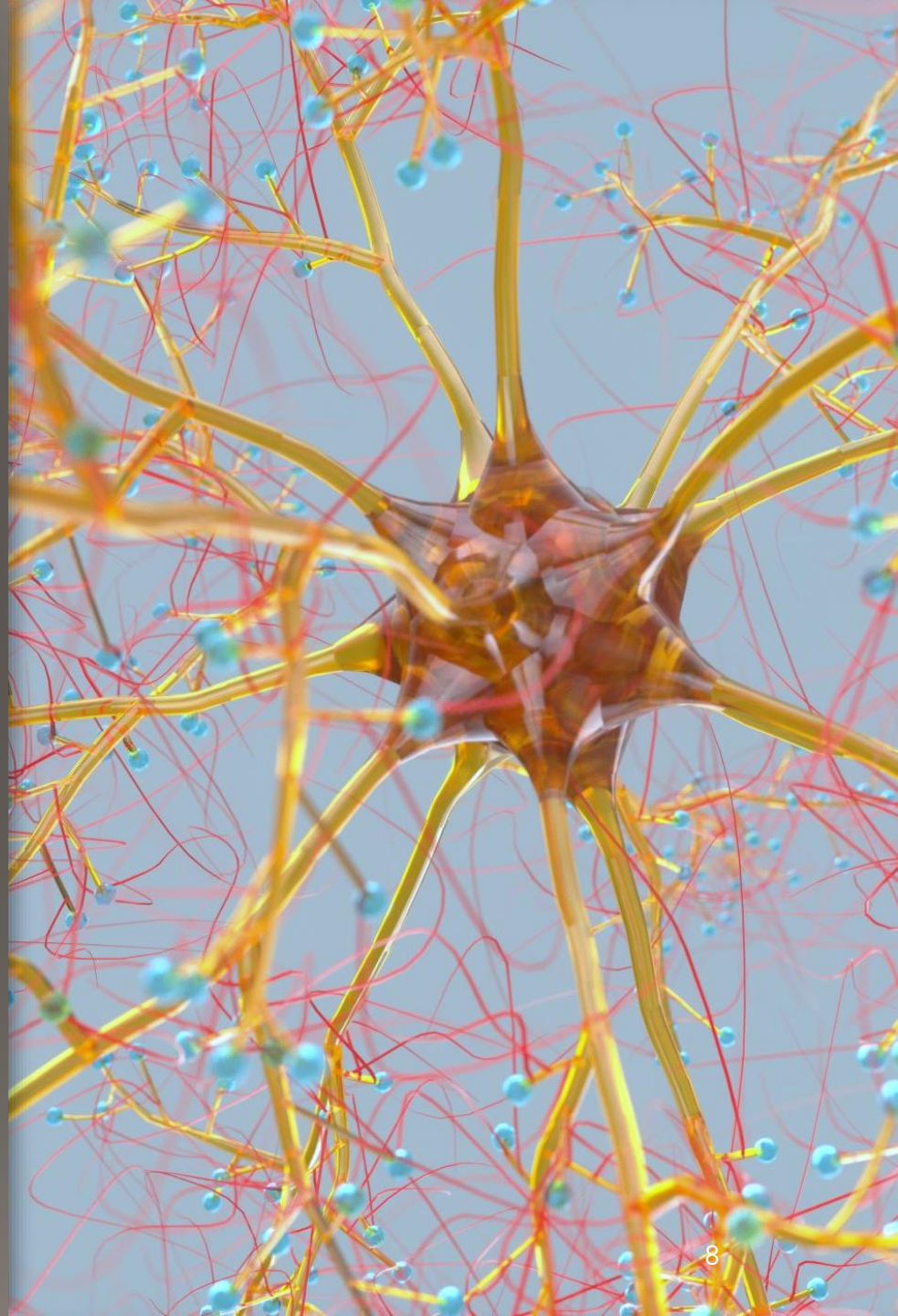
TUMORS WE WILL COVER

- Juvenile Pilocytic Astrocytomas (JPAs)
- Diffuse Intrinsic Pontine Gliomas (DIPG)
- Ependymomas
- Medulloblastomas

Juvenile Pilocytic Astrocytomas

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%



How does Pilocytic Astrocytoma present?

- 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.

BRAIN TUMORS: PILOCYTIC ASTROCYTOMAS

- Data item relationships to watch are **DX YEAR, BEHAVIOR, PRIMARY SITE**.
  - Morphology code is always 9421, but the behavior code changes based on site and dx year.

2017 and Older
Site = C72/C71
Behavior = 3

2018 -2022
Site = C72.3
Behavior = 1
Site = C71/C72 (NOT C72.3)
Behavior = 3

2023+
Site = C72/C71
Behavior = 1

Coding Pilocytic Astrocytoma Histology

- 9421/1
  - Pilocytic astrocytoma
  - Juvenile pilocytic astrocytoma
  - Optic nerve glioma
  - Diffuse astrocytoma, MYB- or MYBL1- altered
  - Diffuse low-grade glioma, MAPK pathway altered
- 9421/3
  - High-grade astrocytoma with piloid features
  - As of 2023, this is the ONLY tumor type associated with this exact histology code with a behavior of 3.
- SSDI help: Brain Molecular Markers
  - 19 – 9421/1 Diffuse astrocytoma, MYB- or MYBL1-altered
  - 20 – 9421/1 Diffuse low-grade glioma, MAPK pathway-altered
  - 99 – If codes 19 or 20 don't apply or are not microscopically confirmed (OPGs for example)
  - Don't use code 86 "benign/borderline"

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**.
- **Only HGAPs will get a behavior code of 3 now.**
- A diagnosis of "juvenile pilocytic astrocytoma or just "pilocytic astrocytoma" will get a behavior code of 1 for 2023+ diagnoses

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).

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.

Pediatric Brain Tumors: Astrocytomas

- BRAF mutations in astrocytomas
  - Associated with poorer prognosis and predicts lack of response to anti-EGFR therapies
  - The most common mutations are:
    - BRAF V600E
    - BRAF KIAA1549
  - BRAF status IS collected as a pediatric SSDI, so it's considered part of both the PDCS and the SSDI manual.
- Mutations vs "wild type"
  - Wild type just means that there's no mutation
- In general, tracking these cytogenetics for brain tumors is really helpful. Put them in your text if you can!

The Pediatric Data Collection System

… a commercial break!

What is the PDCS?

The Pediatric Data Collection System is the very first coding system in the cancer registry management profession dedicated to capturing key information related to the staging and spread of pediatric type tumors.

It is a combination of staging elements and site-specific data items either related to staging, treatment, or surveillance of the disease.

With the impending release of the Comprehensive Abstracting Manual for Pediatric Cancer (CAMP) later this year, the PDCS will become "Appendix 1" of that publication. It's already contained as "Appendix 1" on the NAACCR website.

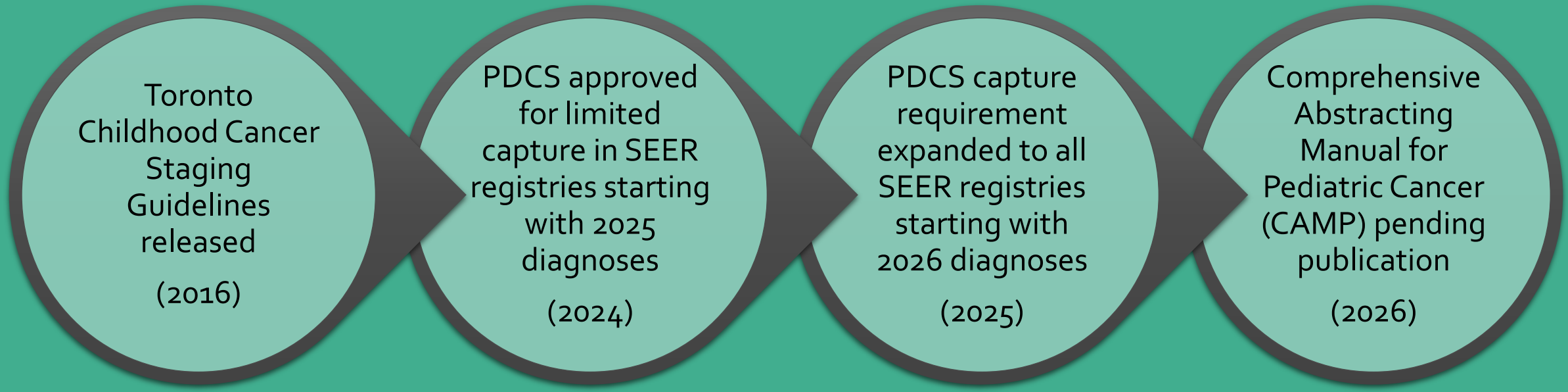
Essentially, we had to "snowball" our efforts. First the PDCS was approved for a small group of registries, then a bigger group of registries, then we release the larger manual, and hopefully expanded requirements for these data items will be implemented nationally!

Pediatric Data Collection System (PDCS)

There are four main data items in the PDCS:

1. Pediatric Primary Tumor
2. Pediatric Regional Nodes
3. Pediatric Mets
4. Applicable SSDIs (schema-dependent)

Timeline of Pediatric Cancer Coding



Now, back to the show

… or your regularly scheduled programming?

PDCS Data Items: Astrocytoma

Pediatric Primary Tumor
Different codes for Brain C70.0, C71.0-C71.9 and CNS Other C70.1,C70.9, C72.0-72.9
–Subsite for Brain sites needs to be determined if Infratentorial or Supratentorial

Pediatric Regional Nodes - N/A (code 888)

Pediatric Mets
Code 10 distant lymph nodes
Code 70 any other mets with or without distant LN

SSDI – BRAF
- Code 0 when BRAF negative/wild type/not detected
- Code 1, 2 or 3 when BRAF identified/detected
- Code 4 when BRAF identified, mutation not known
- Code 9 when insufficient, equivocal, no microscopic confirmation of tumor, BRAF not done

Astrocytoma Case Scenario

- Imaging: Patient with infiltrating process in posterior cranial fossa with involvement of the corpus callosum.
- No other involvement noted
- Diagnosis: Glioblastoma multiforme, BX confirmed.

Data Item	Value	Comments
Pediatric Primary Tumor	500	• Posterior cranial fossa is an infratentorial site • Corpus callosum is a supratentorial site • Both infratentorial and supratentorial sites are involved
Pediatric Regional Nodes	888	Default code for CNS tumors
Pediatric Mets	00	Per imaging there is no other involvement noted
BRAF mutational analysis	9	No mention of BRAF on the pathology report

Staging | Site-Specific Data Items | AJCC TNM Staging | PDCS Staging

Pediatric Primary Tumor: 500

Pediatric Regional Nodes: 888

Pediatric Mets: 00

Stage System:

Stage:

Staged By:

Pediatric ID: 3b: Astrocytoma

BRAF Mutational Analysis: 9

BRAF Mutational Analysis

Code	Description
0	Normal; BRAF negative
1	Abnormal (mutated)/detected: BRAF V600E (c.1799T>A) mutation
2	Abnormal (mutated)/detected, but not BRAF V600E (c.1799T>A) mutation
3	Abnormal (mutated)/detected, KIAA1549: BRAF gene fusion
4	Abnormal (mutated), NOS
7	Test Ordered, Results not in Chart
8	Not Applicable
9	Unknown; Not Documented in Medical Record; BRAF not assessed

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Diffuse Intrinsic Pontine Gliomas (DIPG)

A Video Intro to DIPG

DISCLAIMER: This video is taken from a public YouTube source and is used solely for educational purposes. All individuals shown are consenting participants, and no personal identifying information is displayed. The content is not intended as medical advice or diagnosis and does not replace professional healthcare guidance. The presenter and UNC Health Cancer Care are not liable for any misuse or misinterpretation of the material.

What is DIPG?

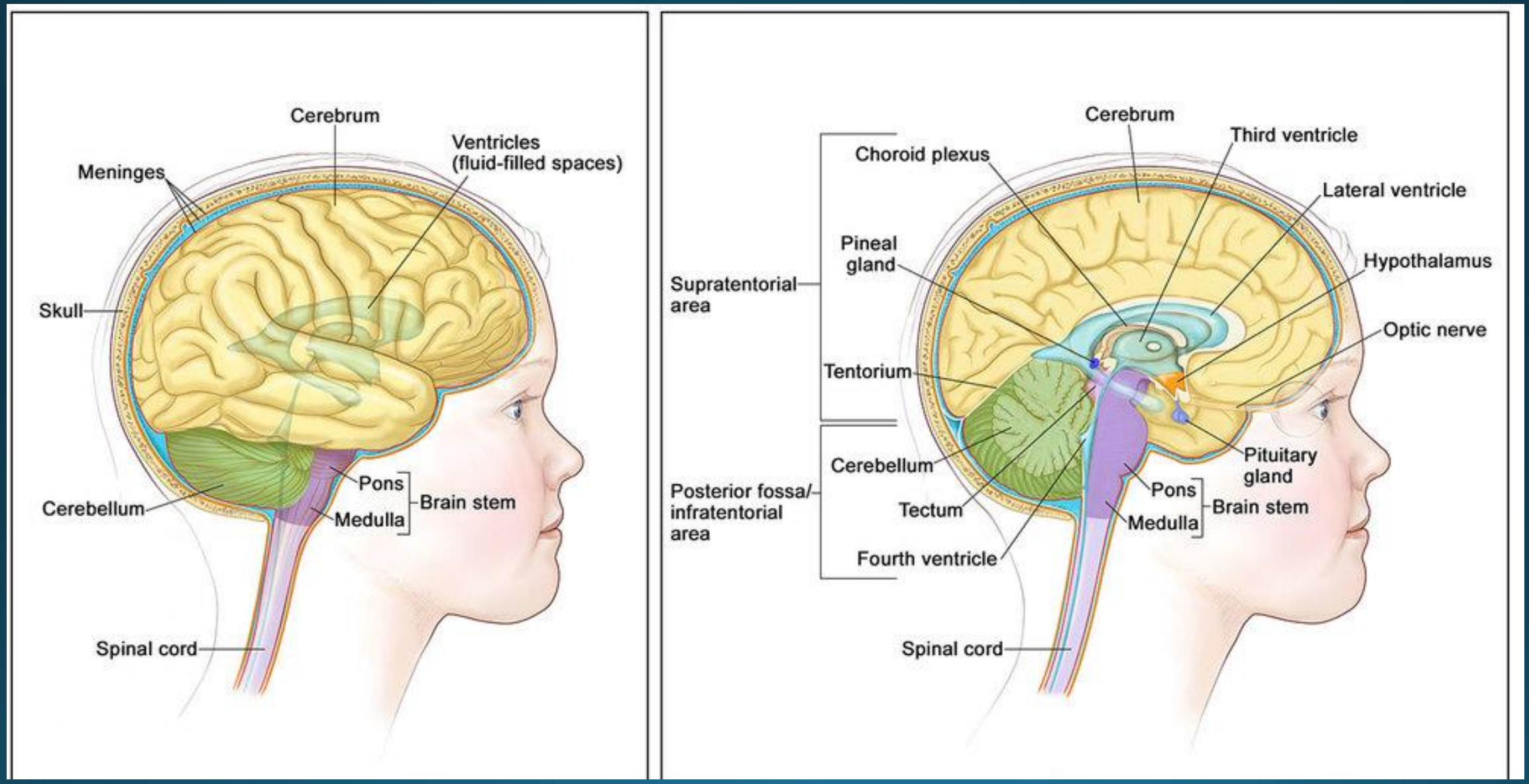
- DIPG is the most common brainstem tumor in children, comprising 75-80% of all pediatric brainstem tumors
- Leading cause of brain tumor-related death in children
- 150-300 diagnoses in USA per year; average age at diagnosis is 6-7yrs
- 30% survival rate <1year (mean range 8-11mos); 90% of patients 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

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 vision, 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.

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


Cerebrum
Ventricles
(fluid-filled spaces)
Meninges
Skull
Pons
Brain stem
Cerebellum
Medulla
Spinal cord
Cerebrum
Third ventricle
Choroid plexus
Lateral ventricle
Pineal
gland
Hypothalamus
Supratentorial
area
Optic nerve
Tentorium
Pituitary
gland
Cerebellum
Pons
Posterior fossa/
infratentorial
area
Brain stem
Tectum
Medulla
Fourth ventricle
Spinal cord

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)
  - 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.

DIPG: Site and Histology

9385/3 holds a variety of different names now:

- Diffuse midline glioma, H3K27 altered
- Diffuse hemispheric glioma, H3G34 mutant
- Infant-type hemispheric glioma
- Diffuse pediatric-type high-grade glioma, H3 and IDH-wildtype

If the location of the tumor is in the pons, then these all clinically qualify as DIPGs.

If they occur in an area outside of the pons, they do NOT qualify as DIPGs.

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

Ependymomas

What are Ependymomas?

- Comprises approximately 9% of all childhood brain and spinal cord tumors, equaling about 200 cases per year in the US
- They arise from ependymal cells that line the ventricles and passageways in the brain and the center of the spinal cord
- In children, 65-75% of ependymomas arise in the posterior fossa around the fourth ventricles

Ependymoma: How Does it Present?

- Posterior fossa (infratentorial) ependymomas:
  - obstructive hydrocephalus caused by obstruction at the level of the fourth ventricle
  - Ataxia
  - neck pain
  - cranial nerve palsies
- Supratentorial ependymomas:
  - Headaches
  - Seizures
  - location-dependent focal neurological deficits
- Spinal cord ependymomas:
  - back pain
  - lower extremity weakness
  - bowel and bladder dysfunction

Ependymoma: Diagnosis

- Every patient with suspected ependymoma will get imaging of the whole brain and spinal cord, likely both CT and MRI
- If feasible, CSF evaluation is conducted
- Molecular studies are done to look for distinctive patterns of gene expression (especially for posterior fossa ependymomas to determine if they are in group A or B) and gene fusion status for supratentorial ependymomas (YAP1 and ZFTA)
  - Group A (PF-EPN-A): primarily occurs in young children; increased occurrence of 1q gain which confers a very poor prognosis; pts with a 1q gain and 6q loss have an even poorer prognosis
  - Group B (PF-EPN-B): primarily occurs in older children and adults; more favorable prognosis; if a 13q loss is seen then its likely a poor prognosis
  - ZFTA fusion-pos (ST-EPN-ZFTA): favorable survival for pts who undergo a complete resection and post-op XRT; less favorable if they forego XRT
  - YAP1 fusion-pos (ST-EPN-YAP1): favorable prognosis, with 5-year survival rates approaching 100%
- Spinal ependymomas, especially when MYCN-amplified (ST-EPN-MYCN) are rare and aggressive, predominantly affecting young adults, and often present with leptomeningeal dissemination and a high rate of recurrence.

Coding Ependymoma Histology

- 9383/1
  - Subependymoma
- 9391/3
  - Supratentorial ependymoma, NOS (C71._)
  - Posterior fossa ependymoma, NOS (C71._)
  - Spinal ependymoma, NOS (C72.0)
- 9394/3
  - Myxopapillary ependymoma
- 9396/3
  - Supratentorial ependymoma, ZFTA fusion-positive
  - Supratentorial ependymoma, YAP1 fusion-positive
  - Posterior fossa group A (PFA) ependymoma
  - Posterior fossa group B (PFB) ependymoma
  - Spinal ependymoma, MYCN-amplified (C72.0)
  - Ependymoma, RELA fusion-positive (old term for ZFTA fusion-pos)

Ependymoma: Treatment

- Treatment depends on location of tumor, morphology, and grade.
- Typically, the best prognosis comes from a total resection of the tumor and adjuvant XRT.
- Resection, either gross total or subtotal will most likely be recommended. Subtotal resections usually mean a poorer prognosis.
- Proton XRT likely. It is not necessary to treat the entire CNS (whole brain and spine) because these tumors usually recur initially at the local site, although posterior fossa ependymomas may disseminate at recurrence, particularly in tumors with 1q gain
  - Lower doses of radiation or chemotherapy-only protocols also confer a poor prognosis.
- There is currently no evidence that systemic treatment is able to successfully treat these tumors, although one phase 3 study was just completed that trialed maintenance chemo vs observation post XRT and surgery.

PDCS Data Items: Ependymoma

Pediatric Primary Tumor
Coded based on a single tumor confined to primary site or crossing the midline without extension to adjacent structures

Pediatric Regional Nodes - N/A (code 888)

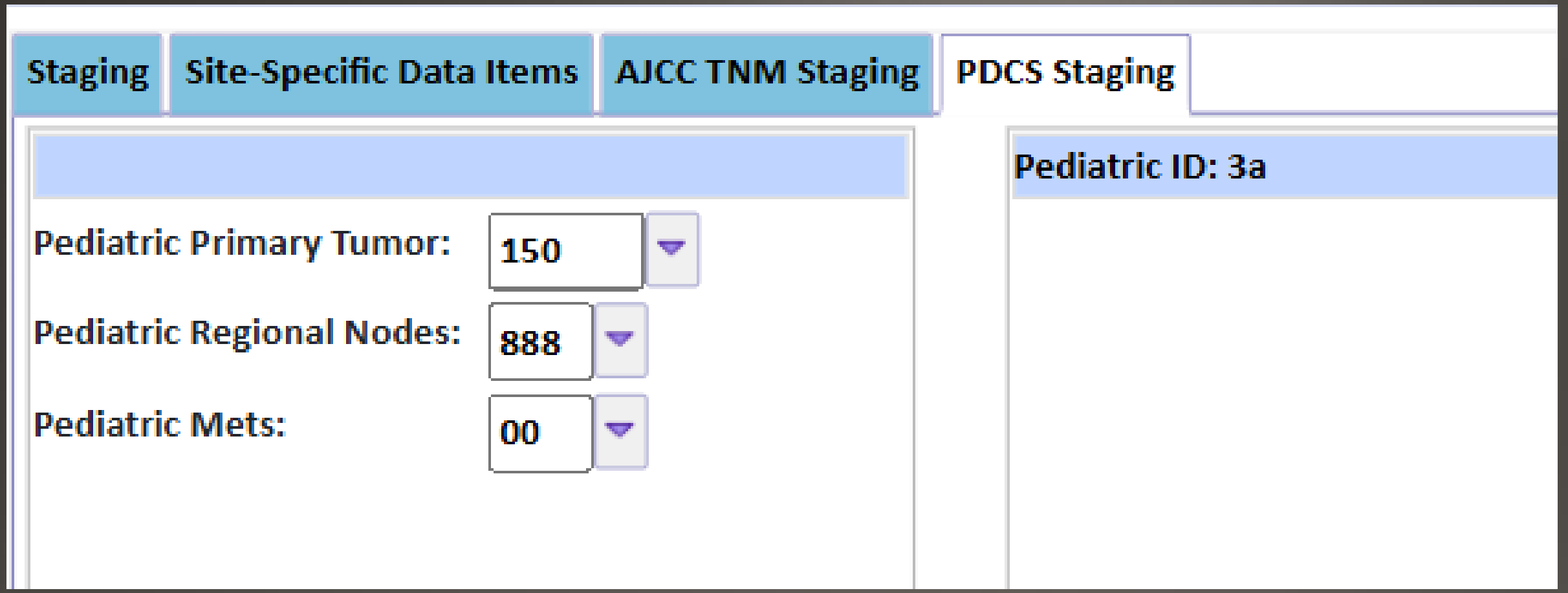
Pediatric Mets
Coded based on tumor extension to adjacent structures/sites, tumor cells in CSF, drop mets or other mets in CSF, extra-neural mets

SSDI - none

Ependymoma Case Scenario

- Imaging: MRI of brain/spine shows 5.6cm spinal cord lesion from C3-C6, favors ependymoma, no further spread
- No other involvement noted
- Diagnosis: Spinal ependymoma, WHO grade 2, GTR confirmed.

Data Item	Value	Comments
Pediatric Primary Tumor	150	All sites • Single tumor confined to the primary site with no invasion or seeding to other structures • Confined to site of origin, NOS • Localized, NOS
Pediatric Regional Nodes	888	Default code for CNS tumors
Pediatric Mets	00	Per imaging there is no other involvement noted



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Medulloblastomas

What are Medulloblastomas?

- Medulloblastomas make up the vast majority of pediatric embryonal tumors.
- Can result from hereditary cancer predisposition syndromes in approximately 5% of patients. Examples include:
  - Turcot syndrome
  - Rubinstein-Taybi syndrome
  - Gorlin syndrome (also known as basal cell nevus syndrome or nevoid basal cell carcinoma syndrome
  - Li-Fraumeni syndrome
  - Fanconi anemia
- They most often arise in the posterior fossa where they constitute approximately 40% of all posterior fossa tumors. In approximately 80% of children, medulloblastomas arise in the region of the fourth ventricle.

Medulloblastoma: How Does it Present?

- Relatively abrupt onset of headaches, especially in the morning on waking
- Nausea and/or vomiting
- Lethargy
- Ataxia
- Some degree of nystagmus
- Papilledema
- More laterally positioned medulloblastomas of the cerebellum may not result in hydrocephalus and, because of their location, are more likely to result in:
  - lateralizing cerebellar dysfunction (appendicular ataxia) manifested by:
    - unilateral dysmetria
    - unsteadiness
    - weakness of the sixth and seventh nerves on the same side as the tumor.
- In infants, the presentation of medulloblastoma is more variable and may include the following:
  - Nonspecific lethargy
  - Psychomotor delays
  - Loss of developmental milestones
  - Feeding difficulties
  - Increase in head circumference

Medulloblastoma: Diagnosis

- Every patient with suspected medulloblastoma will get imaging of the whole brain and spinal cord, likely both CT and MRI
- If feasible, CSF evaluation is conducted
- A biopsy may be done prior to surgery if more easily accessible

Coding Medulloblastoma

- 9470/3
  - Medulloblastoma NOS
  - Classic medulloblastoma
  - Medulloblastoma, histologically defined
- 9474/3
  - Anaplastic/large cell medulloblastoma
- 9471/3
  - Medulloblastoma described as one of the following
    - Desmoplastic nodular medulloblastoma
    - Medulloblastoma with extensive nodularity
    - Medulloblastoma, SHH-activated and TP53- wildtype
- 9477/3
  - Medulloblastoma non-WNT/non-SHH
  - Medulloblastoma group 3
  - Medulloblastoma group 4
- 9476/3
  - Medulloblastoma SHH-activated and TP53-mutant
- 9475/3
  - Medulloblastoma WNT-activated

Medulloblastoma: Treatment

- Treatment depends on location of tumor, morphology, and grade.
- Resection, either gross total or subtotal will most likely be recommended.
  - A significant number of patients with medulloblastoma develop cerebellar mutism syndrome (also known as posterior fossa syndrome) post-surgery
- Proton or Photon XRT is likely.
  - Radiation has a profound effect on children of all ages, but especially for children under 3 years. Debilitating effects on neurocognitive development, growth, and endocrine function have been frequently observed.
- Chemotherapy is usually administered during and after radiation therapy.
- For high risk, persistent and/or recurrent disease, PBSCTs may be given

CNS - Medulloblastoma

Pediatric Primary Tumor
Coded based on a single tumor confined to primary site or crossing the midline without extension to adjacent structures

Pediatric Regional Nodes - N/A (code 888)

Pediatric Mets
Coded based on tumor cells in CSF, intracranial spread beyond a single lesion, drop mets or other mets in CSF, extra neural-mets, spinal mets. This coding system is based on the Chang M definition for Metastases within the CNS at diagnosis.

SSDI - none

Chang M Definition of Mets

Chang M definition of metastases

M0: No visible disease on imaging (MRI brain and
spine) beyond primary site of disease AND no tumor cells into the cerebrospinal fluid (CSF)

M1: Tumor cells in the CSF

M2: Visible metastasis in brain

M3: Visible metastasis in spine OR visible metastasis in cervicomedullary (junction)

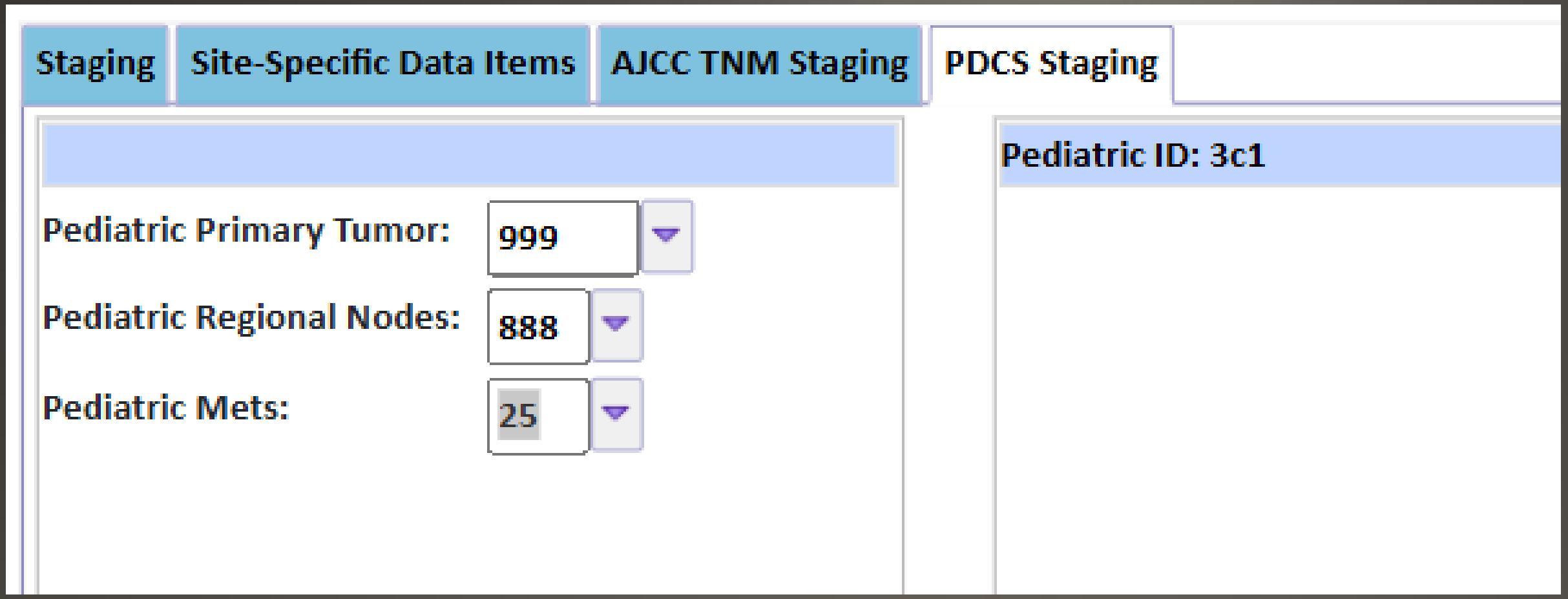
M4: Metastasis outside of the central nervous system

M Category (AJCC)	*Modified Chang*	*M Criteria (AJCC)*
cM0	M0	No distant metastasis
cM1		Distant metastasis
cM1b	M2	Intracranial spread beyond primary site
cM1c	M3	Gross spinal subarachnoid seeding on MRI
cM1d	M4	Metastasis outside CNS (bone marrow, lungs)
pM1		Microscopic confirmation of distant metastasis
pM1a	M1	Microscopic confirmation of tumor cells present in CSF by cytology
pM1b	M2	Microscopic confirmation of intracranial spread beyond primary site
pM1c	M3	Microscopic confirmation of gross spinal subarachnoid seeding
pM1d	M4	Microscopic confirmation of metastasis outside CNS (bone marrow, lungs)

Medulloblastoma Case Scenario

- Imaging: 2.2 cm midline mass in posterior fossa, encroaching 4th ventricle & demonstrating restricted diffusion. Findings most concerning for medulloblastoma, possibly group 4.
- Diagnosis: Resection, medulloblastoma, non-WNT/non-SHH (group 4 by NDA methylation profile), WHO grade 4.

Data Item	Value	Comments
Pediatric Primary Tumor	999	• Single tumor with extension to an adjacent site
Pediatric Regional Nodes	888	Default code for CNS tumors
Pediatric Mets	25	Intracranial spread beyond a single lesion • All sites: bone, major blood vessels, meninges, nerves, tumor invades or encroaches upon ventricular system



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Pediatric Treatment Considerations

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?

Brain Tumors: Intuitive Abstracting

DIAGNOSTIC WORKUP

- **Workup imaging will most likely include:** CT/MRI of brain/spine/orbit
- **Workup labs will most likely include:** if suspicion of CSF involvement, a spinal tap may be done. Cytogenetics are also commonly done and often included on pathology reports. Use the "integrated diagnosis" on path reports for histology codes!

Brain Tumors: Intuitive Abstracting

- **Metastatic disease**
    - This means only CT/MRI of brain/spine/orbit, possibly PET/CT, but likely not
    - Treatment will likely include radiation – proton OR photon
    - Could also be LEPTOMENINGEAL and/or SPINAL (drop mets)

- **Steroids/Decadron**
    - Steroids will likely be given post biopsy or surgery, but it is NOT coded as treatment.
    - It is a standard treatment after brain surgery to *reduce cerebral edema.*

- **Surgical Codes**
    - Removal of the **TUMOR** = codes 21 (STR in brain) or 30 (GTR in brain) or 22 (STR/GTR in spinal cord)
    - Removal of the **LOBE** = codes 40 (STR of lobe) or 55 (GTR/lobectomy)

CNS Biopsy Codes

Open Brain Biopsy

- Coded as 02
- Counts as a diagnostic procedure
- Typical incisional biopsy through a small opening in the skull that accesses brain tissue directly

Stereotactic Needle Biopsy

- Coded as A200
- Counts as a surgical procedure (per SEER) if it's the most advanced procedure done to the tumor
- Done in cases where the tissue is not easily accessible, like the brainstem, or deeper brain locations like the thalamus
- Must specifically be stated as stereotactic in the operative report

Tumor Resections

Subtotal Resection

- Partial or "near total" removal of the TUMOR
  - Brain = A210
  - Spinal cord = A220

Gross Total Resection

- Removal of the entire TUMOR
  - Brain = A300
  - Spinal cord = A220

Lobectomies

Subtotal Lobectomy

- Partial or "near total" removal of the LOBE
  - Brain = A400

Gross Total Lobectomy

- Removal of the entire LOBE
  - Brain = A550

POP QUIZ #1!

A nine-year-old male was given decadron after having a gross total resection of a temporal lobe astrocytoma. The abstractor coded both the GTR and the decadron on the treatment page.

CORRECT or **INCORRECT**

POP QUIZ #1!

A nine-year-old male was given decadron after having a gross total resection of a temporal lobe astrocytoma. The abstractor coded both the GTR and the decadron on the treatment page.

CORRECT or **INCORRECT**

POP QUIZ #2

Patient came to a SEER reporting hospital in February 2024 due to headaches, right sided facial droop and gait instability. Imaging was done that showed a 2cm mass in the pons. A stereotactic incisional biopsy of the tumor was done that confirmed diffuse midline glioma, H3K27m+. No further surgery noted.

What code will you choose for the biopsy?

A. 02 – incisional biopsy

B. A200 – stereotactic biopsy of brain tumor

POP QUIZ #2

Patient came to a SEER reporting hospital in February 2024 due to headaches, right sided facial droop and gait instability. Imaging was done that showed a 2cm mass in the pons. A stereotactic incisional biopsy of the tumor was done that confirmed diffuse midline glioma, H3K27m+. No further surgery noted.

What code will you choose for the biopsy?

A. 02 – incisional biopsy

B. A200 – stereotactic biopsy of brain tumor

POP QUIZ #3

Patient came to reporting hospital in March 2024 with complaints of intermittent headaches and N/V in the mornings. Imaging showed a 1cm mass in the left frontal lobe abutting the meninges. It was biopsied and confirmed to be a pilocytic astrocytoma. A gross total resection of the mass was done in April 2024. Follow up scans confirmed no recurrence or residual tumor.

Which surgical code applies to this scenario?

A. A210 – subtotal resection of the brain tumor
B. A220 – gross total resection of the spinal tumor
C. A300 – gross total resection of the brain tumor
D. A400 – subtotal resection of the lobe
E. A550 – gross total resection of the lobe

POP QUIZ #3

Patient came to reporting hospital in March 2024 with complaints of intermittent headaches and N/V in the mornings. Imaging showed a 1cm mass in the left frontal lobe abutting the meninges. It was biopsied and confirmed to be a pilocytic astrocytoma. A gross total resection of the mass was done in April 2024. Follow up scans confirmed no recurrence or residual tumor.

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POP QUIZ #4

3-year-old female with history of epilepsy secondary to structure abnormality in left temporal lobe was being followed at the reporting hospital. The surgical operative note from October 2022 states that about 70% of the left temporal lobe was resected to remove the diseased tissue due to this condition and an incidental ganglioma was discovered in the resected tissue.

Which surgical code applies to this scenario?

A. A210 – subtotal resection of the brain tumor
B. A300 – gross total resection of the brain tumor
C. A400 – subtotal resection of the lobe
D. A550 – gross total resection of the lobe
E. None of the above – surgery not related to ganglioma

POP QUIZ #4

3-year-old female with history of epilepsy secondary to structure abnormality in left temporal lobe was being followed at the reporting hospital. The surgical operative note from October 2022 states that about 70% of the left temporal lobe was resected to remove the diseased tissue due to this condition and an incidental ganglioma was discovered in the resected tissue.

Which surgical code applies to this scenario?

A. A210 – subtotal resection of the brain tumor
B. A300 – gross total resection of the brain tumor
C. A400 – subtotal resection of the lobe
D. A550 – gross total resection of the lobe
E. None of the above – surgery not related to ganglioma

In Conclusion….

Resources

- The DIPG/DMG Resource Network
- Toronto Childhood Cancer Staging Guidelines
- ICCC Pediatric Groups
- Pediatric Hospital CTRs Facebook Group
- PDQ Cancer Information Summaries: Pediatric Treatment
- NAACCR Pediatric Resources
- Pediatric Data Collection System (PDCS), Appendix 1

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