Data Collection of Primary Central Nervous System (CNS) Tumors

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Portions of this presentation are based on non-malignant CNS tumor data collection rules adopted by the North American Association of Central Cancer Registries (NAACCR) Uniform Data Standards Committee - June 2003.

Part I

- Rationale
- History
- Definition of Reportable Cases
- Casefinding
- Anticipated Impact on Registries

Rationale for Non-malignant CNS Tumor Surveillance and Registration

- Non-malignant CNS tumors cause disruption in normal function similar to that caused by malignant CNS tumors.

- Location of a CNS tumor is as important as tumor behavior (benign or malignant) to morbidity and mortality.

History 1992 -1996

- 1992 Central Brain Tumor Registry of the United States (CBTRUS) formed to report population-based data on primary benign, borderline, and malignant CNS tumors.

- 1996 National Coordinating Council on Cancer Surveillance (NCCCS) formed Brain Tumor Working Group (BTWG) to explore the feasibility of registering non-malignant CNS tumors

History 1998

- BTWG forwarded four recommendations to the NCCCS
- NCCCS
  Accepted recommendations 1 and 2
  Deferred recommendations 3 and 4

BTWG Recommendations (1)

1. The following standard definition is to be used for collecting precise data for all primary intracranial and CNS tumors:

   Primary intracranial and CNS tumors are all primary tumors occurring in the following sites, irrespective of histologic type or behavior:

- Brain
- Spinal cord
- Pituitary gland
- Craniopharyngeal duct
- Meninges
- Cauda equina
- Pineal gland
- Cranial nerves and other parts of the CNS.

BTWG Recommendations (2)

2. Develop a standard site and histology definition for tabulating estimates of CNS tumors to allow comparability of information across registries.

3. All registries, both hospital- and population-based, should collect data on primary CNS tumors.

BTWG Recommendations (3)

4. Develop training for reporting and tabulating primary intracranial and CNS tumors, and develop computerized edit-checking procedures.

History 2000

International Classification of Diseases for Oncology 3rd Edition (ICD-O-3) and World Health Organization (WHO) 2000 Brain Tumor Classification are compatible.

November

- Consensus conference on brain tumor definition convened. Group agrees to:
  - Site definition as in Recommendation 1.
  - Need to develop a standard site and histology definition based on the SEER site and histology validation list.

History 2001-2002

2001 NCCCS

- Accepted Recommendations 1 and 2 as completed.
- Reconvened the BTWG to work on Recommendations 3 and 4.

2002 NAACCR established subcommittee of Registry Operations Committee to:

- Identify changes needed in registry operations for inclusion of non-malignant CNS tumors.
- October: Benign Brain Tumor Cancer Registries Amendment Act (Public Law 107-260) signed by President Bush.

Reportable Brain-Related Tumors (1)

Public Law 107-260 requires reporting of brain-related tumors.

- The term "brain-related tumor" means a listed primary tumor (whether malignant or benign) occurring in any of the following sites:

  (I) The brain, meninges, spinal cord, cauda equina, a cranial nerve or nerves, or any other part of the CNS.

  (II) The pituitary gland, pineal gland, or craniopharyngeal duct.

Reportable Brain-Related Tumors (2)

Brain

- Cerebrum (C71.0)
- Frontal lobe (C71.1)
- Temporal lobe (C71.2)
- Parietal lobe (C71.3)
- Occipital lobe (C71.4).

Reportable Brain-Related Tumors (3)

Brain (continued)

- Ventricle (C71.5)
- Cerebellum (C71.6)
- Brain stem (C71.7)
- Overlapping lesion of the brain (C71.8)
- Brain NOS (C71.9)

Reportable Brain-Related Tumors (4)

- Meninges
  - Cerebral meninges (C70.0)
  - Spinal meninges (C70.1)
  - Meninges NOS (C70.9)
- Spinal cord (C72.0)
- Cauda equina (C72.1)

Reportable Brain-Related Tumors (5)

- Cranial nerves
  - Olfactory nerve (C72.2)
  - Optic nerve (C72.3)
  - Acoustic nerve (C72.4)
  - Cranial nerve NOS (C72.5)

Reportable Brain-Related Tumors (6)

- Other CNS (C72.8, C72.9)
- Pituitary gland (C75.1)
- Craniopharyngeal duct (C75.2)
- Pineal gland (C75.3)

For the sites described, benign, borderline, and malignant tumors are reportable for cases diagnosed on or after January 1, 2004.

History 2003

2003 SEER-supported registries and COC-approved hospital cancer registries will also report non-malignant CNS tumors diagnosed on or after January 1, 2004.

Impact of Collecting Data on Non-malignant CNS Tumors (1)

Annual increase in number of cases estimated by doubling the number of malignant CNS cases diagnosed in the same year

Increase in hospital registry case load will depend on the type of hospital:

- Community hospitals with small or no neurology service will likely experience a small increase in case load.
- Hospitals with a large neurology service will likely experience a larger increase.

Impact of Collecting Data on Non-malignant CNS Tumors (2)

Central registry case load is estimated to increase by 1%.

In 2002, 21 state cancer registries collected data on non-malignant CNS tumors:

- Minimal impact if registry's definition for brain-related sites does not change.

Impact of Collecting Data on Non-malignant CNS Tumors (3)

Central registries adding non-malignant CNS tumors to reportable case definition may have to change state reporting law if law does not allow for collection of data on non-malignant cases.

Impact of Collecting Data on Non-malignant CNS Tumors (4)

All cancer registries must:

- Have the same definition for brain-related tumors.
- Implement data edits created for non-malignant CNS tumors.
- Report rates for these tumors.

Case-finding (1)

Additional or expanded case-finding mechanisms:

- Pathology
- Radiology
- Treatment facilities:
  - Radiation oncology centers and departments
  - Gamma or cyber knife center.

Case-finding (2)

- Disease indices
- Surgery logs
- Diagnostic imaging
- Radiation oncology
- Neurology clinics
- Medical oncology
- Autopsy reports.

Case-finding Sources

- Free-standing radiation therapy centers
- Free-standing Magnetic Resonance Imaging (MRI) centers
- Free-standing gamma or cyber knife centers
- Free-standing oncology centers
- Data exchange with other central registries
- Death clearance process

ICD-9-CM Codes for Case-finding

Table 1: ICD-9-CM Casefinding Codes for Benign and Borderline Intracranial and CNS Tumors	
ICD-9-CM Code	**Description of Neoplasm**
225.0	Benign neoplasm of brain
225.1	Benign neoplasm of cranial nerves
225.2	Benign neoplasm of cerebral meninges; cerebral meningioma
225.3	Benign neoplasm of spinal cord, cauda equina
225.4	Benign neoplasm of spinal meninges; spinal meningioma
225.8	Benign neoplasm of other specified sites of nervous sytem
225.9	Benign neoplasm of nervous system, part unspecified
227.3	Benign neoplasm of pituitary, craniopharyngeal duct, craniobuccal pouch, hypophysis, Rathke's pouch, sella turcica
227.4	Benign neoplasm of pineal gland, pineal body
237.0	Neoplasm of uncertain behavior of pituitary gland and craniopharyngeal duct
237.1	Neoplasm of uncertain behavior of pineal gland
237.5	Neoplasm of uncertain behavior of brain and spinal cord
237.6	Neoplasm of uncertain behavior of meninges: NOS, cerebral, spinal
237.70	Neurofibromatosis, Unspecified von Recklinghausen's Disease
237.71	Neurofibromatosis, Type One von Recklinghausen's Disease
237.72	Neurofibromatosis, Type Two von Recklinghausen's Disease
237.9	Neoplasm of uncertain behavior of other and unspecified parts of nervous system; cranial nerves

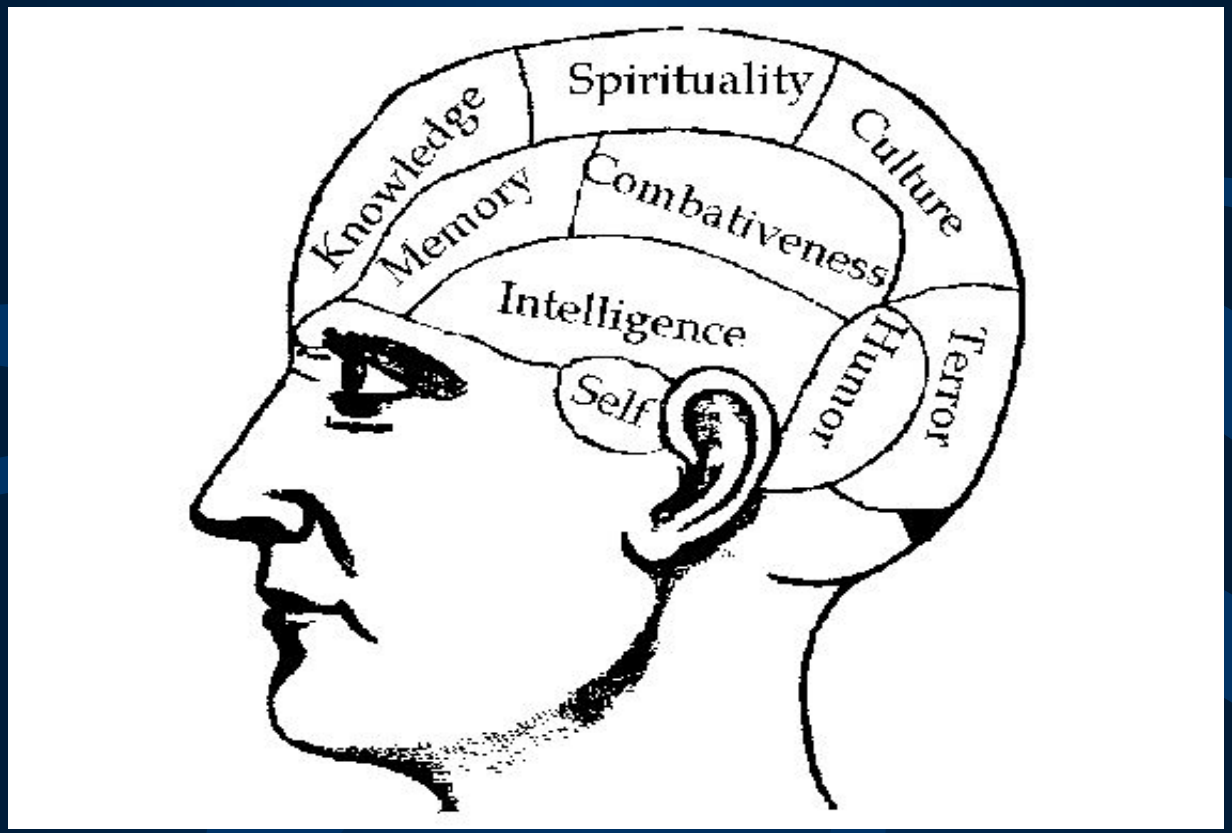
Unusual and Ambiguous Terminology

- If the final pathologic diagnosis is a CNS "neoplasm" or "mass", an ICD-O-3 histology code must exist for the case to be reportable.
- Hypodense mass or cystic neoplasm are not reportable, even for CNS sites.
- A benign meningioma with a skull site should be coded to the cerebral meninges (C70.1).

Part II

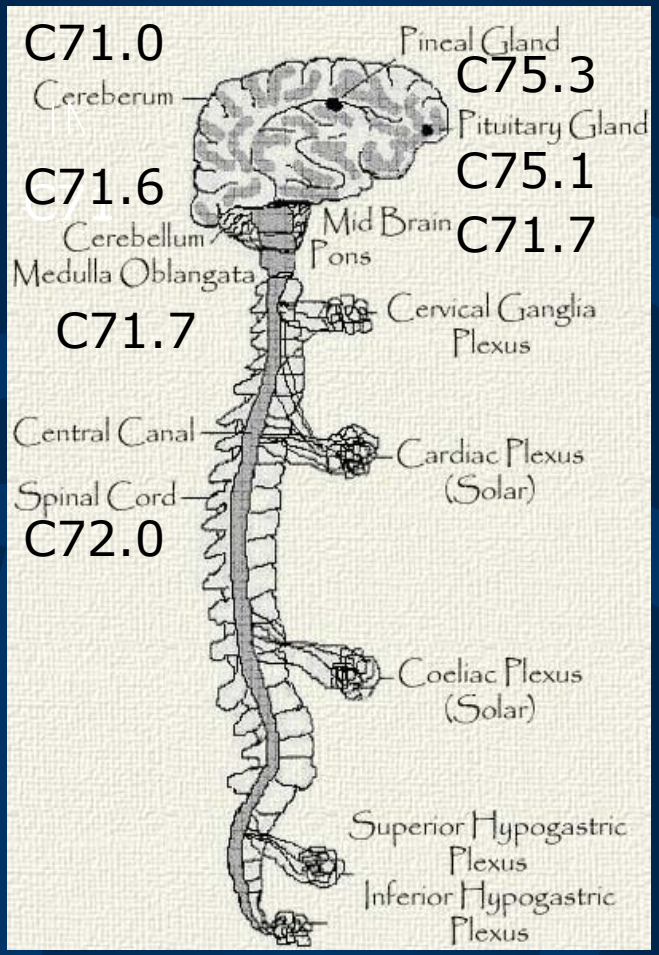
- CNS Anatomy and Function
- Histologies and Primary Sites
- Grading Systems and Coding Grade

CNS Functional Anatomy



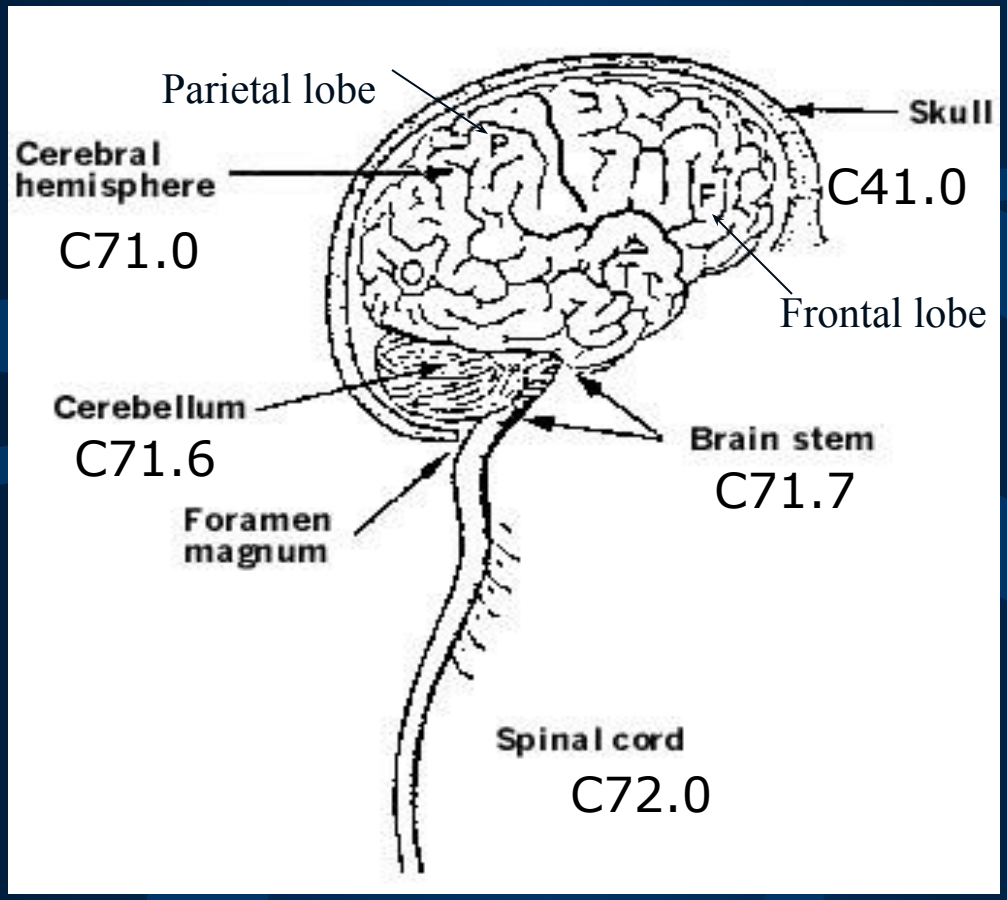
Source: URL: www.solinas.com/solinas/brain.html accessed 7/18/03.

CNS Anatomy



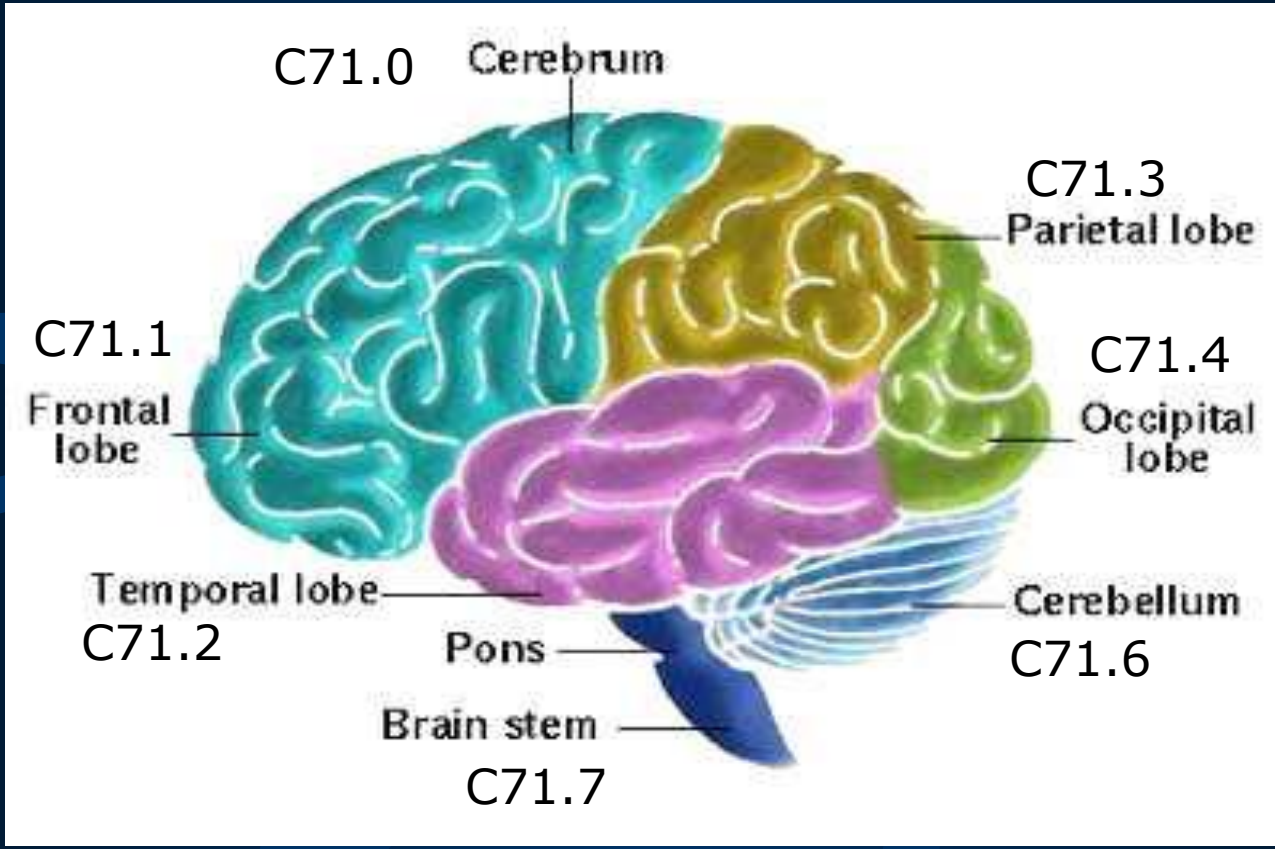
Source: URL: www.universalpeace.ca/principles.htm accessed 7/18/03.

Intracranial Sites



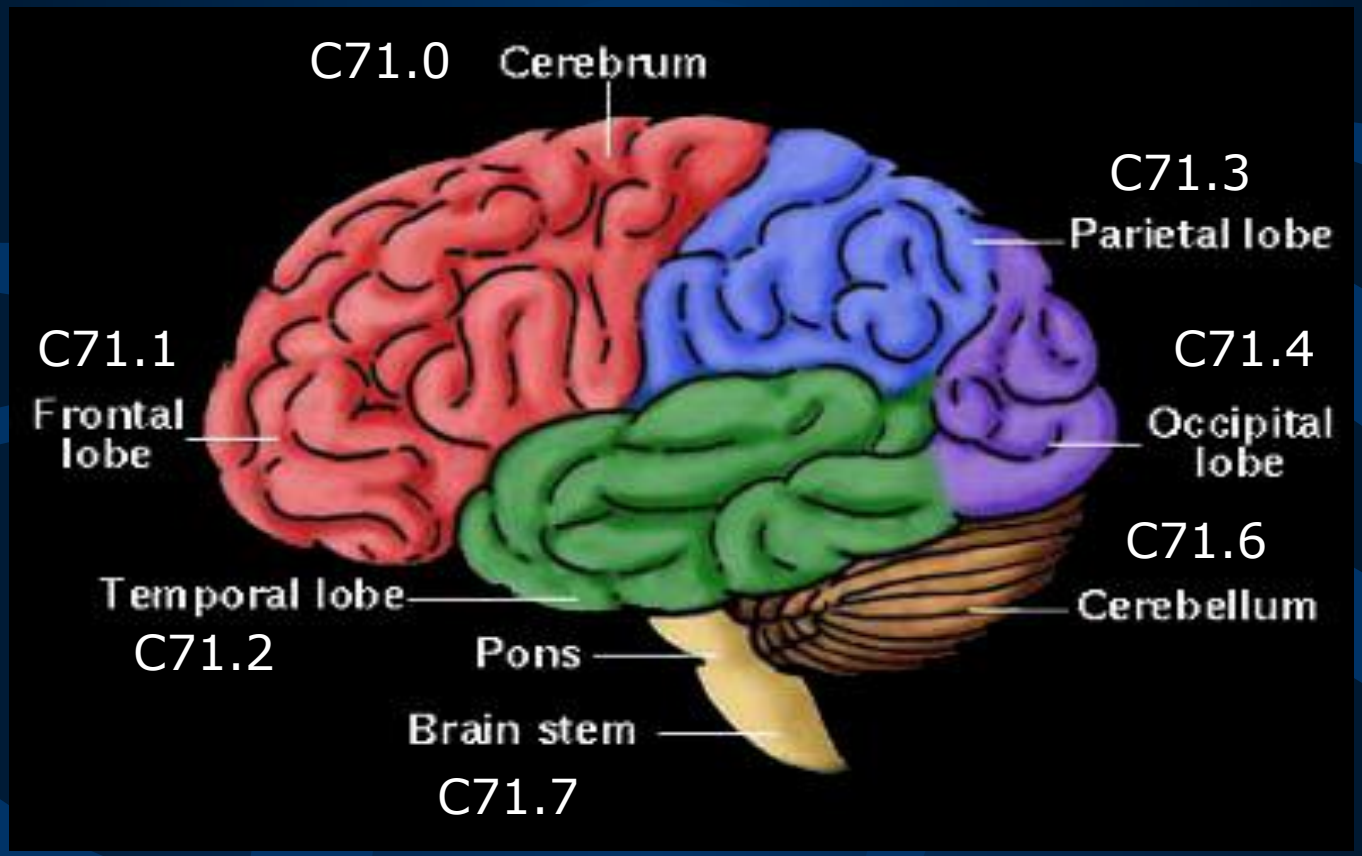
Source: URL: mscenter.ucsf.edu/faq.htm accessed 7/18/03.

Cerebrum



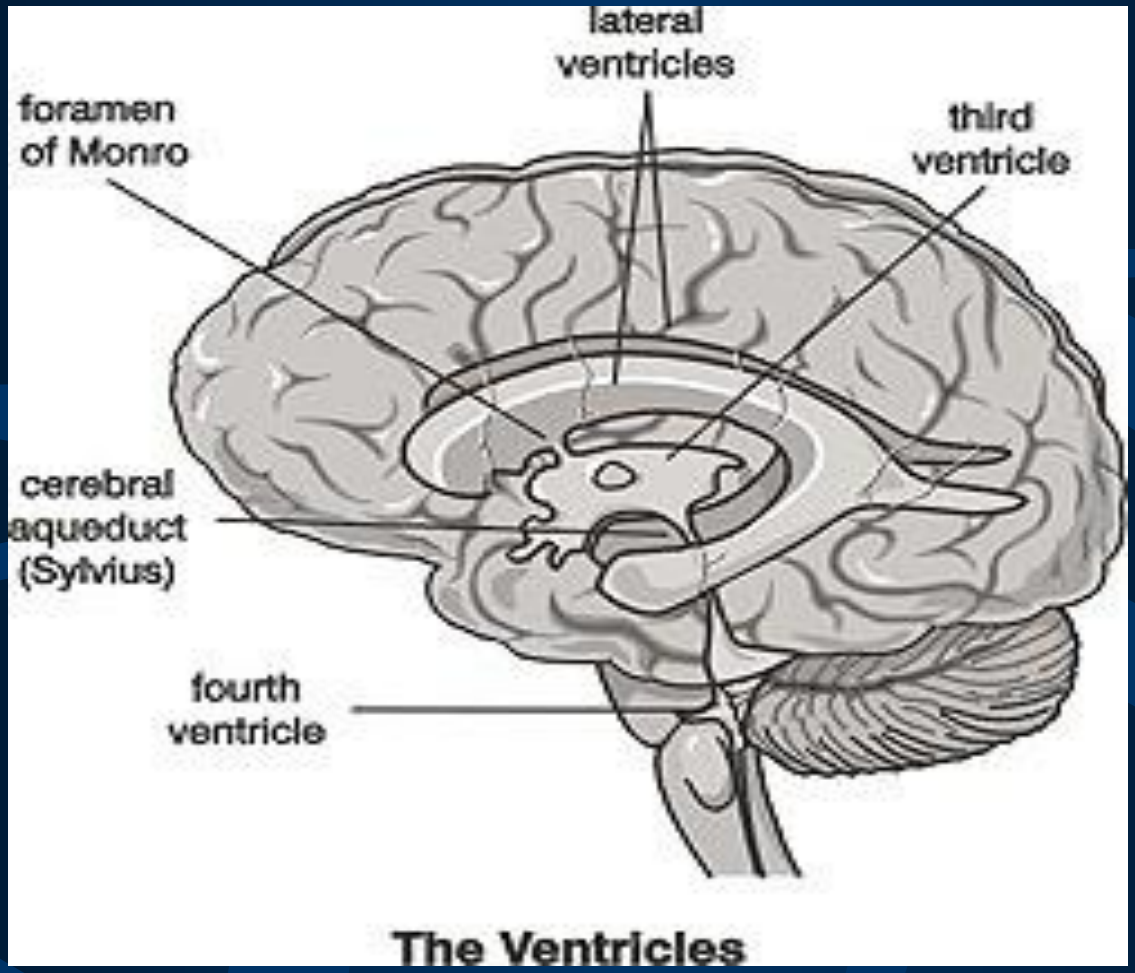
Source: URL: www.sciencebob.com/lab/bodyzone/brainprint.html Accessed 7/18/03.

Cerebellum and Brain Stem



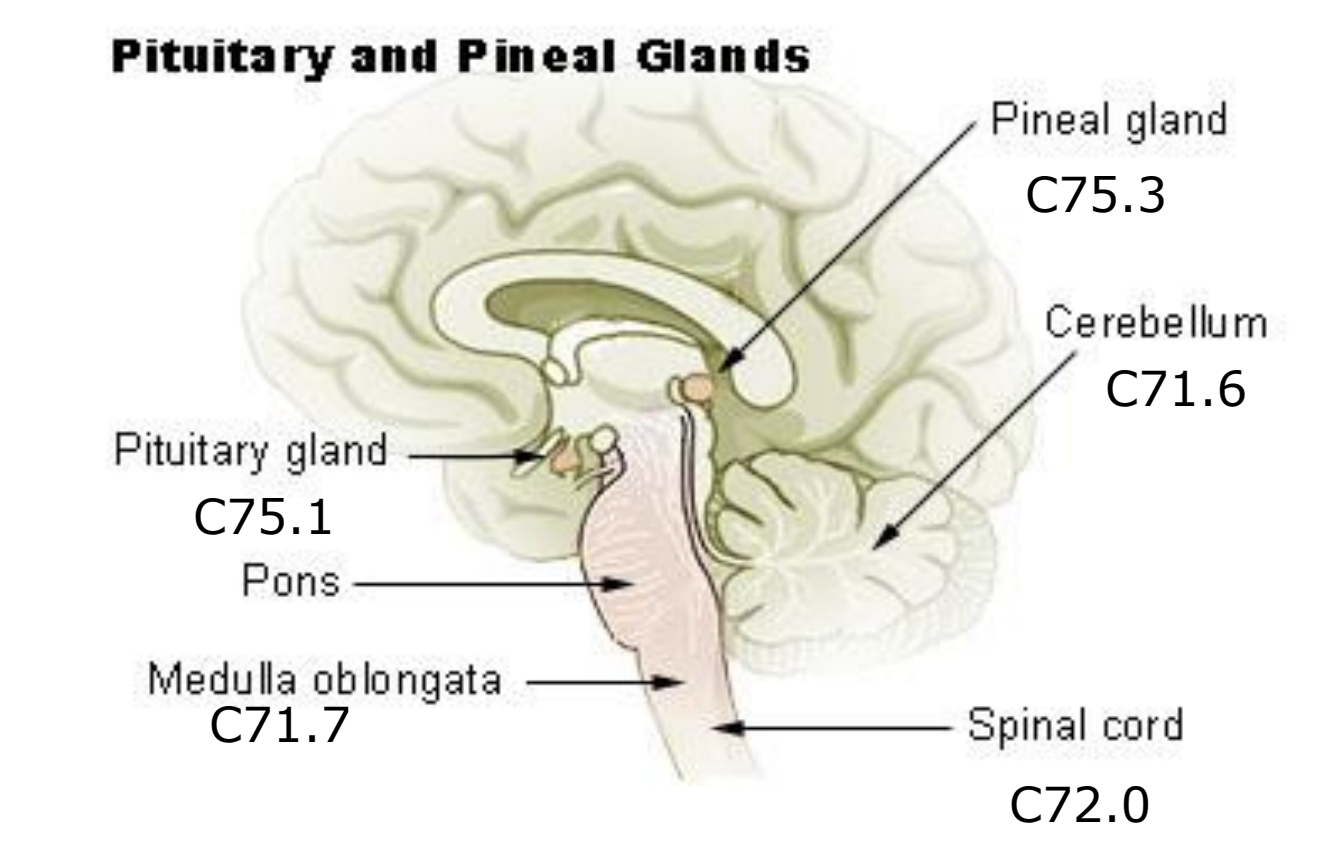
URL: www.sciencebob.com/lab/bodyzone/brain.html 7/18/03

The Ventricular System



http://www.abta.org/primer2.htm

Pineal and Pituitary Glands



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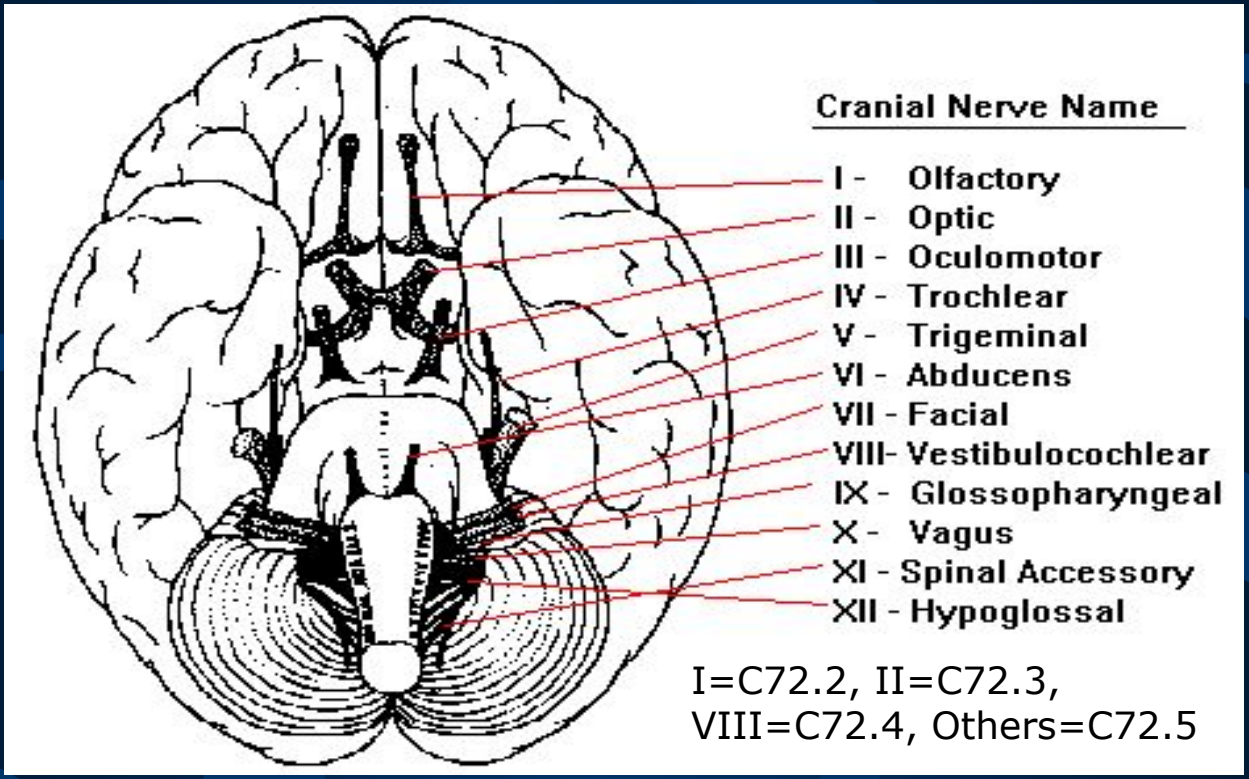
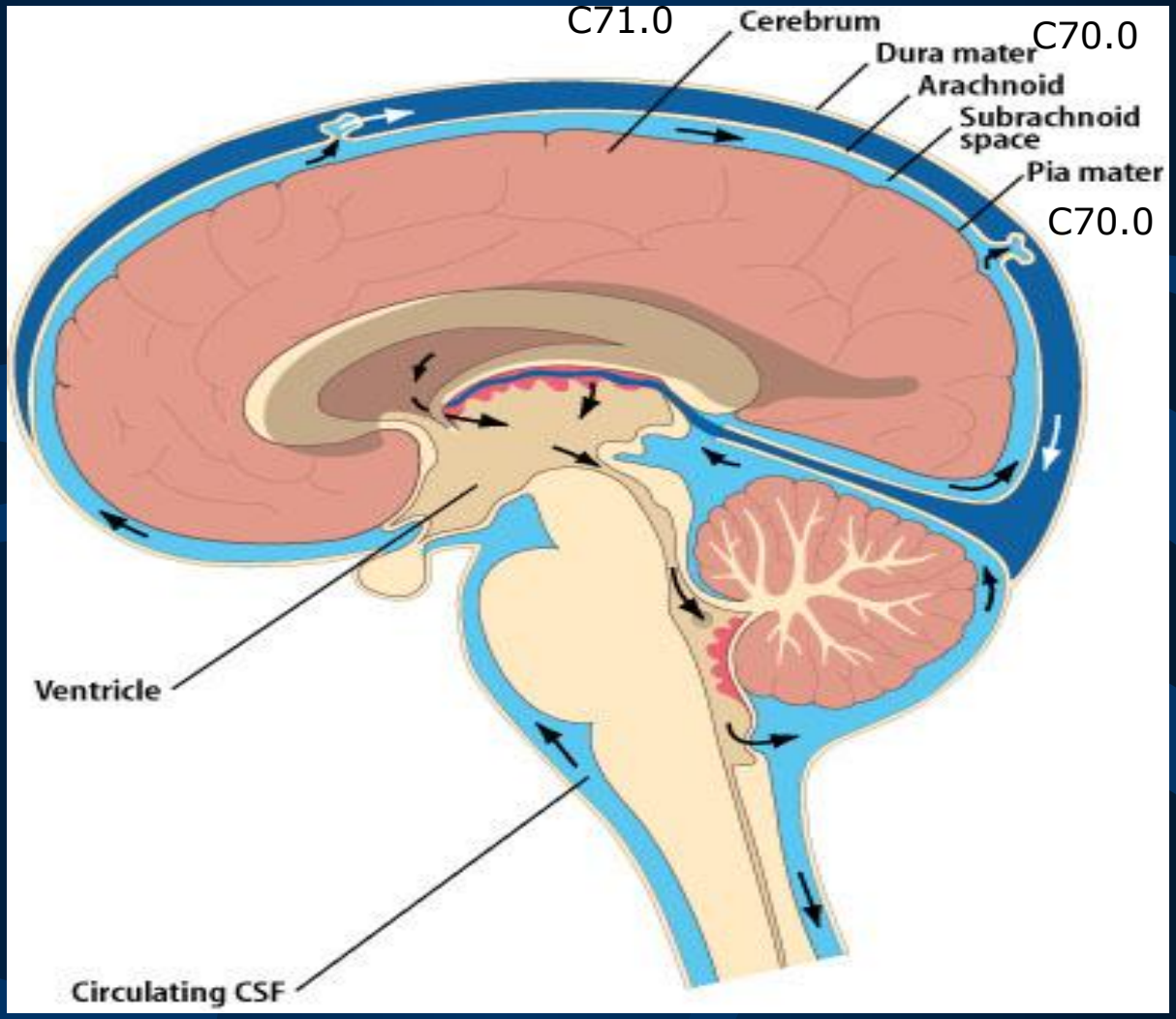
Accessed 7/18/03.

Cranial Nerves



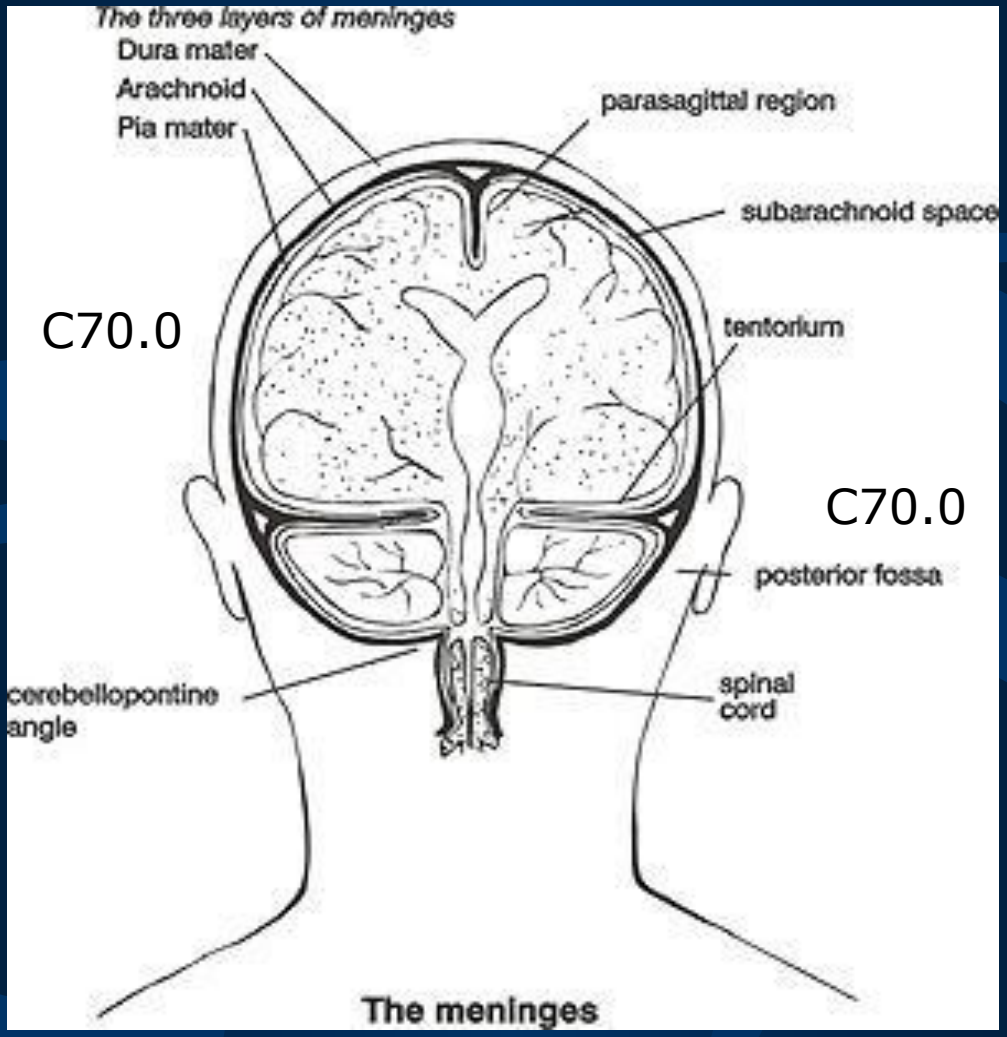
Source: URL: faculty.washington.edu/chudler/cranial.html Accessed 7/18/03.

Meninges



Source: URL: www.cardioliving.com/consumer/Stroke/Hemorrhagic_Stroke.sht Accessed 7/18/03.

Tentorium

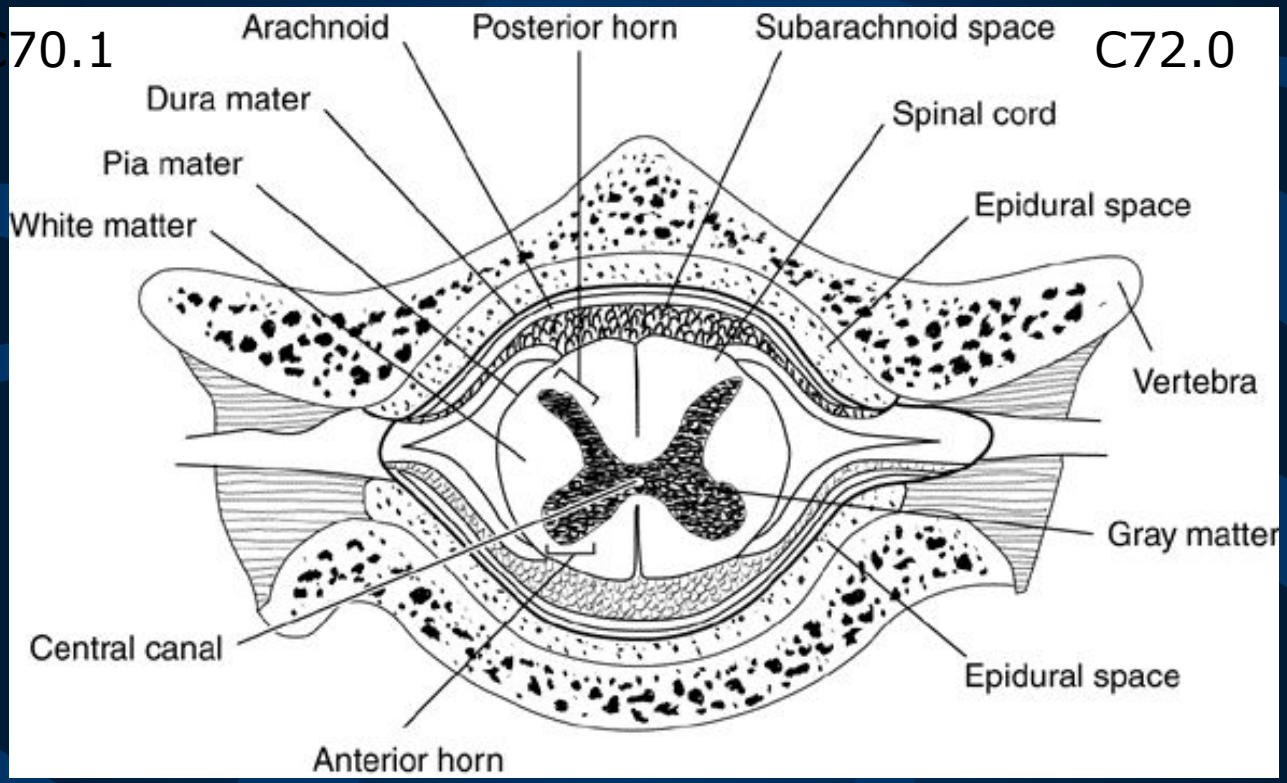


C70.0

C70.0

Source: URL: neurosurgery.mgh.harvard.edu/abta/primer.htm Accessed 7/18/03.

Spinal Cord



Source: URL: www.merck.com/pubs/mmanual/figures/182fig1.htm Accessed 7/18/03

Cellular Classification

Neuroepithelial tumors

- Astrocytomas
- Oligodendrogliomas
- Ependymomas
- Pineal parenchymal tumors

Other CNS tumors

- Sellar tumors
- Hematopoetic tumors
- Germ cell tumors
- Meningiomas
- Tumors of cranial nerves

Glial Tumors (1)

Glial tissue: supportive tissue of brain made up of astrocytes and oligodendrocytes

Glial tumors assigned ICD-O-3 histology codes from glioma series:

- Codes 938 through 948.

Glial Tumors (2)

Astrocytic tumors

- Noninfiltrating
  - Juvenile pilocytic (M9421)
  - Subependymal (M9383)
- Infiltrating
  - Well-differentiated mildly and moderately anaplastic astrocytomas (M9401)
  - Anaplastic astrocytomas
  - Glioblastoma multiforme (M9440)
  - Brain stem gliomas (M9380)

Glial Tumors (3)

Ependymal tumors

- Myxopapillary and well-differentiated ependymomas (M9394)
- Anaplastic ependymomas (M9392)
- Ependymoblastomas (M9392)

Oligodendroglial tumors

- Well-differentiated oligodendrogliomas (M9450)
- Anaplastic oligodendrogliomas (M9451)

Glial Tumors (4)

Mixed tumors

- Mixed astrocytoma-ependymomas
- Mixed astrocytoma-oligodendrogliomas
- Mixed astrocytoma-ependymoma-oligodendrogliomas

Other gliomas

- Ganglioneuromas (M9490)
- Optic nerve gliomas

Non-Glial Tumors (1)

Pineal region tumors

- Parenchymal tumors
  - Pineocytomas (M9361)
  - Pineoblastomas (M9362)
  - Pineal astrocytomas (M9400)
- Germ cell tumors
  - Germinomas (M9064)
  - Embryonal carcinomas (M9070)
  - Choriocarcinomas (M9100)
  - Teratomas (M9080)

Non-Glial Tumors (2)

Meningiomas

- Meningioma: Benign (M953_)
- Malignant meningiomas
  - Anaplastic meningioma
  - Hemangiopericytoma (M9150)
  - Papillary meningioma (M9538)

Choroid plexus tumors

- Choroid plexus papilloma (M9390)
- Choroid plexus carcinoma
- Choroid plexus meningioma (M9538)

Other CNS Tumors (1)

- Craniopharyngiomas (M9350)
  - Rathke pouch tumors
- Chordomas (M9370)
- Schwannomas (M9560)
  - Acoustic schwannomas/neuromas

Other CNS Tumors (2)

Embryonal tumors

- Retinoblastomas (M9510)
- Primitive neuroectodermal tumors (PNETs)
  - Meduloblastomas (M9470)
  - Neuroblastomas (M9500)

Other CNS Tumors (3)

Lymphomas (M9590)

- Arise from
  - Indigenous brain histiocytes (microglia)
  - Rare lymphocytes in meninges
- High incidence in patients with AIDS

Vascular tumors

- Rare, non-malignant tumors
- Arise from blood vessels of brain and spinal cord
- Hemangioblastoma (M9161) most common vascular tumor

Other CNS Tumors (4)

Cysts and tumor-like lesions

- Reportable
  - Dermoid cysts (M9084)
  - Granular cell tumors (M9580)
  - Rathke pouch tumors (M9350)
- Not reportable
  - Epidermoid cysts
  - Colloid cysts
  - Enterogenous cysts
  - Neuroglial cysts
  - Plasma cell granulomas
  - Nasal glial herterotopias
  - Rathke cleft cysts

Childhood versus Adult Tumors

- CNS tumor histology and location are different in adult and children.
- Tumor location and extent of spread affect treatment and prognosis.
- Most common solid tumor in childhood.

Childhood Brain Tumors

- Meduloblastomas are the most common CNS histology in children.
- 50% are infratentorial.
- Common infratentorial tumors:
  - Cerebellar astrocytomas
  - Meduloblastomas
  - Ependymomas
  - Brain stem gliomas
  - Atypical teratoid tumors

Cellular Classification
Childhood Brain Tumors (1)

Supratentorial tumors in children

- Craniopharyngiomas
- Germ cell tumors
- Diencephalic and hypothalamic gliomas
- Low grade astrocytomas
- Mixed gliomas
- Anaplastic astrocytomas
- Oligodendrogliomas
- PNETs
- Meningiomas
- Glioblastoma multiforme
- Low-grade or anaplastic ependymomas
- Choroid plexus tumors
- Pineal parenchymal tumors
- Gangliogliomas
- Desmoplastic infantile gangliogliomas
- Dysembryoplastic neuroepithelial tumors

Cellular Classification
Childhood Brain Tumors (2)

The histopathology of childhood spinal tumors is the same as for childhood brain tumors.

Primary spinal cord tumors comprise approximately 1% to 2% of all childhood CNS tumors.

Cellular Classification
Childhood CNS Tumors

Cause of childhood CNS tumors remains unknown.

American Academy of Pediatrics has outlined guidelines for pediatric cancer centers and their role in the treatment of pediatric cancer patients.

ICD-O-3 Coding Issues (1)

- Some histologies may be difficult to determine if the primary site is intracranial or the skull (C41.0).
- Non-malignant tumors of the skull are not reportable.
  - Chondroma (M9220/0) must originate in a brain-related site to be reportable.
  - Chordoma (M9370/3) and chondrosarcoma (M9220/3) are malignant.
- Tumors in brain-related sites are analyzed separately from those in the skull.

ICD-O-3 Coding Issues (2)

Continue to assign histology code M9421/3 to pilocytic astrocytoma.

When the primary site for intracranial schwannoma (9560/0) is not documented in source documents, the site should be coded to cranial nerves NOS (C72.5).

Grade for CNS Tumors

Sixth digit of ICD-O-3 histology code

- Describes tumor differentiation or grade.
- Is not usually specified for CNS tumors.
- Is always assigned code 9 for non-malignant CNS tumors:
  - Not determined, not stated, or not applicable.
  - Per ICD-O-3, page 30, Rule G, paragraph 1 "Only malignant tumors are graded."
- Not the same as WHO grade.

WHO Grade (1)

WHO grade coded in Collaborative Stage data field:

- Site-specific factor 1 for Brain.

Four-category tumor grading system

Grade I

- Slow growing
- Non-malignant tumors
- Patients have long-term survival.

WHO Grade (2)

Grade II

- Relatively slow growing
- Sometimes recur as higher grade tumors
- May be non-malignant or malignant .

Grade III

- Malignant tumors
- Often recur as higher grade tumors.

Grade IV

- Highly malignant and aggressive.

Kernohan Grade

Defines progressive malignancy for astrocytoma

- Grade 1: benign astrocytomas
- Grade 2: low-grade astrocytomas
- Grade 3: anaplastic astrocytomas
- Grade 4: glioblastoma multiforme

No NAACCR data field for Kernohan grade.

St. Anne-Mayo Grade (1)

- Used for astrocytomas.
- Uses four morphologic criteria:
  - Nuclear atypia
  - Mitosis
  - Endothelial proliferation
  - Necrosis

No NAACCR data field for the St. Anne-Mayo grade.

St. Anne-Mayo Grade (2)

- Grade 1: No criteria
- Grade 2: One criterion, usually nuclear atypia
- Grade 3: Two criteria, usually nuclear atypia and mitosis
- Grade 4: Three or four criteria

Grade for CNS Tumors

- Do not record WHO grade, Kernohan grade, or St. Anne/Mayo grade in the sixth digit histology code data field

Part III

- Laterality
- Multiple Primaries
- Malignant Transformation
- Sequence Numbers
- Date of Diagnosis

Determining Multiple Primaries: Laterality

- Brain is not a paired organ.
- Laterality collected on both non-malignant and malignant tumors.
- Used to determine if multiple non-malignant CNS tumors are counted as multiple primary tumors.
- Not used to determine if multiple malignant tumors of the same intracranial or CNS site are multiple primary tumors.

Coding Laterality (1)

CNS sites to be coded with laterality:

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

Coding Laterality (2)

CNS sites to be coded with laterality (continued):

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

Determining Multiple Primaries: Definitions

Non-malignant tumor

Tumor with ICD-O-3 behavior code 0 (benign) or 1 (borderline).

CNS

Includes intracranial and central nervous system topographic sites.

Determining Multiple Primaries Malignant (1)

NO CHANGES (at this time)

Site

- Rule: Each category (first three characters) as delineated in ICD-O-3 is considered to be a separate site.
- Multiple tumors are:
  - Same: C7**1**.0 Cerebrum, C71.**2** Temporal lobe
  - Different: C7**0**.0 Cerebral Meninges, C7**1**.0 Cerebrum

Determining Multiple Primaries: Malignant (2)

Histology

- Rule: Differences in histologic type refer to differences in the *FIRST THREE* digits of the morphology code.
- Multiple tumors in the same site are:
  - Same: Choroid plexus carcinoma (M9390), Ependymoma (M9391)
  - Different: Astrocytoma (M9400), Gemistocytic astrocytoma (M9411)

Determining Multiple Primaries Non-malignant (1)

NEW RULES

Site

- Rule: Each **sub-site** (fourth-digit level) as delineated in ICD-O-3 is considered a separate site.
  - Same site if separate tumors with the same histology are in the same sub-site.
  - Different site if separate tumors have the same histology in different sub-site
    - C71.**1** Frontal lobe, C71.**4** Occipital lobe
    - C7**0.0** Cerebral Meninges, C70.**1** Spinal meninges.

Determining Multiple Primaries Non-malignant (2)

Site (cont)

- **EXCEPT** NOS (C_ _.9) with specific four-digit site code in same rubric

Example: meninges, NOS (C70.9) with spinal meninges (C70.1) or cerebral meninges (C70.0).

Determining Multiple Primaries Non-malignant (3)

Site (cont)

- Laterality: For non-malignant cases only

  If multiple tumors of the same site and same histologic type are identified and <u>both sides</u> of a site listed as lateral are involved, tumors should be counted as separate primaries.

- Different:
  - Right temporal lobe (**C71.2**) and left temporal lobe (**C71.2**)

Determining Multiple Primaries: Non-malignant (4)

Histology

Table 2: Histologic Groupings to Determine Same Histology for Non-malignant Brain Tumors	
Choroid plexus neoplasms	9390/0, 9390/1
Ependymomas	9393, 9394, 9444
Neuronal and neuronal-glial neoplasms	9384, 9412, 9413, 9442, 9505/1, 9506
Neurofibromas	9540/0,9540/1, 9541, 9550, 9560/0
Neurinomatosis	9560/1
Neurothekeoma	9562
Neuroma	9570
Perineurioma, NOS	9571/0

Determining Multiple Primaries: Non-malignant (5)

Histology

If multiple tumors are in the same site, refer to Table 2, and use the following rules in priority order:

A-1: If the first three digits are the same but the codes are not found in Table 2, then the histology is considered to be the **SAME**.

A-2: If the first three digits are different but the codes are not found in Table 2, then the histology is **considered to be DIFFERENT.**

Determining Multiple Primaries: Non-malignant (6)

Histology (cont.)

B. If **all** histologies are listed in the same histologic group in Table 2, then the histology is considered to be the **SAME.** *

Example: Ependymomas: M9394, Myxopapillary ependymoma and M9444, Chordoid glioma have the same histology

*Note: If two histologies are in the same group in Table 2, code the first or more specific histology.

Determining Multiple Primaries: Non-malignant (7)

Histology (cont)

C: If the first three digits are the same as the first three digits for **any histology** in one of the groupings in Table 2 , then the histology is considered to be the **SAME.***

Example: On table: Neuronal and neurol-glial neoplasm: M9505, ganglioglioma, Not on table: M9507, Pacinian tumor

* Note: If two histologies are in the same group in Table 2, code the first or more specific histology.

Determining Multiple Primaries: Non-malignant (8)

Histology (cont)

D: If the first three digits are the same and the histologies are from two different groups in the histologic groupings table, the histologies are considered to be **DIFFERENT.**

Example: Gliomas: M9442, Gliofibroma; Ependymoma: M9444, Chordoid glioma

Determining Multiple Primaries: Timing (1)

Primary malignant CNS tumors

NO CHANGE

- Malignant tumors of the same site and same histology, diagnosed within 2 months:
  - Tumors are counted as the **SAME** primary.
- Malignant tumors of the same site and same histology, diagnosed more than 2 months apart:
  - Tumors are counted as **DIFFERENT** primary sites.

Determining Multiple Primaries: Timing (2)

Primary non-malignant CNS tumors

NEW

- No timing rule
- If a new non-malignant tumor of the same histology as an earlier tumor that had been diagnosed in the same site is diagnosed subsequently at *any* time, it is considered to be the SAME primary tumor.

General Rules for Determining Multiple Primaries of CNS Sites (1)

Multiple lesions: all **non-malignant**

1. If different sites, then **DIFFERENT** primaries.
2. If different histologies, then **DIFFERENT** primaries.

General Rules for Determining Multiple Primaries of CNS Sites (2)

Multiple lesions: all **non-malignant** (cont.)

3. If same site and same histology:
   a. Laterality is same side, one side unknown or not applicable, then **SAME** primary.
   b. Laterality is both sides, then **DIFFERENT** primaries.

General Rules for Determining Multiple Primaries of CNS Sites (3)

B. Multiple tumors: **One non-malignant** and **one malignant**
   1. Non-malignant tumor followed by malignant tumor: **DIFFERENT** primaries, regardless of timing.
   2. Malignant tumor followed by a non-malignant tumor: **DIFFERENT** primaries, regardless of timing.

Histologic Transformation (1)

Histologic transformation or progression to a higher grade:

- Determined by pathological review.
- Final diagnosis made by review of previous biopsies or excisions and comparison to newly biopsied or resected brain tumor
  - Non-malignant tumor transforms to malignant tumor.
  - Malignant tumors transforms to higher grade tumor.

Histologic Transformation (2)

If a malignant CNS tumor recurs (transforms) as a higher grade tumor,

- SAME tumor.
- Do not change the histology or grade.
- Do not abstract as new primary.

Example: Astrocytoma (M9400) transforms to glioblastoma multiforme (M9440).

Histologic Transformation (3)

Transformation of a non-malignant tumor to a malignant tumor is rare.

Malignant transformations include:

- Changes from WHO grade I to WHO grade II, III, or IV.
- Changes from behavior code 0 or 1 to code 2 or 3.

Complete two abstracts:

- One for the non-malignant tumor
- One for the malignant tumor

Histologic Transformation (4)
Sequence Numbers

- Non-malignant tumors: assigned sequence numbers from the reportable-by-agreement series.
- Malignant tumors: assigned sequence numbers from the malignant series.
  - Example: Patient has a non-malignant CNS tumor that progressed into a malignant CNS tumor:
    - Non-malignant tumor is sequenced as 60.
    - Malignant tumor is sequenced as 00.

Histologic Transformation (5)
Date of Diagnosis

- Non-malignant tumors: First date that a medical practitioner diagnosed the non-malignant tumor either clinically or histologically.

- Malignant tumors: First date that a medical practitioner diagnosed the malignant transformation either clinically or histologically.

Coding Sequence Numbers (1)

- Indicates the sequence of all reportable neoplasms over the lifetime of the person.
- Codes 00 – 35: Malignant and in situ reportable neoplasms.
- Codes 60 – 87: Reportable-by-agreement including non-malignant tumors diagnosed after January1, 2004.

Coding Sequence Numbers (2)

Reportable-by-agreement neoplasms are defined by each facility and/or central cancer registry:

- Non-malignant CNS tumors are assigned reportable-by-agreement sequence numbers even when they are reportable.
- Codes 60 – 87

Coding Sequence Numbers (3)

Sequence numbers for non-malignant CNS tumors are assigned over the lifetime of the person.

- Example: Patient diagnosed with a non-malignant CNS tumor in January, 2003 (not reportable by state or hospital reporting rules) and diagnosed with second non-malignant CNS tumor in 2004:
  - Second is sequence number 62.
  - Complete abstract for the second tumor only.

Assigning Diagnosis Date

- Rules for assigning diagnosis date are the same for malignant and non-malignant tumors.
- Review source records carefully to determine initial diagnosis date, regardless of whether it is a clinical or histological diagnosis.

Part IV

- Staging
- Risk Factors
- Genetic Syndromes
- Diagnostic Tools
- Treatment
- Edits
- Data Analysis

Collaborative Stage (CS)

A computer algorithm uses the collaborative stage (CS) data fields to calculate site-specific American Joint Committee on Cancer (AJCC) TNM stage, SEER Summary Stage 1977, and SEER Summary Stage 2000.

Coding Collaborative Stage (1)

Separate sets of extension codes for:

- Brain and cerebral meninges
- Other parts of the CNS
- Glands: pituitary gland, craniopharyngeal duct, and pineal gland.

Coding Collaborative Stage (2)

- Site-specific codes for lymph nodes
  - Same for the Brain, cerebral meninges and other CNS.
    - Code 88: Not applicable.
  - For pituitary gland, craniopharyngeal duct, and pineal gland
    - Code 99: Not applicable.
- Metastasis at Diagnosis
  - Same for the pituitary gland, craniopharyngeal duct, and pineal gland and other CNS.
  - Different for brain and cerebral meninges.

CS Extension: Brain and Meninges C70.0, C71.0 – C71.9 (1)

05Benign or borderline brain tumors

10 Supratentorial tumor confined to CEREBRAL HEMISPHERE (cerebrum) or MENINGES of cerebral hemisphere one side: frontal lobe, occipital lobe, parietal lobe, or temporal lobe

11 Infratentorial tumor confined to CEREBELLUM or MENINGES of CEREBELLUM on one side: Vermis, lateral lobes, median lobe of cerebellum

CS Extension: Brain and Meninges C70.0, C71.0 – C71.9 (2)

12 Infratentorial tumor confined to BRAIN STEM or MENINGES of BRAIN STEM on one side: medulla oblongata, midbrain (mesencephalon), pons, hypothalamus, or thalamus

15 Confined to brain, NOS, Confined to meninges, NOS

20 Infratentorial tumor: Both cerebellum and brain stem involved with tumor on one side

30 Confined to ventricles - Tumor invades or encroaches upon ventricular system

CS Extension: Brain and Meninges C70.0, C71.0 – C71.9 (3)

40 Tumor crosses the midline: involves the contralateral hemisphere, involves corpus callosum (including splenium)

50 Supratentorial tumor extends infratentorially to involve cerebellum or brain stem

51 Infratentorial tumor extends supratentorially to involve cerebrum (cerebral hemisphere)

60 Tumor invades bone (skull), major blood vessel(s), meninges (dura), nerves, NOS (cranial nerves), or spinal cord/canal

CS Extension: Brain and Meninges C70.0, C71.0 – C71.9 (4)

70 Circulating cells in cerebral spinal fluid; nasal cavity; nasopharynx; posterior pharynx; or outside CNS

80 Further contiguous extension

95 No evidence of primary tumor

99 Unknown extension; Primary tumor cannot be accessed; Not documented in patient record

CS Extension: Other CNS C70.1-9, C72.0–C72.9 (1)

- Spinal meninges, meninges NOS
- Spinal cord
- Caudia equina
- Olfactory, acoustic, cranial nerve, NOS
- Overlapping brain and CNS
- Nervous system, NOS

CS Extension: Other CNS
C70.1-9, C72.0–C72.9 (2)

05 Benign or borderline tumors

10 Tumor confined to tissue or site of origin

30 Localized, NOS

40 Meningeal tumor infiltrates nerve; nerve tumor infiltrates meninges (dura)

50 Adjacent connective/soft tissue; adjacent muscle

60 Brain, for cranial nerve tumors; major blood vessel(s); sphenoid and frontal sinuses (skull)

CS Extension: Other CNS C70.1-9, C72.0–C72.9 (3)

70 Brain except for cranial nerve tumors; bone, other than skull; eye

80 Further contiguous extension

95 No evidence of primary tumor

99 Unknown extension; primary tumor cannot be assessed; not documented in patient record

CS Extension: Other Endocrine C75.1, C75.2, C75.3

00 In situ; non-invasive; intraepithelial

05 Benign or borderline tumors

10 Invasive carcinoma confined to gland of origin

30 Localized, NOS

40 Adjacent connective tissue

60 Pituitary and craniopharyngeal duct: Cavernous sinus; infundibulum; pons; sphenoid body and siunses

Pineal: Infratentorial and central brain

80 Further contiguous extension

95 No evidence of primary tumor

99 Unknown extension

CS Lymph Nodes

Describes tumor involvement of regional lymph nodes.

- Code for CS Lymph Nodes is 88 (not applicable) for meninges, brain, spinal cord, cranial nerves, and other parts of the CNS.
- Code for CS Lymph Nodes is 99 (unknown, not stated) for pituitary gland, craniopharyngeal duct, and pineal gland.

CS Metastasis at Diagnosis Brain and Meninges
C70.0, C71.0-9

00 No; None

10 Distant metastases

85 "Drop" metastases

99 Unknown; distant metastasis cannot be assessed; not documented in patient record

CS Metastasis at Diagnosis Other CNS and Other Endocrine C70.1-9, C72.0—9, C75.1, C75,2, C75.3

00 No; None

10 Distant lymph node(s)

40 Distant metastasis except lymph nodes (code 10)

Distant metastasis, NOS

Carcinomatosis

50 (40) + (10)

99 Unknown; distant metastasis cannot be assessed; not documented in patient record

CS Site-specific Factor 1 (1)
C70.0-C70.9, C71.0-C71.9, C72.0-C72.9

010 WHO Grade I
020 WHO Grade II
030 WHO Grade III
040 WHO Grade IV
999 Clinically diagnosed; grade unknown;
Not documented in the medical record;
Grade unknown, NOS

CS Site-specific Factor 1 (2)
C70.0-C70.9, C71.0-C71.9, C72.0-C72.9 C75.1- C75.3

- Code the WHO grade for CNS tumors in CS Site-specific factor 1.
- Do not code WHO grade in the sixth digit histology data field.

Possible Risk Factors

- Genetic predispositions for the development of brain tumors have been identified.
- Population-based studies suggest that no more than 4% are attributed to heredity.
- Several environmental factors that may be associated with CNS tumors.

Possible Risk Factors

- Epstein-Barr virus in the DNA of primary lymphoma suggests a viral etiology for CNS tumors.

Reference: "Surveillance of Primary Intracranial and Central Nervous System Tumors: Recommendations from the Brain Tumor Working Group."

Genetic Syndromes

Genetic syndromes associated with multiple CNS tumors are:

- Neurofibromatosis I (von Recklinghausen's disease)
- Neurofibromatosis II (bilateral acoustic neurofibromatosis)
- Von Hippel-Lindau disease
- Tuberous sclerosis (Bourneville-Pringle syndrome)
- Gorlin syndrome (Nevoid Basal Cell Carcinoma syndrome
- Hermans-Grosfeld-Spaas-Valk disease
- Li-Fraumeni syndrome
- Familial retinoblastoma
- Turcot syndrome (Adenomatous Polyposis syndrome)
- Cowden disease

Diagnostic Tools – Physical Exam

Neurological examination

- Eye movements
- Vision
- Hearing
- Reflexes
- Balance and coordination
- Sense of smell and touch
- Abstract thinking
- Memory

Diagnostic Tools: Radiology

- Computerized tomography (CT) scan
- Magnetic resonance imaging (MRI)
- Positron emission tomography (PET)
- Single photon emission computed tomography (SPECT)
- Magnetoencephalography (MEG)
- Angiography

Diagnostic Tools: Laboratory tests

- Audiometry
- Electroencephalogram (EEG)
- Endocrine evaluation
- Evoked potentials
- Lumbar puncture
- Myelogram
- Perimetry

Diagnostic Tools

Needle biopsy

- Needle inserted through a burr hole and tissue extracted for tissue diagnosis.

Stereotactic biopsy

- Computer used to guided needle biopsy to extract tissue.

College of American Pathologist (CAP) Protocols

Site-specific checklists

- Required to be completed in the health record in hospitals with COC-approved cancer programs for cases diagnosed January 1, 2004 and later.

www.cap.org/cancerprotocols/protocols_index.html.

Brain and Spinal Cord CAP Protocols (1)

Macroscopic

- Specimen type
- Specimen size
- Tumor site
- Tumor size

Brain and Spinal Cord CAP Protocols

Microscopic

- Histologic type
- Histologic grade
- Margins
- Additional studies*
- Additional pathologic findings*
- Comments*

*Not required for COC approval.

Treatment (1)

- Watchful waiting
- Surgery
- Radiation
- Chemotherapy
- Hormonal therapy
- Immunotherapy
- Hematologic Transplant and Endocrine procedures

Treatment (2)

Inoperable or inaccessible tumors may be treated with primary radiation and other systemic therapy:

- Chemotherapy, immunotherapy, and hormone therapy.

Shunt insertion to reduce intracranial swelling is not coded as surgical treatment.

Surgical Procedure of Primary Site

- Brain: Site-specific surgery codes
  - Meninges
  - Brain
  - Spinal cord, cranial nerves, other CNS.
- All Other Sites: Site-specific surgery codes
  - Pituitary gland
  - Craniopharyngeal duct
  - Pineal gland.

Surgical Procedure of Primary Site C70-0-C70.9, C71.0-C71.9, C72.0-C72.9 (1)

- Code 10: Tumor destruction, NOS
  - Laser surgery
  - Laser surgery with photodynamic therapy
  - Ultrasonic aspirator.

No specimen sent to pathology from surgical procedure.

Surgical Procedure of Primary Site
C70-0-C70.9, C71.0-C71.9, C72.0-C72.9 (2)

- 20:Local Excision (biopsy) of tumor, lesion, or mass
  - Specimen sent to pathology from surgical event.
- 40: Partial resection
- 55: Gross total resection
- 90: Surgery, NOS

Surgical Procedure of Primary Site
C75.1, C75.2, C75.3 (1)

- Code 10: Local tumor destruction, NOS
- Code 11: Photodynamic therapy
- Code 12: Electrocautery; fulguration
- Code 13: Cryosurgery
- Code 14: Laser

No specimen is sent to pathology from surgical events 10-14.

Surgical Procedure of Primary Site
C75.1, C75.2, C75.3 (2)

- Code 20: Local tumor excision, NOS
- Code 26: Polypectomy
- Code 27: Excisional biopsy

  Any combination of 20 or 26-27 WITH
  - 21: Photodynamic therapy (PDT)
  - 22: Electrocautery
  - 23: Cyrosurgery
  - 24: Laser ablation

Surgical Procedure of Primary Site
C75.1, C75.2, C75.3 (3)

- Code 25: Laser excision

  Specimen sent to pathology from surgical event 20-27.

- Code 30: Simple or partial surgical removal of primary site.

Surgical Procedure of Primary Site
C75.1, C75.2, C75.3 (4)

- Code 40: Total surgical removal of primary site; enucleation
- Code 50: Surgery stated to be "debulking"
- Code 60: Radical surgery

  Partial or total removal of the primary site WITH resection in continuity (partial or total removal) with other organs
- Code 90: Surgery, NOS

Surgical Margins of the Primary Site

Code final status of surgical margins

- COC-required data item.
- Serves as quality control measure for pathology reports.
- May be prognostic factor in recurrence.

Scope of Regional Lymph Node Surgery

Identifies removal, biopsy, or aspiration of regional lymph node(s):

- NPCR-, COC-, and SEER-required data item.

- Code 9: Meninges, brain, and spinal cord; cranial nerves; and other parts of the CNS.
- Code as appropriate: Pituitary gland, craniopharyngeal duct, and pineal gland.

Radiation Therapy (1)

Radiation codes indicate type of radiation therapy performed as part of the first course of treatment.

- Records modality of radiation therapy used to deliver significant regional dose to the primary volume of interest.
- COC-required data item.
- SEER collects these data from COC-approved facilities
- NPCR: Supplementary or recommended.

Radiation Therapy (2)

Beam radiation

- Codes 20 – 29:
  - Conventional radiation therapy: from an external beam directed at the tumor.
  - Energy is orthovoltage, cobalt, photon, and/or electron.
- Code 30: Boron neutron capture therapy (BNCT)
- Code 31: Intensity-modulated radiation therapy (IMRT)

Radiation Therapy (3)

Beam radiation

- Code 32: Conformal radiation

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

Radiation Therapy (3)

- Tumors typically treated with stereotactic radiosurgery include:

- Acoustic neuroma
- Chordoma
- Pineal tumor
- Astrocytoma
- Craniopharyngioma
- Hemangioblastoma
- Pituitary adenomal tumor

Radiation Therapy (4)

Radioactive implants

- Code 50: Brachytherapy, radiation implants, radiation seeding, radioactive implants, interstitial implants, intracavitary radiation NOS
- Code 51: Intracavitary radiation with low dose rate applicators (Cesium-137, Fletcher applicator)

Radiation Therapy (5)

Radioactive implants (continued)

- Code 52: Intracavitary radiation with high dose rate applicator
- Code 53: Interstitial radiation with low dose rate sources
- Code 54: Interstitial radiation with high dose rate sources
- Code 55: Low dose rate interstitial or intracavitary radium

Chemotherapy (1)

Record type of chemotherapy administered as first course of treatment:

- Code 01: Chemotherapy, NOS
- Code 02: Single-agent chemotherapy
- Code 03: Multi-agent chemotherapy

Chemotherapy (2)

- Blood-brain barrier
  - Protects the brain from foreign substances, including chemotherapy.
  - May be disrupted by receptor-mediated permeabilizers.
- Intrathecal chemotherapy
  - Drugs directly injected into the cerebrospinal fluid by spinal injection or Ommaya reservoir.

Chemotherapy (3)

- Interstitial chemotherapy
  - Administered directly to involved tissues.
  - Polymer wafers soaked in a chemotherapeutic agent are inserted in the tumor bed after tumor resection.

Hormone Therapy

Record systemic hormonal agents administered as first course of treatment.

- Tamoxifen and RU-486 (Mifepristone) may be used to treat meningioma.
- Steroids given to treat swelling caused by CNS tumors are not coded as hormone therapy.

Immunotherapy (1)

Record whether immunotherapeutic agents were administered as first course of treatment:

- Angiogenesis inhibitors block the development of new blood vessels and starve the tumor.
- Interleukins are growth factors that manipulate the tumor's ability to grow.

Immunotherapy (2)

- Gene therapy replaces or repairs the gene responsible for tumor growth.
- Vaccine therapy allows the immune system to detect the tumor antigens and attack the tumor cells.

Hematologic Transplant and Endocrine Procedures

Identify systemic therapeutic procedures administered as first course of treatment:

- Code 10: Bone marrow transplant, NOS
- Code 11: Autologous bone marrow transplant
- Code 12: Allogeneic bone marrow transplant
- Code 20: Stem cell harvest
- Code 30: Endocrine surgery and/or endocrine radiation therapy
- Code 40: Combination of endocrine surgery and/or radiation with transplant procedure

Technical Issues
Edit Checks

- NAACCR Edits Committee is developing and modifying data edits to accommodate data collection of non-malignant CNS tumors.

Technical Issues
Data Analysis Recommendations

- Report and analyze data for non-malignant CNS tumors separately from malignant tumors.
- Footnote that pilocytic astrocytomas are included in the analysis for malignant brain tumors for continuity of trends.
- Review the standard site and histology groupings for tabulating estimates of these tumors to allow comparability of information across registries.

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