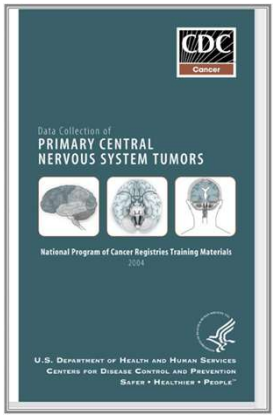

CNS Part 1
NPCR DQE Findings

- Barbara J. Dearmon-Neyland, BS, ODS-C
- FCDS Webcast Series 2026
- August 12, 2026

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CDC & Florida DOH Attribution

- "Funding for this conference was made possible (in part) by the Centers for Disease Control and Prevention. The views expressed in written conference materials or publications and by speakers and moderators do not necessarily reflect the official policies of the Department of Health and Human Services, nor does the mention of trade names, commercial practices, or organizations imply endorsement by the US Government."
- FCDS would also like to acknowledge the Florida Department of Health for its support of the Florida Cancer Data System, including the development, printing and distribution of materials for the 2024 FCDS Webcast Series under state contract COHAW. The findings and conclusions in this series are those of the author(s) and do not necessarily represent the official position of the Florida Department of Health.

2

2

FLccSC
Fundamental Learning Collaborative for the Cancer Surveillance Community

FLccSC LMS – CEU Quiz – FCDS IDEA

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NCRA CEU# 2026-041

2 CEUs Awarded; meets 2 Category A

Log in to FLccSC to take the quiz to get the certificate

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3

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Objectives

- Brain and CNS Tumor Facts
- Brain and CNS Tumors Reportability
- Review of Anatomy, Brain, and CNS Tumors
- NPCR DQE Findings
- Review STRs Non-Malignant CNS Tumors

4

8/12/2026

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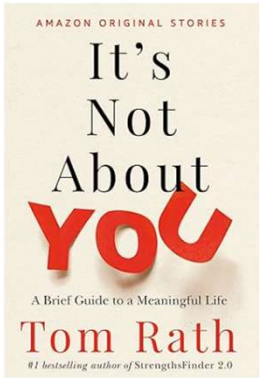
Did You Know? Brain and Non-Malignant Tumor Facts

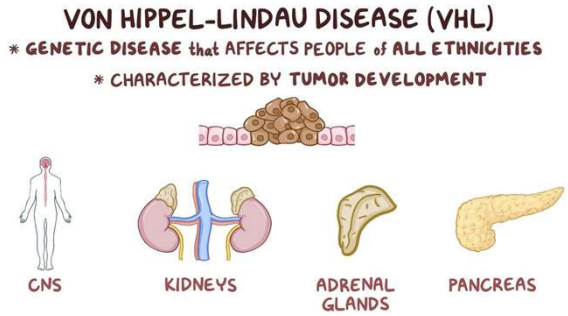
https://seer.cancer.gov/statfacts/html/brain.html

5

Von Hippel-Lindau Disease (VHL)

https://www.osmosis.org/learn/von_Hippel-Lindau_disease





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MyTumorID

- National Brain Tumor Society
- Biomarker
- Biomarker testing
- Genetic testing for inherited cancer risk
- Mutation
- Next generation sequencing

https://braintumor.org/brain-tumors/diagnosis-treatment/mytumorid/biomarker-testing/

7

Journal of Neuro-Oncology (2025) 172:567–578
https://doi.org/10.1007/s11060-025-04944-y

RESEARCH

Cancers of the brain and central nervous system: global patterns and trends in incidence

Adalberto M. Filho[1] · Ariana Znaor[1] · Ceren Sunguc[1] · Mariam Zahwe[1] · Rafael Marcos-Gragera[2,3,4,5,6] · Jonine D. Figueroa[7] · Freddie Bray[1]

Received: 2 December 2024 / Accepted: 16 January 2025 / Published online: 30 January 2025

Abstract

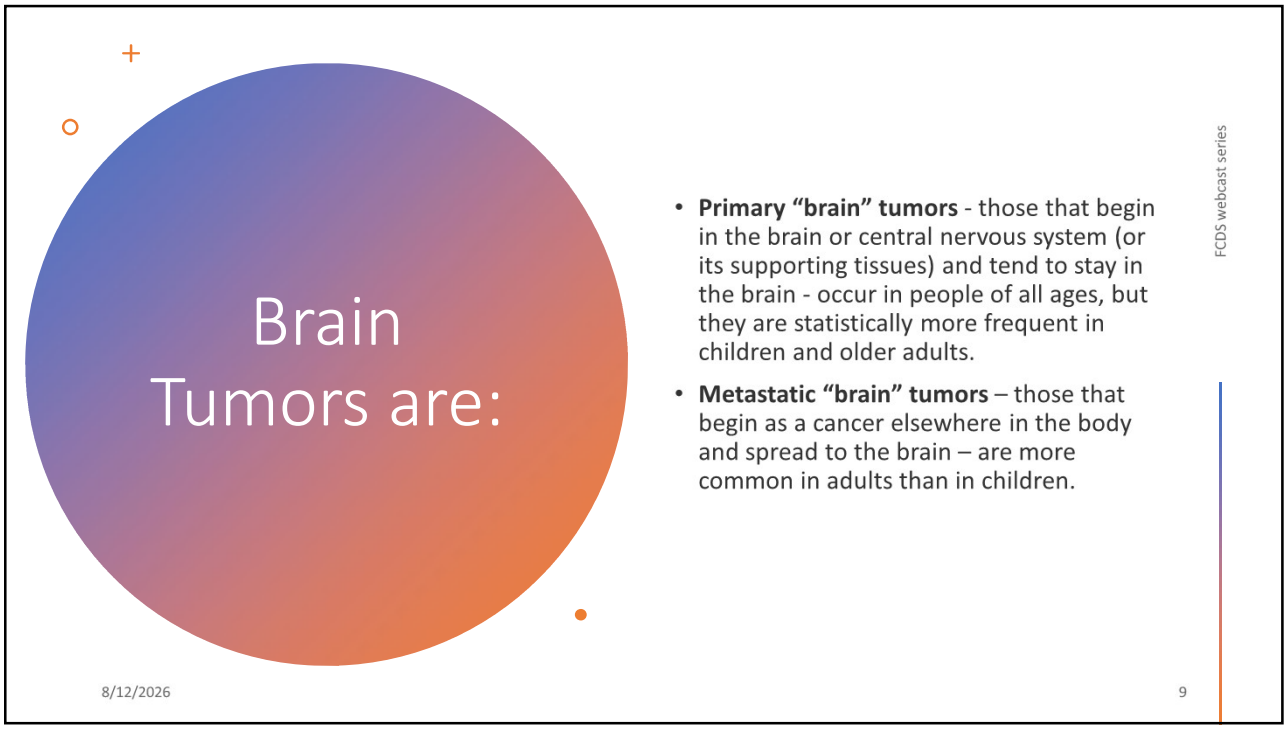
Background Global comparisons of the burden and impact of cancers of the brain and central nervous system (CNS) are critical for developing effective control strategies and generating etiological hypotheses to drive future research.

Methods National incidence estimates were obtained from GLOBOCAN 2022, and recorded incidence data from the Cancer in Five Continents series, both developed and compiled by the International Agency for Research on Cancer. We examined the estimated age-standardized incidence rates in 185 countries, as well as time trends in recorded incidence in 35 countries, quantifying the direction and change in the magnitude of the rates using the estimated average percentage change (EAPC).

Results In 2022, 322,000 new cases of brain and CNS tumors were estimated globally. By world region, the highest incidence rate was seen in Northern America (5.46 per 100,000), Eastern Asia (3.95), and Western Europe (5.56). Africa had relatively lower incidence rates. By country and age group, Austria and the U.S. exhibited the highest rates in boys (3.5 in both), while in adolescents and young adults (AYA), Norway had the highest incidence rates in both males (4.7) and females (3.8). Among adults (+ 40yo), the highest rates in males were observed in the Northern European countries of Norway (18.6), Lithuania (18.4), and Latvia (16.7). In terms of time trends, incidence rates tended to be rather stable in most world regions over the last decade, though increases were observed in selected countries. Trends-based predictions indicate that if incidence rates remain stable, population ageing and growth would mean there would be 474,000 new cases by the year 2045, a 47% increase from 2022.

Conclusion While the increased incidence rates in certain populations require further study, the future predictions based on

8



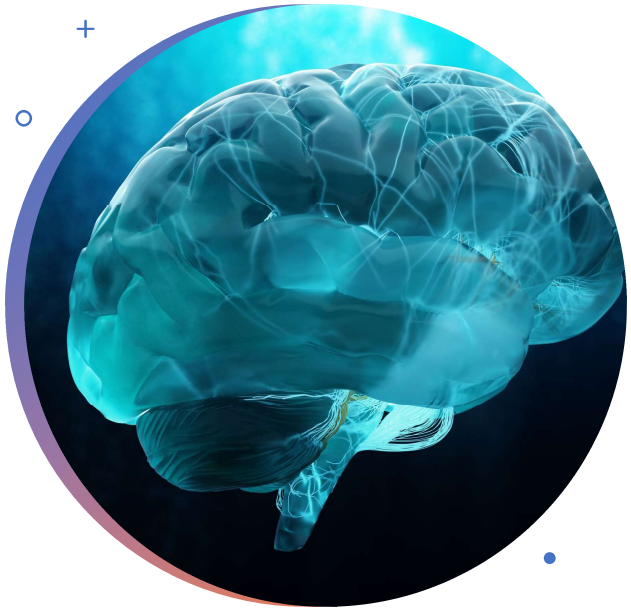
9

National Brain Tumor Society - Facts

- An estimated 1 million Americans are living with a primary brain tumor
- Approximately 72% of all brain tumors are benign
- Approximately 28% of all brain tumors are malignant
- Approximately 59% of all brain tumors occur in females and 41% occur in males
- Median age at diagnosis for primary brain tumor is 61 years

https://braintumor.org/brain%20tumors/about%20brain%20tumors/brain%20tumor%20facts/

10



Brain Cancer Survival Rates

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The overall five-year relative survival rate 76%.

A non-malignant brain tumor average five-year survival rate of 91.8%; for meningioma, it is 88.2%.

For malignant brain tumors, the five-year survival rate is 35.7%.

Glioblastoma, the most common malignant type, five-year survival rate of 6.9%

In 2025, brain cancer was the 9th leading cause of death for all genders and ages

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Symptoms and Complications

- CNS tumors are associated with a range of symptoms and complications such as edema, seizures, endocrinopathy, fatigue, psychiatric disorder, venous thromboembolism that can seriously impact quality of life.
- Symptoms depend on the size and location of the tumor, growth of tumor, swelling in the brain (edema), or blockage in the flow of cerebrospinal fluid.
- Symptoms of increased intracranial pressure include headaches which tend to be worse with activity, at night or early in the morning, convulsions, vomiting, changes in personality, behavior, memory, mental ability, drowsiness, lethargy and coma.

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Tumor Location and Symptoms

- Symptoms are often tumor location-specific or provide clues
- Symptoms on the right side of the body may occur if the tumor is located on the left side of the brain and vice-versa.
  - The speech center in most people is on the left side of the brain. If the tumor is located in the frontal lobe, which controls intellectual function, thought process, behavior, and memory, those activities may be affected.
  - Tumors in the cerebellum cause balance and motor function problems.
- Similarity to closed head injury victims (motorcycle crash).

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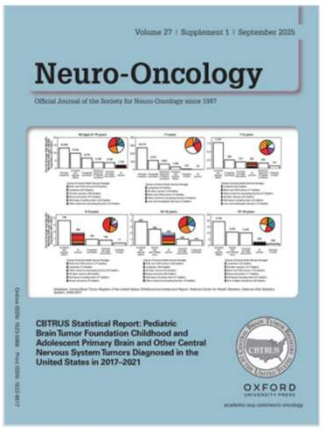
CBTRUS Central Brain Tumor Registry of the United States 2017-2021



NEURO-ONCOLOGY
Volume 27 | Issue Supplement 1 | September 2025

The **CBTRUS Statistical Report: Pediatric Brain Tumor Foundation Childhood and Adolescent Primary Brain and Other Central Nervous System Tumors Diagnosed in the United States in 2017–2021**, published as a Supplement to the Society for Neuro-Oncology journal, *Neuro-Oncology*, is available as a Free to View web publication through a link to Oxford University Press.

Citation: Mackenzie Price, Christine Ann Pittman Ballard, Julia R. Benedetti, Carol Kruchko, Jill S. Barnholtz-Sloan, Quinn T. Ostrom, CBTRUS Statistical Report: ediatric Brain Tumor Foundation Childhood and Adolescent Primary Brain and Other Central Nervous System Tumors Diagnosed in the United States in 2017-2021 , *Neuro-Oncology*, Volume 27, Issue Supplement_1, September 2025, Pages i1–i42, https://doi.org/10.1093/neuonc/noaf168

https://cbtrus.org/reports/

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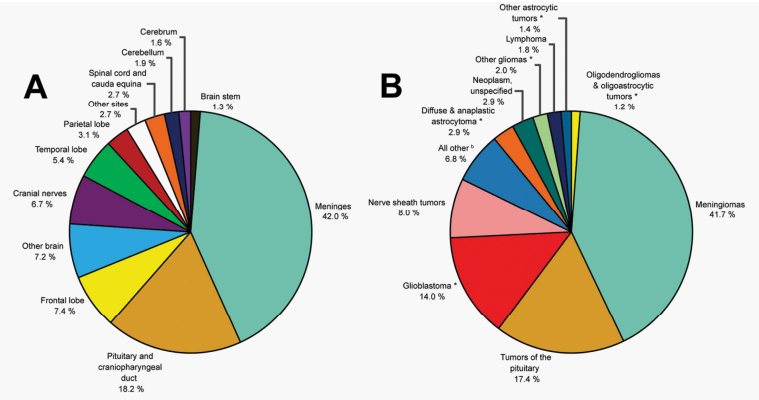
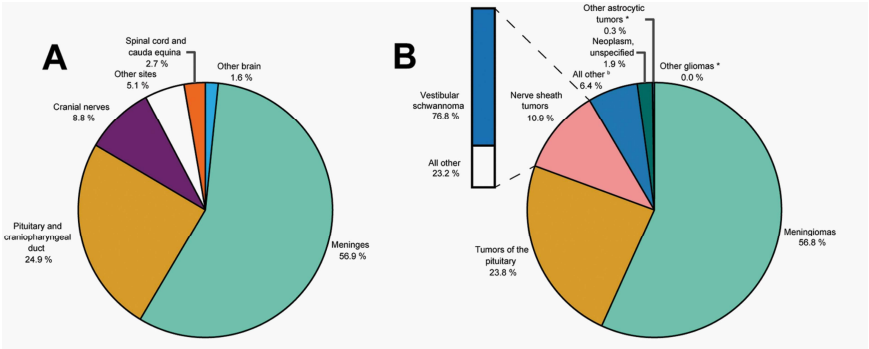
CBTRUS Distribution[a] of All Primary Brain and Other CNS Tumors (Malignant and Non-Malignant Combined)



* All or some of the codes used to define this histopathology are included in the CBTRUS definition of gliomas, including ICD-O-3 histopathology codes 9380-9384 and, 9391-9460 (Table 2).
a. Percentages may not add up to 100% due to rounding.
b. Includes ependymal tumors, neuronal and mixed neuronal-glial tumors, choroid plexus tumors, tumors of the pineal region, embryonal tumors, other tumors of cranial and spinal nerves, mesenchymal tumors, primary melanocytic lesions, other hematopoietic neoplasms, germ cell tumors, craniopharyngioma, hemangioma, all other (Table 2).
Data Source: CBTRUS Statistical Report: US Cancer Statistics – NPCR and SEER, 2017–2021
Abbreviations: CBTRUS, Central Brain Tumor Registry of the United States; NPCR, National Program of Cancer Registries; SEER, Surveillance, Epidemiology, and End Results Program.

15

CBTRUS Figure 12. Distribution[a] of All Non-Malignant Primary Brain and Other Central Nervous System Tumors ...



* All or some of this histopathology is included in the CBTRUS definition of gliomas, including ICD-O-3 histopathology codes 9380-9384 and, 9391-9460 (Table 2).
a. Percentages may not add up to 100% due to rounding.
b. Includes cases with behavior code /0 or /1 in ependymal tumors, neuronal and mixed neuronal-glial tumors, choroid plexus tumors, tumors of the pineal region, other tumors of cranial and spinal nerves, mesenchymal tumors, primary melanocytic lesions, lymphoma, other hematopoietic neoplasms, germ cell tumors, craniopharyngioma, hemangioma, all other (Table 2).
Data Source: CBTRUS Statistical Report: US Cancer Statistics – NPCR and SEER, 2017–2021
Abbreviations: CBTRUS, Central Brain Tumor Registry of the United States; NPCR, National Program of Cancer Registries; SEER, Surveillance, Epidemiology, and End Results Program.

Neuro Oncol, Volume 26, Issue Supplement_6, October 2024, Pages vi1–vi85, https://doi.org/10.1093/neuonc/noae145

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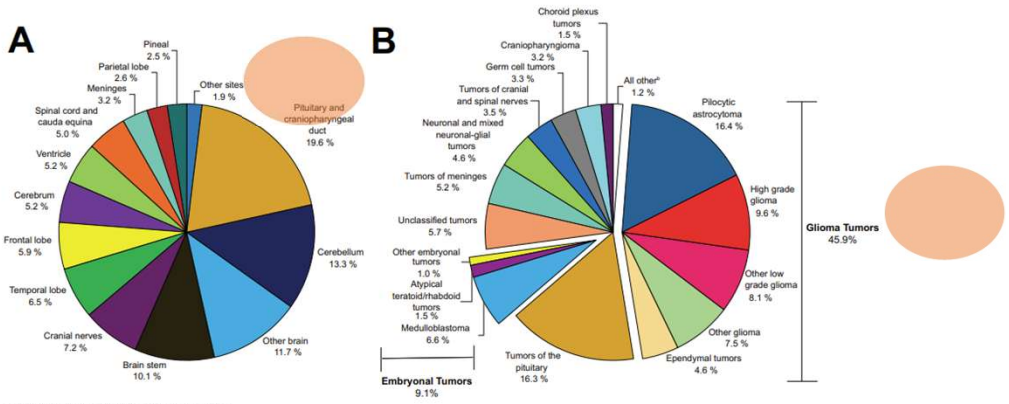
Supplementary Figure 3. Distributiona in Children and Adolescents Ages 0-19 Years of All Primary Malignant and Non-Malignant Brain and Other Central Nervous System Tumors (Five-Year Total=24,614; Annual Average Cases=4,923) by A) Site and B) Histopathology



[a]Percentages may not add up to 100% due to rounding
[b]All other includes the following histopatholoiges: lymphoma and hemopoetic neoplasms and tumors of the pineal region.
Database: *Central Brain Tumor Registry of the United States Childhood and Adolescent Report: US Cancer Statistics - CDC's National Program of Cancer Registries and NCI's Surveillance, Epidemiology and End Results Program, 2017-2021.*

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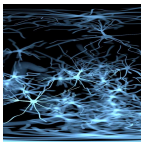
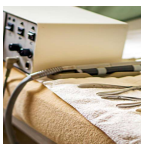
Glioblastoma: Deadly Brain Tumor



Aggressive Tumor Growth

Glioblastoma grows quickly and invades surrounding brain tissue, making it especially difficult to treat and remove completely.

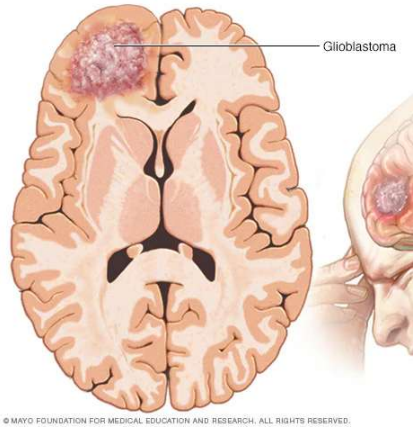


Challenging Treatment

Standard treatment involves surgery, radiation, and chemotherapy, but complete removal is rarely possible and recurrence is common.

Poor Prognosis

Despite advanced therapies, glioblastoma has a poor prognosis due to high recurrence rates and resistance to treatment.



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Non-Malignant Meningioma Facts

Most Common Non-Malignant Brain Tumor

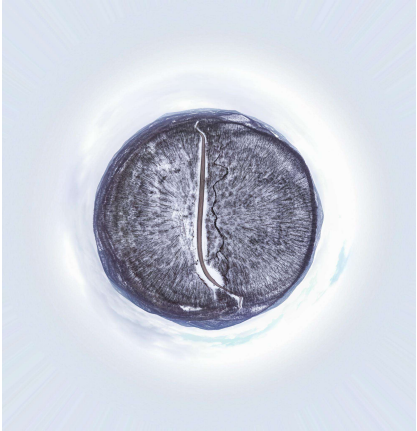
Non-malignant meningiomas are the most frequent primary non-malignant brain tumors, accounting for a significant portion of cases.

High Prevalence in Brain Tumors

They make up 39.7% of all brain tumors and 55% of all non-malignant tumors, highlighting their prevalence.

Favorable Prognosis

Patients with non-malignant meningiomas typically have a favorable outlook, with a five-year relative survival rate of 91.8%.



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NCCN Treatment Options- Meningiomas

- Observation or Active Surveillance
- Surgery
- Radiation
- Systemic therapy
- Clinical trials
- https://www.nccn.org/

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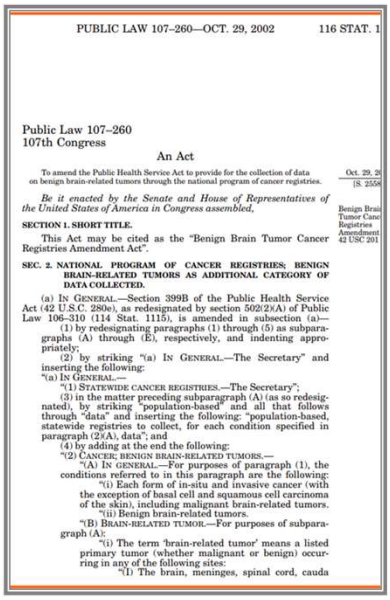
Reportability – Brain and CNS Tumors



PUBLIC LAW 107–260—OCT. 29, 2002 116 STAT. 1

Public Law 107–260
107th Congress

An Act

To amend the Public Health Service Act to provide for the collection of data on benign brain-related tumors through the national program of cancer registries. Oct. 29, 2[S. 255

Be it enacted by the Senate and House of Representatives of the United States of America in Congress assembled,

Benign Brai Tumor Canc Registries Amendment 42 USC 201

SECTION 1. SHORT TITLE.

This Act may be cited as the "Benign Brain Tumor Cancer Registries Amendment Act".

SEC. 2. NATIONAL PROGRAM OF CANCER REGISTRIES; BENIGN BRAIN-RELATED TUMORS AS ADDITIONAL CATEGORY OF DATA COLLECTED.

(a) IN GENERAL.—Section 399B of the Public Health Service Act (42 U.S.C. 280e), as redesignated by section 502(2)(A) of Public Law 106–310 (114 Stat. 1115), is amended in subsection (a)—

(1) by redesignating paragraphs (1) through (5) as subparagraphs (A) through (E), respectively, and indenting appropriately;

(2) by striking "(a) IN GENERAL.—The Secretary" and inserting the following:

"(a) IN GENERAL.—

"(1) STATEWIDE CANCER REGISTRIES.—The Secretary";

(3) in the matter preceding subparagraph (A) (as so redesignated), by striking "population-based" and all that follows through "data" and inserting the following: "population-based, statewide registries to collect, for each condition specified in paragraph (2)(A), data"; and

(4) by adding at the end the following:

"(2) CANCER; BENIGN BRAIN-RELATED TUMORS.—

"(A) IN GENERAL.—For purposes of paragraph (1), the conditions referred to in this paragraph are the following:

"(i) Each form of in-situ and invasive cancer (with the exception of basal cell and squamous cell carcinoma of the skin), including malignant brain-related tumors.

"(ii) Benign brain-related tumors.

"(B) BRAIN-RELATED TUMOR.—For purposes of subparagraph (A):

"(i) 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

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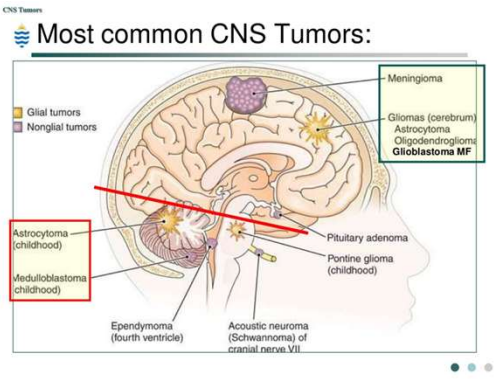
CNS Tumor Sites and Reportability

CNS tumors include tumors in the following sites:

- Meninges (C700-C709)
- Brain (C710–C719)
- Spinal Cord, Cranial Nerves, and Other Parts of the Central Nervous System (C720–C729)
- Pilocytic Astrocytoma/juvenile pilocytic astrocytoma (9421/1)
- High-grade Astrocytoma with Piloid Features (HGAP) (9421/3)



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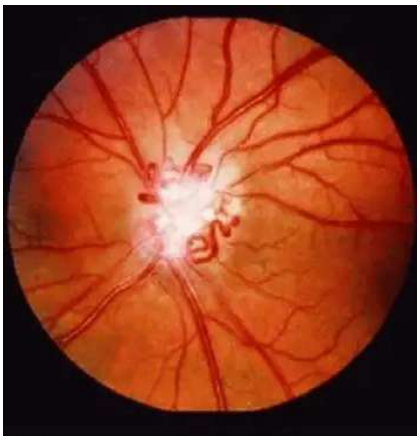
Reportability: Brain and CNS Tumors

- Is an optic nerve meningioma reportable if stated to arise in the "intraorbital segment" of the optic nerve meninges?



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Coding Brain and CNS Tumors

Understanding Neuroanatomy

WHO Classification of Brain and CNS Tumors

Standardized Coding Rules

- STRs
- STORE Manual
- SSDI
- ICD-0-3 Histology Codes
- Summary Stage

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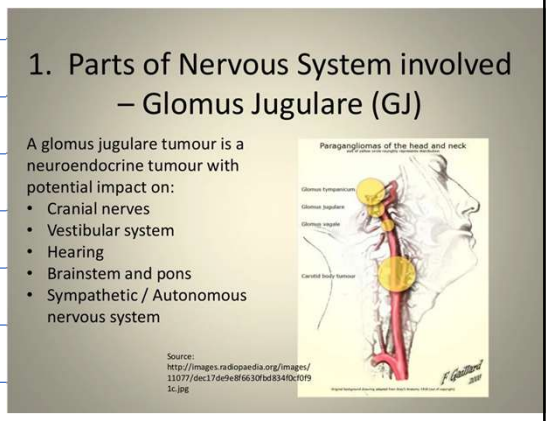
Types of Benign Brain Tumors

- Chordomas
- Craniopharyngiomas 9350/1
- Gangliocytomas 9492/0
- Glomus jugulare 8690/1 *(refer to Head and Neck Table 9)*
- Meningiomas 9530/0
- Pineocytomas 9361/1
- Pituitary adenomas 8272/0
- Schwannomas 9560/0



https://www.aans.org/patients/conditions-treatments/brain-tumors/

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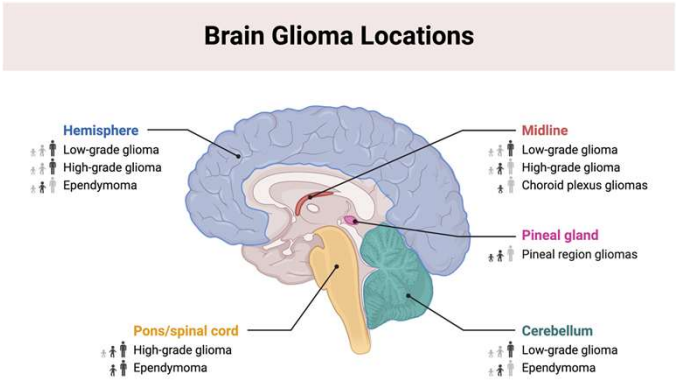
Types of Malignant Brain Tumors

- Gliomas
- Astrocytomas
- Ependymomas
- Glioblastoma multiforme (GBM)
- Oligodendrogliomas
- Medulloblastomas
- Hemangioblastomas
- Rhabdoid tumors



26

Anatomy Brain and CNS Tumors

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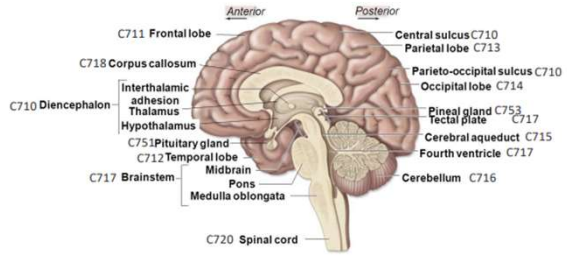
Brain and Associated Structures by Topography Codes

Primary Site

Brain and Associated Structures with Topography Codes



Extra-axial: Outside the brain parenchyma

Intra-axial: Within the brain

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Reportable Brain & CNS Sites

Use this quick reference to identify primary sites and reportable subsites for Brain/CNS cases.

Brain/CNS Reportability Snapshot

Primary site categories shown below include ICD-O-3 topography/site codes for reportable Brain/CNS subsites.

6	Primary site groups
C70–C75	Includes brain, meninges, CNS, glands
✓	Quick reference for casefinding

Brain
C71.0–C71.9 | NC, stem, lobes, NOS
Brain NOS C719
Brain stem C717
Cerebellum NOS C716
Cerebrum C710
Frontal lobe C711; Parietal C713
Occipital C714; Temporal C712
Overlapping lesion of brain C718
Ventricle NOS C715

Cranial Nerves
C72.3–C72.5 | individual nerve sites
Abducens VI C725; Accessory XI C725
Acoustic VIII C724; Cranial nerve NOS C725
Facial VII C725; Glossopharyngeal IX C725
Hypoglossal XII C725; Oculomotor III C725
Olfactory I C722; Optic II C723
Trigeminal V C725; Trochlear IV C725; Vagus X C725

Meninges
C70.0–C70.9
Cerebral meninges C700
Meninges NOS C709
Spinal meninges C701

Intracranial Duct & Glands
C75.1–C75.3
Craniopharyngeal duct C752
Pineal gland C753
Pituitary gland C751

Spinal Sites
C72.0–C72.1
Cauda equina C721
Conus medullaris / filum terminale C720

Ill-Defined Sites
C72.8–C72.9 | CNS NOS / overlapping lesion
Nervous system NOS C729
Overlapping lesion of brain and central nervous system C728

Tip: Confirm reportability use current STRs/state guidance and applicable coding manuals before abstraction.

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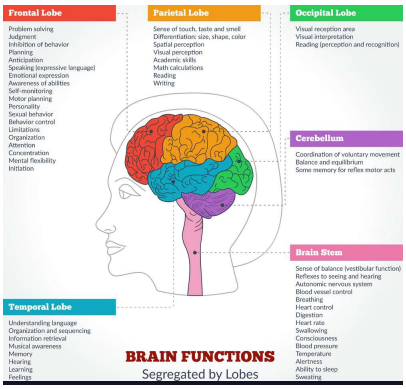
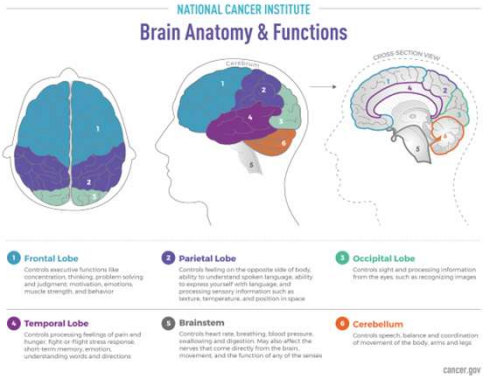
Brain Anatomy and Function as Related to Symptoms

VectorStock – Human Brain Anatomy and Functions – Vector Image





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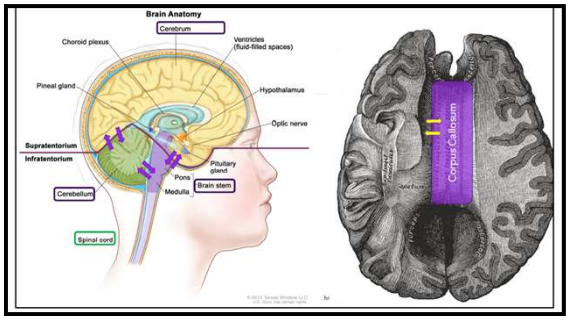
Tentorium C700



Supratentorial, NOS C710

Supratentorial subsites

- All subsites coded C711-C715
- Primary site C710 (excluding hypothalamus, pallium, thalamus)
- Anterior cranial fossa (C719)
- Corpus callosum (C718)
- Middle cranial fossa (C719)
- Tapetum (C718)
- Suprasellar (C719)

Infratentorial, NOS C717

Infratentorial subsites

- All subsites coded C716-C717
- Hypothalamus (C710)
- Pallium (C710)
- Posterior cranial fossa (C719)
- Thalamus (C710)

Supratentorial and Infratentorial subsites are based on Summary Stage 2018.

https://seer.cancer.gov/manuals/2026/AppendixC/Coding_Guidelines_Brain_Malig_2026.pdf

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Non-Malignant CNS Tumors

- Benign and borderline primary intracranial and central nervous system (CNS) tumors with a behavior code of /0 or /1 in ICD-O-3 **are reportable as of 01/01/2004.**
- **Cases diagnosed before 1/1/2004 are not reportable to FCDS**.
- FCDS requires reporting of all benign, borderline, and malignant tumors of the Brain, Central Nervous System, Cranial Nerves, Intracranial Glands, Meninges and Peripheral Nerve Tumors.
- CDC published a reference manual in 2004 entitled "Data Collection of Primary Central Nervous System Tumors." The manual is free of charge in PDF format on the CDC NPCR Website at https://stacks.cdc.gov/view/cdc/11345
- This document and ICD-O-3 are the primary references when determining case reportability for primary brain and CNS tumors

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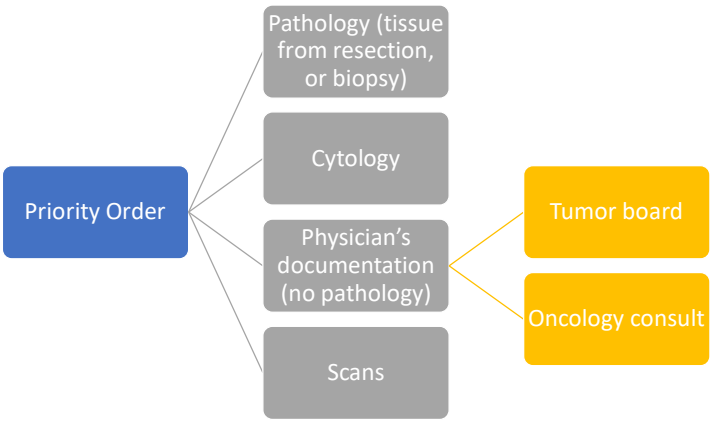
Assigning Behavior for Non-Malignant CNS Tumors



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NPCR and Registry Audits

Cancer Surveillance Support

NPCR plays a vital role in supporting cancer surveillance nationwide, gathering critical health data for analysis and action.

Registry Data Auditing

Audits performed by NPCR ensure the accuracy, completeness, and consistency of central cancer registries.

Impact on Public Health

High-quality audited data aids research, public health decisions, and resource allocation for effective cancer prevention and control.

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National Program of Cancer Registries
Data Quality Evaluation 2024 – 2025: Validation

Florida Cancer Data System

Diagnosis Year: 2019-2021

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Data Elements Reviewed During Reconsolidation

Race	Cancer Identification	Staging/Prognostic Factors	Treatment Data	
Race 1	Date of Diagnosis	Tumor Size Summary	Date of 1st Course RX—CoC	RX Date—Chemo
Race 2	Primary Site	Summary Stage 2018	**OR**	RX Summ—Chemo
Race 3	Laterality	Regional Lymph Nodes Positive	Date of Initial Rx—SEER	RX Date—Hormone
Race 4	Diagnostic Confirmation	Regional Lymph Nodes Examined	**AND**	RX Summ—Hormone
Race 5	Histologic Type ICD-O-3		RX Summ--Treatment Status	RX Date — BRM
	Behavior Code ICD-O-3		RX Date – Surgery	RX Summ—BRM
	Grade Clinical		RX Date Mst Defn Srg	RxSumm—Systemic/Sur Seq
	Grade Pathological		RX Summ—Surg Prim Site	RX Summ—TransplntEndocr
	Grade Post Therapy		RX Summ—Scope Reg LN Sur	RX Date — Other
			RX Summ—Surg Other Reg/Dis	RX Summ—Other
			Reason No Surgery	
			RX Date—Radiation	
			Phase I Radiation Treatment Modality	

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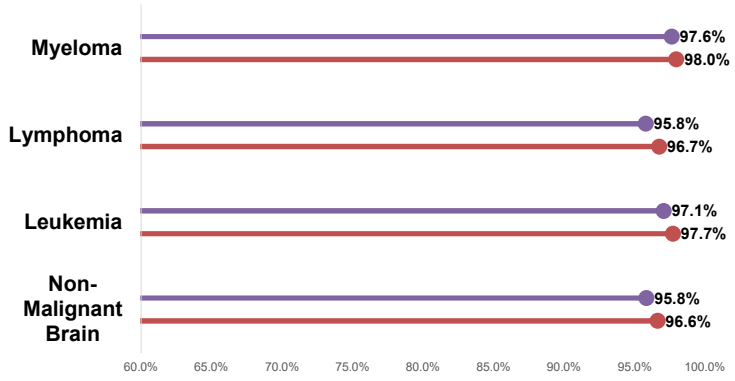
Accuracy Proportions of Major Errors & Major + Minor Errors by Primary Site



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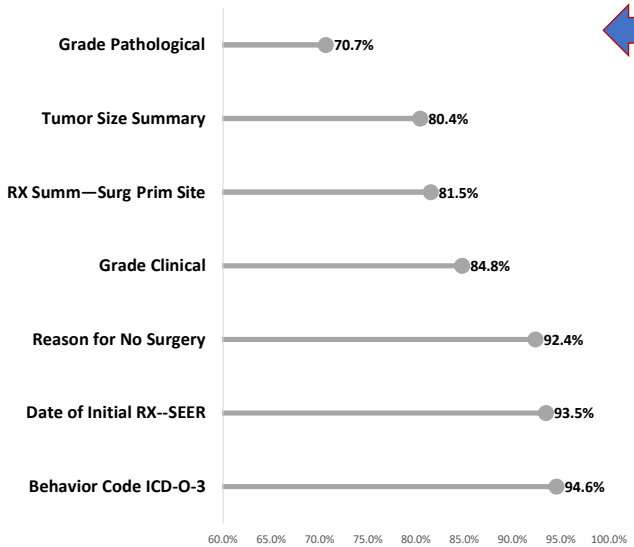
Data Elements with Lowest Accuracy Proportions: Brain (Non-Malignant)



CAP Approved — CentralNervousSystem.Bx.Res_1.0.0.1.REL_CAPCP

2. WHO Classification of Tumours Editorial Board. *WHO Classification of Tumours of the Endocrine and Neuroendocrine Tumours*, 5th Edition, Volume 9. Lyon, France: IARC Press; 2022.

H. Histologic Grade

Below is a list of possible WHO grades for CNS tumors. The WHO grading of some of the more common CNS tumors is shown in Table 1. There is no formal TNM-based classification and staging system for CNS tumors.

CNS WHO Grades for CNS Tumors
CNS WHO grade 1
CNS WHO grade 2
CNS WHO grade 3
CNS WHO grade 4
CNS WHO grade not assigned

Table 1. CNS WHO Grading System for Some of the More Common Tumors of the CNS[1,2]

Group	Type	Grade 1	Grade 2	Grade 3	Grade 4
Adult-type diffuse gliomas	Astrocytoma, IDH-mutant		X	X	X
	Oligodendroglioma, IDH-mutant and, 1p/19q co-deleted		X	X	
	Glioblastoma, IDH-wildtype				X
Pediatric-type diffuse low-grade gliomas	Diffuse glioma, MYB- or MYBL1-altered	X			
	Angiocentric glioma	X			
	Polymorphous low-grade neuroepithelial tumor of the young	X			
	Diffuse low-grade glioma, MAPK pathway-altered*				

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NPCR DQE Audit – Findings

Coding Primary 95.1%

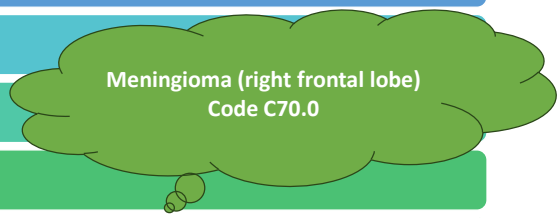
Date of diagnosis 94.4%

Histologic Type ICD-0-3 was 93.5%

Clinical Grade (Malignant) 85.%

Clinical Grade (Non-Malignant) 79.9%

RX Summ—Surgery of Primary Site 85.4%



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CNS Tumors Coding Grade

National Program of Cancer Registries (NPCR) Data Quality Corner No.4

Coding Clinical Grade (NAACCR data item #3843) for Malignant and Non-malignant Brain Tumors

Overview

Grade Clinical (NAACCR data item #3843)[1,2]:

- Is required by NPCR, CoC, and SEER.
- Is applicable for diagnosis years 2018 and forward.
- Records the grade of a solid tumor before any treatment (surgical resection or initiation of any treatment including neoadjuvant).
- Along with Grade Pathological (NAACCR data item #3844) and Grade Post-Therapy Path (NAACCR data item #3845), replaces Grade (NAACCR data item #440).
- Includes the WHO grading system specifically for brain and central nervous system (CNS) tumors, which is based on the microscopic characteristics of the tumor cell.
- Measures the tumor's aggressiveness as an important prognostic indicator for many cancers, including the brain.

Valid Codes[3]

Code	Grade 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 progression or recurrence.
3	WHO Grade III: Tumors with histologic and/or molecular genetic evidence of malignancy that are 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," not otherwise specified (NOS).
H	Stated as "high grade," NOS.
A	Well differentiated.
B	Moderately differentiated.
C	Poorly differentiated.
D	Undifferentiated, anaplastic.
9	Grade cannot be assessed; unknown.

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Table 1: Accuracy Proportion of Clinical Grade for Malignant and Non-malignant Brain Tumors

Characteristic	Malignant (gliomas and embryonal tumors only)	Non-malignant
Diagnosis years	2019	2018–2020
Number of registries	15	12
Number of cases evaluated	1057	912
Number of cases with major errors	149	183
Accuracy proportion		
Overall	85.9%	79.9%
Minimum	74.0%	66.3%
Mean	89.0%	79.4%
Maximum	95.3%	100.0%

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WHO Grade Definitions

WHO Grade	Definitions	
WHO Grade 1	Non-malignant (/0 or /1)	Circumscribed tumor of low proliferative potential associated with the possibility of cure following resection
WHO Grade 2	Malignant or Non-malignant	Infiltrative tumor with low proliferative potential with an increased risk of recurrence
WHO Grade 3	Malignant (/3)	Tumors with histologic evidence of malignancy, including nuclear atypia and mitotic activity, are associated with aggressive clinical course
WHO Grade 4	Malignant (/3)	Tumors that are cytologically malignant, with mitotic activity, are associated with rapid clinical progression and potential dissemination

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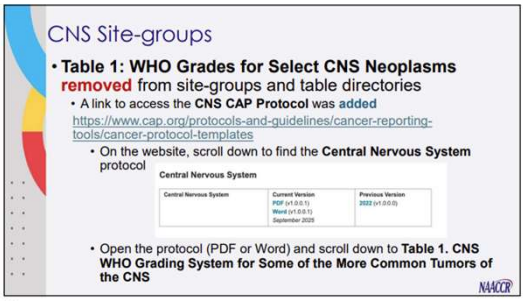
CNS WHO Grade

Table 1. CNS WHO Grading System for Some of the More Common Tumors of the CNS[1,2]

Group	Type	Grade 1	Grade 2	Grade 3	Grade 4
Adult-type diffuse gliomas	Astrocytoma, IDH-mutant		X	X	X
	Oligodendroglioma, IDH-mutant and, 1p/19q co-deleted		X	X	
	Glioblastoma, IDH-wildtype				X
Pediatric-type diffuse low-grade gliomas	Diffuse glioma, MYB- or MYBL1-altered	X			
	Angiocentric glioma	X			
	Polymorphous low-grade neuroepithelial tumor of the young	X			
	Diffuse low-grade glioma, MAPK pathway-altered*				
Pediatric-type diffuse high-grade gliomas	Diffuse midline glioma, H3 K27-altered				X
	Diffuse hemispheric glioma, H3 G34-mutant				X
	Diffuse pediatric-type high-grade glioma, H3/IDH-wildtype				X



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CNS Grade Coding Practices

- Codes 1–4 take priority over A–D, L, and H.
- CNS WHO classifications
- Code the WHO grading system for selected tumors of the CNS as noted in the AJCC 8th edition Table 72.2 when the WHO grade is not documented in the record.
- A list of the histologies that have a default grade can also be found in the Brain/Spinal Cord CAP Protocol
- **For benign tumors ONLY (behavior 0), code 1 can be automatically assigned for all histologies.**
- Code 9 when grade from primary site is not documented, clinical workup is not done (for example, cancer is an incidental finding during surgery for another condition), or grade checked “not applicable” on CAP Protocol (if available) and no other grade information is available

(https://www.cap.org/protocols-and-guidelines/cancer-reporting-tools/cancer-protocol-templates).

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Assign WHO Grade 1

- Angiocentric glioma
- Craniopharyngioma
- Choroid plexus papilloma
- Desmoplastic infantile astrocytoma and ganglioglioma
- Dysembryoplastic neuroepithelial tumor
- Dysplastic gangliocytoma of cerebellum (D=Lhemitte-Duclos)
- Gangliocytoma
- Granular cell tumor
- Hemangioblastoma
- Meningioma

WHO Grade Definitions

WHO Grade	Definition
WHO Grade I	Non-malignant (/0 or /1)
WHO Grade II	Malignant or Non-malignant
WHO Grade III	Malignant (/3) (See Malignant CNS site-group)
WHO Grade IV	Malignant (/3) (See Malignant CNS site-group)

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Assign WHO Grade 1 or 2

- Myxopapillary ependymoma
- Neurofibroma
- Oligodendroglioma IDH mutant and 1p/19q-codeleted (2)
- Papillary glioneuronal tumor
- Papillary tumor of the pineal region
- Perineuroma
- Pilocytic astrocytoma
- Pineal parenchymal tumor of intermediate differentiation
- Pineocytoma
- Pituicytoma
- Pleomorphic xanthroastrocytoma (2)
- Rosette-forming glioneuronal tumor
- Schwannoma

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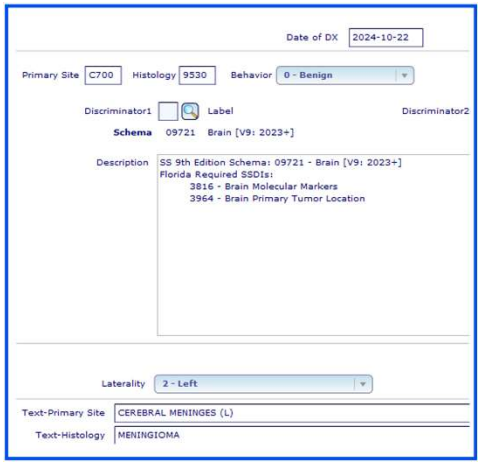
Discrepancy on Clinical Grade



10/22/24 CT HEAD: "LIKELY PARTIALLY CALCIFIED **MENINGIOMA** ALONG THE LEFT FRONTAL CONVEXITY MEASURES 5 X 6 MM. ENHANCING MASS WITHIN THE LEFT PAROTID GLAND MEASURES 1.5 X 1.1 X 2 CM. MEDIASTINAL LYMPHADENOPATHY IS PARTIALLY IMAGED, MEASURING UP TO 2 X 2.2 CM IN THE PRETRACHEAL REGION.

For benign tumors ONLY (behavior 0), code 1 can be automatically assigned for all histologies.

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Using Ambiguous Terminology

- The 'ambiguous terminology' list of words and phrases for the presence or absence of disease is applied only when 'definitive terminology' is NOT used to describe the presence or absence of a tumor or a specific histologic type/subtype
- **Ambiguous terms that do not constitute a diagnosis without additional information**
  - Suggestive
  - Concerning
  - Likely

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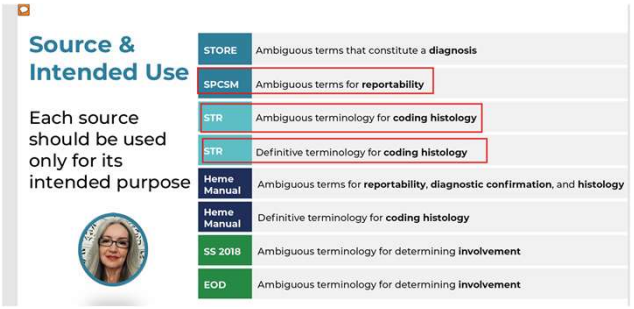
Use of Ambiguous Terminology - Reportability

Apparent(ly)
Appears
Comparable with
Compatible with
Consistent with
Favor(s)
Malignant appearing
Most likely
Presumed
Probable
Suspect(ed)
Suspicious (for)
Typical (of)



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

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Use of Ambiguous Terminology - Reportability

Equivalent to "Not diagnostic for" malignancy or reportable diagnosis. These phrases are NOT reportable when no other information is available.

- **Highly suspicious** for, but not diagnostic of [malignancy or reportable diagnosis]
- **Most compatible with** a [non-reportable diagnosis] such as a [reportable diagnosis]
- **High probability for** [malignancy or reportable diagnosis]
- **Equivalent to "**Differential diagnoses"
- Differential consideration

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Pop Quiz Reportability

1. **Patient presents to the ER on 2/3/25. CT of the head shows a lesion in the brain.**
   - A Reportable
   - B. Not Reportable
2. **The patient returns on 3/3/25 for an MRI of the brain, which shows a tumor in the right frontal lobe. The patient is referred for a biopsy.**
   - A. Reportable
   - B. Not reportable
3. **Patient presents for a stereotactic biopsy on 4/3/25. Pathology shows an astrocytoma, IDH-mutant, WHO Grade 2. What is the date of diagnosis?**
   - A. 2/3/25
   - B. 3/3/25
   - C. 4/3/25

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Pop Quiz

1. Patient presents to the ER on 2/3/25. CT of the head shows a **lesion** in the brain.
   - A Reportable
   - **B. Not Reportable**

   "Mass" and **"lesion"** are **not** reportable terms for intracranial and CNS because they are **not** listed in ICD-O-3.2 with behavior codes of /0 or /1
2. The patient returns on 3/3/25 for an MRI of the brain, which shows a **tumor** in the right frontal lobe. The patient is referred for a biopsy.
   - **A. Reportable**
   - B. Non reportable

   Neoplasm and tumor are reportable terms for intracranial and CNS because they are listed in ICD-O-3.2 with behavior codes of /0 and /1
3. Patient presents for a stereotactic biopsy on 4/3/25. Pathology shows an astrocytoma, IDH-mutant, WHO Grade 2. What is the date of diagnosis?
   - A. 2/3/25
   - **B. 3/3/25**
   - C. 4/3/25

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Treatment – Site Specific Surgery Codes

- Non-Malignant Brain and CNS Tumors

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National Program of Cancer Registries (NPCR) Data Quality Corner No. 2

Treatment Summary Surgery of Primary Site (NAACCR Data Item #1290) for Non-Malignant Brain Tumors

Overview

RX Summ—Surgical Procedure of Primary Site 03-2022 (NAACCR data item # 1290)[1]:

- Is required by NPCR, CoC, and SEER.
- Is applicable for diagnosis years 2003–2022.
- Records the surgical procedure(s) performed to the primary site during first-course treatment.
- Compares the efficacy of treatment options.
- Documents the most invasive surgical procedure for the primary site if only one procedure is collected in the registry software.
- Was replaced in 2023 by RX Summ – Surg 2023 (NAACCR data item #1291) to capture code structure, not to the descriptions or definitions for brain/CNS sites.

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Valid Codes (RX Summ—Surgery of Primary Site 03-2022 for diagnosis years 2003–2022)

No specimen sent to pathology	Specimen submitted to pathology	Unknown surgery
00 – No surgery **10** – Tumor destruction, NOS	**20** – Local excision of tumor, lesion, or mass; excisional biopsy • **21** – Subtotal resection of tumor, lesion, or mass in brain • **22** – Resection of tumor of spinal cord or nerve **30** – Radical, total, gross resection of tumor, lesion, or mass in 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)	**90** – Surgery, NOS; type unknown **99** – Unknown if surgery performed, death certificate ONLY

Valid Codes (RX Summ – Surg 2023 for diagnosis years 2023+)

No specimen sent to pathology	Specimen submitted to pathology	Unknown surgery
A000 – No surgery **A100** – Tumor destruction, NOS	**A200** – Local excision of tumor, lesion, or mass; excisional biopsy • **A210** – Subtotal resection of tumor, lesion, or mass in brain • **A220** – Resection of tumor of spinal cord or nerve **A300** – Radical, total, gross resection of tumor, lesion, or mass in brain **A400** – Partial resection of lobe of brain when surgery can't	**A900** – Surgery, NOS **A990** – Unknown if surgery performed, death certificate ONLY

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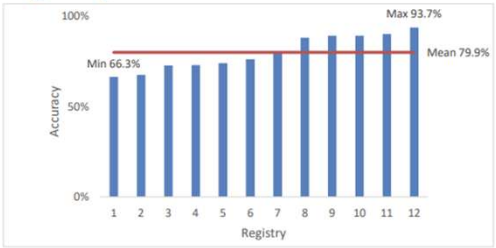
NPCR DQE Audit – Coding Surgery

Figure 1: Accuracy of RX Summ -- Surgery of Primary Site by registry, diagnosis years 2018–2020



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Site Specific Surgery Codes - Brain

- **2018-2020 Surgery Codes**
- Use code 20 if no description of the procedure is available; includes excisional biopsy; for *SEER registries, assign 20 for Stereotactic Biopsy of the Brain*
- Use code 21 when only a subtotal resection is performed
- Use code 30 when a radical, total or gross resection of tumor is performed

- **2026 Surgery Codes**
- A200
- A210 subtotal resection
- A220 resection of tumor of spinal cord or nerve
- A300 Radical, total gross resection of tumor, lesion or mass in brain
- A400 Partial resection of lobe of brain, when the surgery cannot be coded as A200-A300

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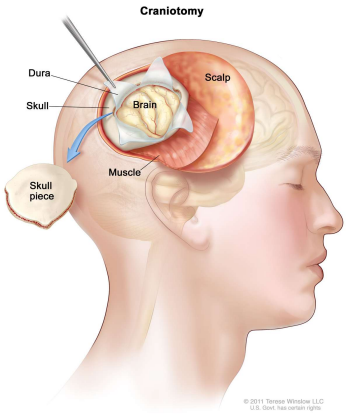
Brain and CNS Tumors –Cancer Directed Surgery Coding

- A000 None; no surgery of primary site; autopsy ONLY
- A100 Tumor destruction, NOS

[SEER Note: Local tumor destruction, NOS; laser interstitial thermal therapy (LITT) - code A100 if no specimen sent to pathology.]

- No specimen sent to pathology from surgical event A100
- **Do not record stereotactic radiosurgery** (SRS), Gamma knife, Cyber knife, or Linac radiosurgery as surgical tumor destruction. All of these modalities are recorded in the radiation treatment fields



https://seer.cancer.gov/manuals/2024/appendixc.html

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SEER Program Coding and Staging Manual 2025

Surgery Codes

Brain [and other parts of central nervous system]
Meninges C700-C709,
Brain C710–C719,
Spinal Cord, Cranial Nerves and Other Parts of Central Nervous System
C720-C729

Do not code laminectomies for spinal cord primaries.

Codes

A000 None; no surgery of primary site; autopsy ONLY

A100 Tumor destruction, NOS
[***SEER Note:*** Local tumor destruction, NOS; laser interstitial thermal therapy (LITT) - code A100 if no specimen sent to pathology.]
No specimen sent to pathology from surgical event A100

Do not record stereotactic radiosurgery (SRS), Gamma knife, Cyber knife, or Linac radiosurgery as surgical tumor destruction. All of these modalities are recorded in the radiation treatment fields.

A200 Local excision of tumor, lesion, or mass, excisional biopsy
A210 Subtotal resection of tumor, lesion or mass in brain
A220 Resection of tumor in spinal cord or nerve
[***SEER Note:*** Assign code A200 for stereotactic biopsy of brain tumor. This includes a Stealth or StealthStation guided needle biopsy, a type of stereotactic biopsy.]

STORE 2025 *APPENDIX A: Site-Specific Surgery Codes*

Brain

Meninges C70.0–C70.9, Brain C71.0–C71.9,
Spinal Cord, Cranial Nerves and Other Parts of Central Nervous System C72.0–C72.9

Do not code laminectomies for spinal cord primaries.

Codes
A000 None; no surgery of primary site

A100 Tumor destruction, NOS
No specimen sent to pathology from surgical event A100.

Do not record stereotactic radiosurgery (SRS), Gamma knife, Cyber knife, or Linac radiosurgery as surgical tumor destruction. All of these modalities are recorded in the radiation treatment fields.

A200 Local excision of tumor, lesion or mass; excisional biopsy
A210 Subtotal resection of tumor, lesion or mass in brain
A220 Resection of tumor of spinal cord or nerve

Stereotactic Biopsy of the Brain

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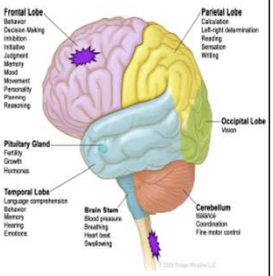
Surgery Codes

Code	Procedure	Specifics
A200	Local excision of tumor, lesion or mass; excisional biopsy	Used when the surgeon describes the procedure "biopsy," or "excisional biopsy", or when there are no details about the procedure. Unknown whether total or partial tumor resected.
A210	Subtotal resection of tumor, lesion or mass in brain	Near total, partial, subtotal, debulking, open biopsy (if residual tissue).
A220	Resection of tumor of spinal cord nerve	
A300	Radical, total, gross resection of tumor, lesion or mass in brain	The resection of the brain tissue surrounding the tumor is limited to ensure clean margins.



NAACCR

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CNS Cancer Directed Surgery

- A550 Gross total resection of lobe of brain (lobectomy)
- Codes A300-A550 are not applicable for spinal cord or spinal nerve primary sites.
- Specimen sent to pathology from surgical events A200-A550
- A900 Surgery, NOS

[SEER Note: Laser interstitial thermal therapy (LITT) - code A900 if specimen sent to pathology.]

- A990 Unknown if surgery performed; death certificate ONLY

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Tumor Size Summary

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- Record the most accurate measurement of a solid tumor, usually on the surgical resection specimen
- Coding priority order
- STORE manual 2026, page 121, Record size in specified order Rule #3. **If no surgical resection, then code the largest measurement of tumor from imaging**, physical exam, imaging, or other diagnostic procedures in this order of priority before any other form of treatment
- Imaging can be used to code tumor size if no information is available from pathology or operative report

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Brain

Meninges C70.0–C70.9, Brain C71.0–C71.9,
Spinal Cord, Cranial Nerves and Other Parts of Central Nervous System C72.0–C72.9

Do not code laminectomies for spinal cord primaries.

Codes
A000 None; no surgery of primary site

A100 Tumor destruction, NOS
No specimen sent to pathology from surgical event A100.

Do not record stereotactic radiosurgery (SRS), Gamma knife, Cyber knife, or Linac radiosurgery as surgical tumor destruction. All of these modalities are recorded in the radiation treatment fields.

A200 Local excision of tumor, lesion or mass; excisional biopsy
A210 Subtotal resection of tumor, lesion or mass in brain
A220 Resection of tumor of spinal cord or nerve

A300 Radical, total, gross resection of tumor, lesion or mass in brain

A400 Partial resection of lobe of brain, when the surgery cannot be coded as A200-A300.

A550 Gross total resection of lobe of brain (lobectomy)
Codes A300-A550 are not applicable for spinal cord or spinal nerve primary sites.
Specimen sent to pathology from surgical events A200–A550.

A900 Surgery, NOS

A990 Unknown if surgery performed

Coding LITT for Glioblastoma Brain

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- **Laser interstitial thermal therapy**, or LiTT, is a minimally invasive procedure that uses heat to destroy abnormal cells, such as tumors or those causing seizures.
- It is sometimes called laser ablation surgery, stereotactic laser ablation, or MRI-guided laser ablation

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Case 1 Non-Malignant CNS Tumor Question

NPCR DQE Audit

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Case 1 – Question 1 NPCR DQE Audit

- **04/01/2021**- CT HEAD- LARGE HYPODENSE CYSTIC 4.1 X 3.8 X 3.0 CM LESION INVOLVING SUPEROMEDIAL RIGHT CEREBELLUM, VERMIS CYSTIC LESION LIKELY REPRESENTS NEOPLASTIC ETIOLOGY SUCH AS PRIMARY CYSTIC BRAIN MASS OR CYSTIC METASTASIS. **04/01/2021-** MRI BRAIN- PRIMARILY CYSTIC MASS WITHIN THE CEREBELLUM MEASURES 4.2 X 3.2 X 3.7 CM AND APPEARS TO SHOW A SMALL NODULE OF ENHANCEMENT POSTERIORLY; 2ND SOLID-APPEARING ENHANCING LESION WITHIN THE INFERIOR MIDLINE CEREBELLUM MEASURING 0.7 X 0.5 X 0.8 CM

- **04/12/21** BRAIN TUMOR XS 2 SUBOCCIPITAL CRANIOTOMY FOR RESECTION OF **CEREBELLAR** BRAIN TUMORS HEMANGIOBLASTOMA IS FAVORED, VHL DELETION, **08/26/2021**- CRANIOTOMY FOR SRS, BRAIN **SRS**, 001600CGY FOR 1FX

- **What is the date of diagnosis?**
  - A. 4/1/2021
  - B. 4/12/2021

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Case 1 – Question 1 – Correct Answer NPCR DQE Audit

- **01/01/2021**- CT HEAD- LARGE HYPODENSE CYSTIC 4.1 X 3.8 X 3.0 CM **LESION** INVOLVING SUPEROMEDIAL RIGHT CEREBELLUM, VERMIS AS NEW FROM PRIOR STUDY, WITH RESULTING MASS EFFECT, EFFACEMENT OF FOURTH VENTRICLE. CYSTIC LESION LIKELY REPRESENTS NEOPLASTIC ETIOLOGY SUCH AS PRIMARY CYSTIC BRAIN MASS OR CYSTIC METASTASIS.

- **04/12/21** BRAIN TUMOR XS 2 SUBOCCIPITAL CRANIOTOMY FOR RESECTION OF **CEREBELLAR** BRAIN TUMORS HEMAGIOBLASTOMA IS FAVORED, VHL DELETION, **08/26/2021**- CRANIOTOMY FOR SRS, BRAIN **SRS**, 001600CGY FOR 1FX

Data Items	Coded	Recoded
Date of Diagnosis	20210401	20210412

The correct answer is B for date dx 4/12/2021

Rationale. Likely is an ambiguous term that does not constitute a diagnosis of cancer. The date of diagnosis is the date of surgery when the diagnosis was confirmed to be hemangioblastoma

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Case 1 – Question 2
NPCR DQE Audit

- **01/01/2021**- CT HEAD- LARGE HYPODENSE CYSTIC 4.1 X 3.8 X 3.0 CM LESION INVOLVING SUPEROMEDIAL RIGHT CEREBELLUM, CYSTIC LESION LIKELY REPRESENTS NEOPLASTIC ETIOLOGY SUCH AS PRIMARY CYSTIC BRAIN MASS OR CYSTIC METASTASIS. **04/01/2021**- HCH- MRI BRAIN- PRIMARILY CYSTIC MASS WITHIN THE CEREBELLUM MEASURES 4.2 X 3.2 X 3.7 CM AND APPEARS TO SHOW A SMALL NODULE OF ENHANCEMENT POSTERIORLY; 2ND SOLID-APPEARING ENHANCING LESION WITHIN THE INFERIOR MIDLINE CEREBELLUM MEASURING 0.7 X 0.5 X 0.8 CM. **04/12/21** BRAIN TUMOR XS 2 SUBOCCIPITAL CRANIOTOMY FOR RESECTION OF CEREBELLAR BRAIN TUMORS HEMAGIOBLASTOMA IS FAVORED, VHL DELETION

- **What is coded for tumor size summary?**
  - A. 999
  - B. 042
  - C. 038

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Case 1 – Question 2 – Correct Answer
NPCR DQE Audit

01/01/2021- CT HEAD- LARGE HYPODENSE CYSTIC 4.1 X 3.8 X 3.0 CM LESION INVOLVING SUPEROMEDIAL RIGHT CEREBELLUM, CYSTIC LESION LIKELY REPRESENTS NEOPLASTIC ETIOLOGY SUCH AS PRIMARY CYSTIC BRAIN MASS OR CYSTIC METASTASIS. **04/01/2021**- HCH- MRI BRAIN- PRIMARILY CYSTIC MASS WITHIN THE CEREBELLUM MEASURES **4.2** X 3.2 X 3.7 CM AND APPEARS TO SHOW A SMALL NODULE OF ENHANCEMENT POSTERIORLY; 2ND SOLID-APPEARING ENHANCING LESION WITHIN THE INFERIOR MIDLINE CEREBELLUM MEASURING 0.7 X 0.5 X 0.8 CM.

04/12/21 BRAIN TUMOR XS 2 SUBOCCIPITAL CRANIOTOMY FOR RESECTION OF CEREBELLAR BRAIN TUMORS HEMAGIOBLASTOMA IS FAVORED, VHL DELETION

Data Items	Coded	Recoded
Tumor Size Summary	999	042

The correct answer is B. 042 for Tumor Size Summary

Recoded from 999 to 042 when the largest tumor size is from a brain MRI study which indicates that patient has multiple brain masses and the largest is 4.2 cm. **STORE manual 2026, page 121**, Information on size from imaging/radiographic techniques can be used to code the tumor size when there is no more specific size information from pathology or operative report.

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Case 1 – Question 3
NPCR DQE Audit

- **01/01/2021**- CT HEAD- LARGE HYPODENSE CYSTIC 4.1 X 3.8 X 3.0 CM LESION INVOLVING SUPEROMEDIAL RIGHT CEREBELLUM, VERMIS AS NEW FROM PRIOR STUDY, WITH RESULTING MASS EFFECT, EFFACEMENT OF FOURTH VENTRICLE. CYSTIC LESION LIKELY REPRESENTS NEOPLASTIC ETIOLOGY SUCH AS PRIMARY CYSTIC BRAIN MASS OR CYSTIC METASTASIS.

- **04/12/21** BRAIN TUMOR XS 2 SUBOCCIPITAL CRANIOTOMY FOR RESECTION OF **CEREBELLAR** BRAIN TUMORS HEMAGIOBLASTOMA IS FAVORED, VHL DELETION, **08/26/2021**- CRANIOTOMY FOR Stereotactic Radiosurgery, BRAIN **SRS**, 001600CGY FOR 1FX

- **What is coded for radiation modality?**
  - A. 02, photons
  - B. 01, external beam, NOS
  - C. 00, none

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Case 1 – Question 3- Correct Answer
NPCR DQE Audit

- **01/01/2021**- CT HEAD- LARGE HYPODENSE CYSTIC 4.1 X 3.8 X 3.0 CM LESION INVOLVING SUPEROMEDIAL RIGHT CEREBELLUM, VERMIS AS NEW FROM PRIOR STUDY, WITH RESULTING MASS EFFECT, EFFACEMENT OF FOURTH VENTRICLE. CYSTIC LESION LIKELY REPRESENTS NEOPLASTIC ETIOLOGY SUCH AS PRIMARY CYSTIC BRAIN MASS OR CYSTIC METASTASIS.

- **04/12/21** BRAIN TUMOR XS 2 SUBOCCIPITAL CRANIOTOMY FOR RESECTION OF **CEREBELLAR** BRAIN TUMORS HEMAGIOBLASTOMA IS FAVORED, VHL DELETION, **08/26/2021**- CRANIOTOMY FOR SRS, BRAIN **SRS**, 001600CGY FOR 1FX

Data Items	Coded	Recoded
Phase I Radiation Treatment Modality	00	02

The Correct Answer was A. Code 02

Recoded from 00 to 02 when text fields indicate that stereotactic radiosurgery (SRS) was done on 08/26/2021. SRS is coded as radiation.

CTR Guide to Coding Radiation Therapy Treatment in the STORE, states that SRS modality is coded to 02 (photons). [SPCSM 2021, pages 195-196]

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Case 1 – Summary NPCR DQE Audit

01/01/2021- CT HEAD- LARGE HYPODENSE CYSTIC 4.1 X 3.8 X 3.0 CM LESION INVOLVING SUPEROMEDIAL RIGHT CEREBELLUM, VERMIS AS NEW FROM PRIOR STUDY, WITH RESULTING MASS EFFECT, EFFACEMENT OF FOURTH VENTRICLE. CYSTIC LESION LIKELY REPRESENTS NEOPLASTIC ETIOLOGY SUCH AS PRIMARY CYSTIC BRAIN MASS OR CYSTIC METASTASIS.

04/12/21 BRAIN TUMOR XS 2 SUBOCCIPITAL CRANIOTOMY FOR RESECTION OF **CEREBELLAR** BRAIN TUMORS HEMAGIOBLASTOMA IS FAVORED, VHL DELETION, **08/26/2021**- CRANIOTOMY FOR SRS, BRAIN **SRS**, 001600CGY FOR 1FX

Data Items	Coded	Recoded
RX Summ—Surg/Rad Seq	0	3
Reason No Radiation	1	0
Phase I Radiation Treatment Modality	00	02
Grade Clinical	L	9
Grade Pathological	L	1
Date of Diagnosis	20210401	20210412
Primary Site	C719	C716
Tumor Size Summary	999	042

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Case 2 Non-Malignant CNS Tumors Questions

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Case 2 – Question 1
NPCR DQE Audit

- 67-YEAR-OLD SINGLE WHITE NON-HISPANIC MALE WHO WAS FOUND TO HAVE A BRAINSTEM CAVERNOMA, 5/28/2021 MRI- BRAINSTEM CAVERNOMA 12 MM 6/13/2021 CT BRAIN - CAVERNOMA 15 MM

- 6/14/2021 BRAIN TUMOR EXCISION -SAW BULGING EXOPHYTIC CAVERNOMA WITH DARK BLOOD TINGE ON THE PART THAT WAS BULGING AND THEN THE FLOOR OF THE 4TH VENTRICLE HAD HEMOSIDERIN IN IT, 6/14/2021 PATHOLOGY BRAIN TUMOR EXCISION – CAVERNOMA

- **What is the Primary Site Code?**
  - A. C712
  - B. C717
  - C. C710

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Case 2 – Question 1 Answer
NPCR DQE Audit

67-YEAR-OLD SINGLE WHITE NON-HISPANIC MALE WHO WAS FOUND TO HAVE A BRAINSTEM CAVERNOMA

5/28/2021 MRI- BRAINSTEM CAVERNOMA 12 MM 6/13/2021 CT BRAIN - CAVERNOMA 15 MM

6/14/2021 BRAIN TUMOR EXCISION -SAW BULGING EXOPHYTIC CAVERNOMA WITH DARK BLOOD TINGE ON THE PART THAT WAS BULGING AND THEN THE FLOOR OF THE 4TH VENTRICLE HAD HEMOSIDERIN IN IT, 6/14/2021 PATHOLOGY BRAIN TUMOR EXCISION - CAVERNOMA

Data Items	Coded	Recoded
Primary Site	C710	C717

The Correct Answer is B. C717 Brain stem

Brain	Cerebrum	C710
	Frontal lobe	C711
	Temporal lobe	C712
	Parietal lobe	C713
	Occipital lobe	C714
	Ventricle, NOS	C715
	Cerebellum, NOS	C716
	Brain stem	C717

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Case 2 – Question 2
NPCR DQE Audit

- 67-YEAR-OLD SINGLE WHITE NON HISPANIC MALE WHO WAS FOUND TO HAVE A BRAINSTEM CAVERNOMA, 5/28/2021 MRI- BRAINSTEM CAVERNOMA 12 MM 6/13/2021 CT BRAIN - CAVERNOMA 15 MM

- 6/14/2021 BRAIN TUMOR EXCISION -SAW BULGING EXOPHYTIC CAVERNOMA WITH DARK BLOOD TINGE ON THE PART THAT WAS BULGING AND THEN THE FLOOR OF THE 4TH VENTRICLE HAD HEMOSIDERIN IN IT, 6/14/2021 PATHOLOGY BRAIN TUMOR EXCISION – CAVERNOMA

- **The Pathologic Grade should be coded 1?**
  - True
  - False

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Case 2 – Question 2 – Answer
NPCR DQE Audit

67-YEAR-OLD SINGLE WHITE NON-HISPANIC MALE WHO WAS FOUND TO HAVE A BRAINSTEM CAVERNOMA

5/28/2021 MRI- BRAINSTEM CAVERNOMA 12 MM 6/13/2021 CT BRAIN - CAVERNOMA 15 MM

6/14/2021 BRAIN TUMOR EXCISION -SAW BULGING EXOPHYTIC CAVERNOMA WITH DARK BLOOD TINGE ON THE PART THAT WAS BULGING AND THEN THE FLOOR OF THE 4TH VENTRICLE HAD HEMOSIDERIN IN IT, 6/14/2021 PATHOLOGY BRAIN TUMOR EXCISION - CAVERNOMA

Data Items	Coded	Recoded
Grade Pathological	1	9

The correct answer is False

Note 8: Code 9 (unknown) when
- Grade from primary site is not documented
- No resection of the primary site (see exception in Note 7, Surgical resection, last bullet)
- Neo-adjuvant therapy is followed by a resection (see Grade Post Therapy Path (yp))
- Grade checked "not applicable" on CAP Protocol (if available) and no other grade information is available
- Clinical case only (see Grade Clinical)

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Case 2 – Question 3
NPCR DQE audit

- 67-YEAR-OLD SINGLE WHITE NON-HISPANIC MALE WHO WAS FOUND TO HAVE A BRAINSTEM CAVERNOMA, 5/28/2021 MRI- BRAINSTEM CAVERNOMA 12 MM 6/13/2021 CT BRAIN - CAVERNOMA 15 MM

- 6/14/2021 BRAIN TUMOR EXCISION -SAW BULGING EXOPHYTIC CAVERNOMA WITH DARK BLOOD TINGE ON THE PART THAT WAS BULGING AND THEN THE FLOOR OF THE 4TH VENTRICLE HAD HEMOSIDERIN IN IT, 6/14/2021 PATHOLOGY BRAIN TUMOR EXCISION – CAVERNOMA

- **What is Coded for Cancer Directed Surgery?**
  - A. 10, tumor destruction
  - B. 20, local excision of tumor (subtotal resection)
  - C. 30, radical, total, gross resection tumor, lesion, mass

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Case 2 – Question 3 – Answer
NPCR DQE Audit

67-YEAR-OLD SINGLE WHITE NON-HISPANIC MALE WHO WAS FOUND TO HAVE A BRAINSTEM CAVERNOMA

5/28/2021 MRI- BRAINSTEM CAVERNOMA 12 MM 6/13/2021 CT BRAIN - CAVERNOMA 15 MM

6/14/2021 BRAIN TUMOR EXCISION -SAW BULGING EXOPHYTIC CAVERNOMA WITH DARK BLOOD TINGE ON THE PART THAT WAS BULGING AND THEN THE FLOOR OF THE 4TH VENTRICLE HAD HEMOSIDERIN IN IT, 6/14/2021 PATHOLOGY BRAIN TUMOR EXCISION - CAVERNOMA

Data Items	Coded	Recoded
RX Summ—Surg Prim Site	30	20

The correct answer is B. 20.

A100 Tumor destruction, NOS

No specimen sent to pathology from surgical event A100.

Do not record stereotactic radiosurgery (SRS), Gamma knife, Cyber knife, or Linac radiosurgery as surgical tumor destruction. All of these modalities are recorded in the radiation treatment fields.

A200 Local excision of tumor, lesion or mass; excisional biopsy

A210 Subtotal resection of tumor, lesion or mass in brain

A220 Resection of tumor of spinal cord or nerve

A300 Radical, total, gross resection of tumor, lesion or mass in brain

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Case 2 – Summary NPCR DQE Audit

Data Items	Coded	Recoded
RX Summ—Surg Prim Site	30	20
Grade		
Pathological	1	9
Primary Site	C710	C717

- 67-YEAR-OLD SINGLE WHITE NON-HISPANIC MALE WHO WAS FOUND TO HAVE A BRAINSTEM CAVERNOMA
- 5/28/2021 MRI- BRAINSTEM CAVERNOMA 12 MM 6/13/2021 CT BRAIN - CAVERNOMA 15 MM
- 6/14/2021 BRAIN TUMOR EXCISION -SAW BULGING EXOPHYTIC CAVERNOMA WITH DARK BLOOD TINGE ON THE PART THAT WAS BULGING AND THEN THE FLOOR OF THE 4TH VENTRICLE HAD HEMOSIDERIN IN IT, 6/14/2021 PATHOLOGY BRAIN TUMOR EXCISION - CAVERNOMA

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Sequence – Non-Malignant CNS Tumors

- Assign sequence number 60 when there are no prior or subsequent non-malignant intracranial/ CNS tumors
- If another reportable non-malignant tumor, change the sequence number of the first primary from 60 to 61
- Sequence multiple non-malignant tumors chronologically as 61 (first of two or more), 62 (second), etc.

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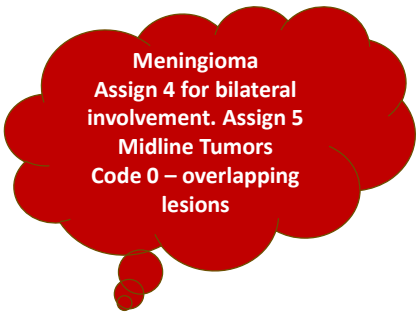
Brain and CNS Sites (require Laterality)



- C. 47.1-C47.2 Peripheral nerves
- C70.0 Cerebral meninges, NOS
- C71.0 – C714 Cerebrum, frontal lobe, temporal lobe, parietal, occipital lobes
- C72.2 – C72.5 Olfactory, optic, Acoustic and cranial nerves
- **** excludes diagnoses before 2004***

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Coding Laterality C716 Cerebellum

- Primary tumors arising in the left cerebellum (C716). The cerebellum is not a lateral brain site. The coding instructions for Laterality indicate, "Assign code 0 when the primary site is not a paired site."
- While some brain subsites are lateral sites, C716 is not among them.
- The Sites for Which Laterality Codes Must Be Recorded (2026 SEER Manual) and Table 5: Paired Sites (Solid Tumor Rules Manual, Malignant CNS) both confirm C716 is not a site for which laterality is coded.
- Code the Laterality as 0 (Not a paired site).

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SSDI Data Items – Brain and CNS Tumors (2023)

- Grade
  - Clinical, Pathologic, Post Therapy Clin, Post Therapy Path
- Brain Molecular Markers
- Chromosome 1p Status
- Chromosome 19q Status
- MGMT
- Brain Primary Tumor Location (2024)

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Brain and CNS Tumor SSDI – Required for FCDS

DS Requires the Following SSDIs for Cases Diagnosed/Treated 2018 and Forward:

Core/Derived	Item #	Item Name	Length	S
C	1068	Grade Post Therapy Clin (yc)	2	
D	3800	Schema ID	5	
C	1172	Post Transplant Lymphoproliferative Disorder (PTLD)	1	
C	1174	PD-L1	5	
C	3816	Brain Molecular Markers	2	
C	3817	Breslow Tumor Thickness	4	
C	3827	Estrogen Receptor Summary	1	
C	3829	Esophagus and EGJ Tumor Epicenter	1	
C	3835	Fibrosis Score	1	
C	3838	Gleason Patterns Clinical	2	
C	3839	Gleason Patterns Pathological	2	
C	3840	Gleason Score Clinical	2	
C	3841	Gleason Score Pathological	2	
C	3842	Gleason Tertiary Pattern	2	
C	3843	Grade Clinical	1	
C	3844	Grade Pathological	1	
C	3845	Grade Post Therapy Path (yp)	1	
C	3855	HER2 Overall Summary (breast)	1	
C	3890	Microsatellite Instability (MSI)	1	
C	3915	Progesterone Receptor Summary	1	
C	3920	PSA (Prostatic Specific Antigen) Lab Value	5	
C	3932	LDH Pretreatment Lab Value	7	
C	3956	P16 (cervix, anus, vulva)	2	
C	3960	Histologic Subtype (appendix)	1	
C	3964	Brain Primary Tumor Location	1	

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Diffuse Low Grade Glioma MAPK Pathway altered - Benign

08/17/2025 - Path Report #1

FINAL DIAGNOSIS

A, B. BRAIN, DESIGNATED AS "RIGHT OCCIPITAL TUMOR", RESECTION:

INTEGRATED DIAGNOSIS: Diffuse low-grade glioma, MAPK pathway-altered (see Comment).
HISTOLOGICAL CLASSIFICATION: Glial neoplasm
MOLECULAR INFORMATION:
IDH: Negative for IDH1/2 mutations by sequencing
1p/19q: Negative for co-deletion by fluorescence in situ hybridization
FGFR1 FUSION: Positive for activating alteration by sequencing
MGMT PROMOTER METHYLATION: Positive for MGMT gene promoter methylation by methylation specific PCR

This SSDI does not apply to all brain and CNS histologies. Assign code 85 if the histology does not apply. **For benign or borderline, assign Code 86.**

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EOD Primary Tumor

EOD Primary Tumor

- Benign and Borderline tumors **are always coded 050**
- Regional nodes examined 99
- Regional nodes positive 99
- SS2018 – are always coded to 8

- **Brain Molecular Markers**
  - **Note 1:** Physician statement of histologic subtype can be used to code this data item when no other information is available
  - **Code 86** when there is a benign (/0) or borderline (/1) tumor
  - *Excludes: 9421/1 (see codes 19-20 when microscopically confirmed)*

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Non-Malignant Solid Tumor Rules

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Review Solid Tumor Rules: Non-Malignant CNS Tumors

Introduction

Note 1: This site group **includes** the following **primary sites**: 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: Malignant CNS neoplasms have a separate set of rules.

Note 3: Non-malignant central nervous system (CNS) neoplasms (previously called benign and borderline) are **reportable** for cases **diagnosed 1/1/2004** and later.

Note 4: **Pilocytic astrocytoma/juvenile pilocytic astrocytoma:**

- For cases diagnosed prior to 1/1/2023, these neoplasms are reportable in North American as malignant 9421/3 for all CNS sites with the exception of the optic nerve:
  - WHO Classification Tumors of the Central Nervous System and IARC designate pilocytic astrocytoma as a synonym for optic glioma
  - When the primary site is optic nerve and the diagnosis is either optic glioma or pilocytic astrocytoma, the behavior is non-malignant and coded 9421/1
  - Beginning with cases diagnosed 1/1/2023 forward, pilocytic astrocytoma/juvenile pilocytic astrocytoma are to be reported as 9421/1 for ***all*** CNS sites.

Note 5: Neurofibromatosis NOS, Neurofibromatosis 1 (NF1), Neurofibromatosis 2 (NF2), and schwannomatosis are familial tumor syndromes and are not reportable conditions. People with NF1 and NF2 have a high risk of developing reportable and non-reportable tumors. Tumors associated with NF1 and NF2 are reportable when they meet the behavior (/0 or /1), site (within the CNS), and histology reportability requirements (see Reportability Criteria).

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Non-Malignant CNS and Peripheral Nerves

- **Note 5**: Pilocytic astrocytoma/juvenile pilocytic astrocytoma Beginning with cases diagnosed 1/1/2023 forward, pilocytic astrocytoma/juvenile pilocytic astrocytoma are to be reported as 9421/1 for ***all*** CNS sites
  - When the primary site is optic nerve (C723) and the diagnosis is either optic glioma or pilocytic astrocytoma, the behavior is non-malignant and coded 9421/1
- Neurofibromatosis NOS, Neurofibromatosis 1 (NF1), Neurofibromatosis 2 (NF2), and schwannomatosis are familial tumor syndromes and **are not reportable conditions**

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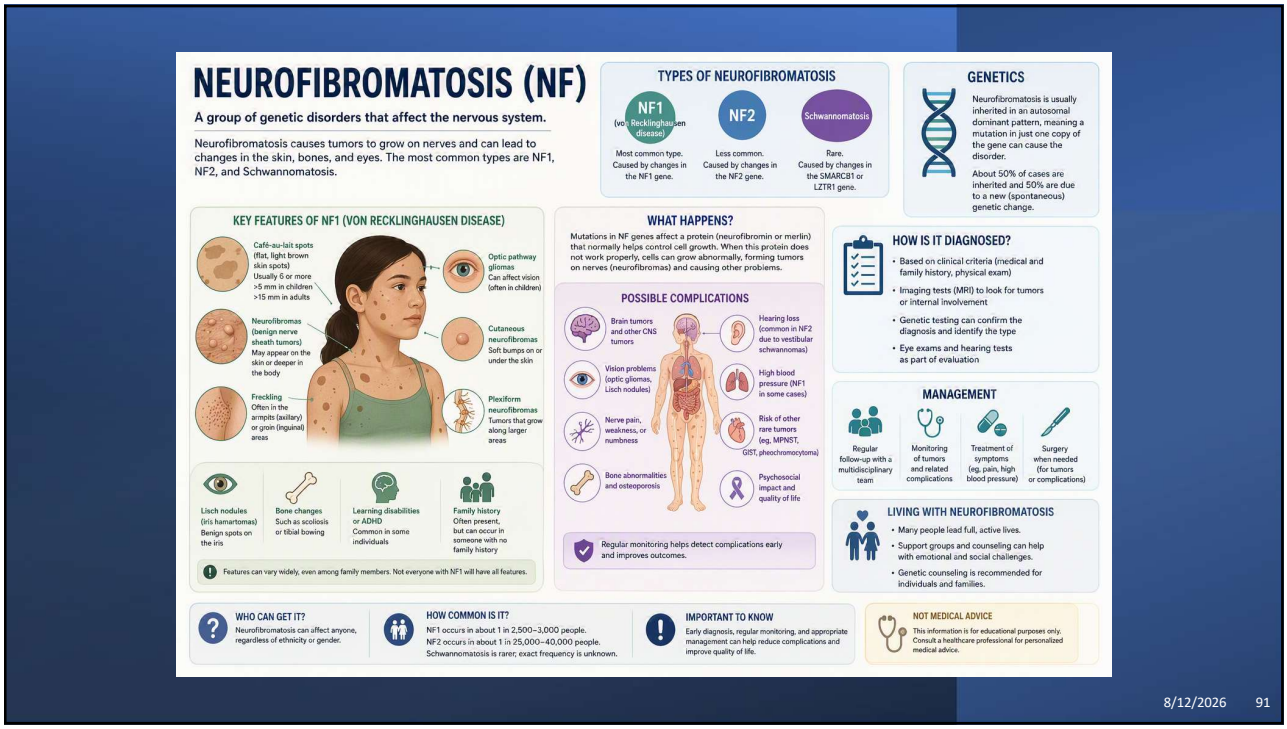

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Non-Malignant CNS Site Group Non-Reportable Neoplasms

Non-Malignant CNS Site-group Instructions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Table 3: Non-Reportable Neoplasms

Non-reportable Histology Term	Non-reportable Histology Code	Definitions and Sites
Neurofibromatosis, NOS [1]	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) [1]	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) [1]	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.
Neuroglial cyst	No code	Ventricles
Osteochondroma	9210/0	Originates in the cartilage around bone, site not reportable for non-malignant neoplasms
Rathke cleft cyst	No code	Sella turcica C751. This is a Rathke cleft CYST, not a Rathke cleft tumor.
Schwannomatosis [1]	No code	A form of neurofibromatosis newly named/discovered

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CNS Solid Tumor Rules – Coding Neurofibromatosis

Rule H2 (single tumor)	Code the reportable CNS tumor (Table 5 in the Site-group Instructions) when a patient has any of the following: • Neurofibromatosis type 1 (NF1) • Neurofibromatosis type 2 (NF2) • Schwannomatosis
Rule H5	Code meningioma 9530/0 when there are multiple meningiomas with the following diagnosis • Benign meningioma • Lymphoplasmacyte-rich meningioma • Meningioma with no mention of behavior • Metaplastic meningioma • Microcystic meningioma • Secretory meningioma
Rule H7	Code the reportable CNS tumor (Table 5 in the Site-group Instructions) when a patient has any of the following: • Neurofibromatosis type 1 (NF1), • Neurofibromatosis type 2 (NF2) • Schwannomatosis

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Non-Malignant CNS and Peripheral Nerves

- The terms tumor, mass, tumor mass, lesion, and neoplasm are not used in a standard manner in clinical diagnoses, scans, or consults. **Disregard the terms unless there is a physician's statement that the term is a non-malignant**.
- **Tumor/neoplasm**
- o These terms are used ONLY for determining multiple primaries
- o DO NOT USE these terms for casefinding or determining reportability

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Non-Malignant CNS Tumors Reportability

CNS neoplasms must meet all three criteria/conditions below to be reported as non-malignant:

1. The behavior must non-malignant /0 or /1.
   A. Pathology designates the tumor as non-malignant (/0 or /1) OR
   B. Diagnostic imaging definitively states the tumor as non-malignant (/0 or/1)
   C. The tumor is a WHO Grade I (See Section 1: Table 1)

Note 1: Always code the behavior code reported by the pathologist.

Note 2: Never report a malignant /3 behavior code for a meningioma based on tumor extension to brain, skin of scalp, or other regional organs/tissue. Non-malignant CNS tumors can extend to the regional tissue and bone. Tumor extension is usually equivalent to atypical meningioma 9539/1.

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Priorities for Coding Primary Site

- Use the list below in **hierarchical order**:
- 1. **Resection**
  - A. Operative report(s)
  - B. Pathology report(s)
- 2. **Biopsy**
  - A. Operative report(s)
  - B. Pathology report(s)
- 3. Resection and/or biopsy performed, but operative report(s) and pathology are not available (minimal information):
  - A. Tumor Board
  - B. Code from physician's documentation of **original diagnosis** from operative or pathology report
  - C. Physician's **documentation of primary site** in the medical record

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Priority Order for Cases Diagnosed by Imaging

- For cases diagnosed by imaging (no pathology/resection or biopsy) use information from scans in the following priority order:
  - A. MRI
  - B. CT
  - C. PET
  - D. Angiogram

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Non-Reportable Neoplasms Brain

Use **Table 4** for **non-malignant neoplasms ONLY**. The table identifies **histology/site** combinations which are **not reportable.** This table was created from WHO with the cooperation of the Central Brain Tumor Registry of the United States (CBTRUS).

Non-reportable Histology Term	Non-reportable Histology Code	Definitions and Sites
Carcinomas	**8010-8060, 8071-8671, 8940-8941**	Brain C710-C719 **Site/histology edit** carcinomas/brain
Carcinomas	**8010-8671, 8940-8941**	Cerebral meninges, spinal meninges, meninges NOS C700-C709 **Site/histology edit** carcinomas/meninges
Carcinomas	**8010-8671, 8940-8941**	C721-C729 (Other central nervous system) **Site/histology edit** carcinomas/other CNS
Colloid cyst	**No code**	
Epidermoid tumor/cyst	**No code**	
Glomus tympanicum, glomus jugulare	**8690/1**	These tumors occur in the inner ear, the aortic body and other paraganglia respectively; these sites are **not reportable** as non-malignant. See Head and Neck Table 9 for malignant glomus jugular tumors.
Hygroma	**9173/0**	
Hypothalamic hamartoma	**No code**	Occurs in hypothalamus
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
Neurofibromatosis, type 2 (NF2)	**No code***	
Neuroglial cyst	**No code**	
Osteochondroma	**9210/0**	Originates in the cartilage around bone, site not reportable for non-malignant neoplasms
Rathke cleft cyst	**No code**	Sella turcica C751. This is a Rathke cleft CYST, not a Rathke cleft tumor.
Schwannomatosis	**No code***	A form of neurofibromatosis newly named/discovered

***The code which is currently in ICD-O-3 for neurofibromatosis will not be used in ICD-O updates or in future ICD-O editions**

Refer to the STRs Manual

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Non-Malignant CNS and Peripheral Nerves – STRs Reportability

- **Note 1**: A neoplasm arising in a cranial nerve is coded to the specific nerve in which it arises.
- **Note 2:** Neoplasms, commonly meningiomas, arising in the dura/meninges of an intracranial nerve are coded to cerebral meninges C700.
- **Note 3**: It is important to check the operative report to determine whether the surgery is intracranial or intradural.
- **Note 4**: This table is used for non-malignant neoplasms ONLY

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CNS Histology Unlikely Combination

Error:3269 Force: Y This is an unlikely combination for site/histology/behavior. Please review.

Discrepant Data:

```
Edit: Primary Site, Morphology-Type, Beh ICDO3, 2024 (SEER) [Tag:N7040]
[Agency:FCDS]
M: Refer to PathCHART database for information on site and histology
E: 3269: Primary Site: C719, Histologic Type ICD-O-3: 9084/0 is an unlikely
combination for Date of Diagnosis: Y:2024 M:01 D:07. Please review.
                                    Date of Diagnosis(390)   [Y:2024 M:01 D:07]
                                          Primary Site(400)  [C719]
                                 Histologic Type ICD-O-3(522)  [9084]
                                   Behavior Code ICD-O-3(523)  [0]
                                     Over-ride Site/Type(2030) []
                                          Record Type(10)     []
```

Primary Site	ICD-O-3 Site Code	ICD-O-3.2 Morphology Code	ICD-O-3.2 Preferred Term	WHO Term(s)	CPC Validity Status
Cerebellum, NOS	C71.6	9084/0	Dermoid cyst, NOS		Valid

Primary Site	ICD-O-3 Site Code	ICD-O-3.2 Morphology Code	ICD-O-3.2 Preferred Term	WHO Term(s)	CPC Validity Status
Pineal gland	C75.3	9084/0	Dermoid cyst, NOS		Valid
Parathyroid gland	C75.0	9084/3	Teratoma with malignant transformation		Valid
Parathyroid gland	C75.0	9084/3		Neuroectodermal-type tumors	Valid

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Table 8: Non-Malignant CNS Tumors with the Potential of Transforming to Malignant Behavior

Original Histology and Code	Transformed Histology and Code
Chondroma **9220 (/0)**	Chondrosarcoma **9220 (/3)**
Ganglioglioma **9505 (/1)**	Anaplastic ganglioglioma **9505 (/3)**
Hemangioma **9120 (/0)**	Angiosarcoma **9120 (/3)**
Hemangiopericytoma **9150(/1)**	Anaplastic hemangiopericytoma **9150 (/3)**
Leiomyoma **8890 (/0)**	Leiomyosarcoma **8890 (/3)**
Lipoma **8850 (/0)**	Liposarcoma **8850 (/3)**
Osteoma **9180 (/0)**	Osteosarcoma **9180 (/3)**
Perineurioma **9571 (/0)**	Malignant perineurioma **9571 (/3)**
Rhabdomyoma **8900 (/0)**	Rhabdomyosarcoma **8900 (/3)**

Original Histology and Code	Transformed Histology and Code
Teratoma **9080(/1)**	Immature teratoma **9080 (/3)**
Teratoma, mature **9080 (/0)**	Immature teratoma **9080 (/3)**

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Non-Malignant CNS STRs – Histology Coding Rules

Rule	Description
H1	Code meningioma 9530/0 when diagnosis is a meningioma, benign, metaplastic, microcystic, secretory, two subtypes or lymphoplasmacyte-rich
H2	Code the reportable CNS tumor – **use Table 5**
H3	Code the histology when only one histology is present
H4	Code the subtype/variant when there is a NOS and a single subtype/variant of that NOS, example Meningioma 9530/0 and a subtype/variant of meningioma
H5	Code meningioma 9530/0 when there are multiple meningiomas
H6	Code meningiomas 9530/1 when there are multiple meningiomas of uncertain behavior

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Non-Malignant CNS STRs – Histology Coding Rules

Rule	Description
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
Rule H8	Code the histology when only one histology is present in all tumors. When the histology is not listed in Table 6 use the ICD-O and all updates
Rule H9	**Code the subtype/variant when there is a NOS and a single subtype/variant of that NOS present in all tumors, such as the following** Choroid plexus papilloma 9390/0 and a subtype/variant of choroid plexus papilloma • Craniopharyngioma 9350/1 and a subtype/variant of craniopharyngioma • Gangliocytoma 9492/0 and a subtype/variant of gangliocytoma

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Non-Malignant CNS Site-group Instructions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Table 5: Specific Histologies, NOS, and Subtypes/Variants

Specific or NOS Term, Code, and Synonym(s)	Subtype(s)/Variant(s) and Synonym(s)
Craniopharyngioma 9350 (/1)	Adamantinomatous craniopharyngioma **9351 (/1)** Papillary craniopharyngioma **9352 (/1)**
Desmoplastic infantile astrocytoma and ganglioglioma 9412 (/1) • DIAG	
Diffuse astrocytoma, MYB- or MYBL1 altered 9421 (/1) [1] • Diffuse low-grade glioma, MAPK pathway-altered • Juvenile pilocytic astrocytoma • Pilocytic astrocytoma [2]	Angiocentric glioma **9431 (/1)** • Angiocentric neuroepithelial tumor • Monomorphous angiocentric glioma
Dysembryoplastic neuroepithelial tumor 9413 (/0) [3] • DNET	
Gangliocytoma 9492 (/0)	Dysplastic cerebellar gangliocytoma **9493 (/0)** • Lhermitte-Duclos disease
Ganglioglioma 9505 (/1)	
Granular cell tumor of the sellar region 9582 (/0)	
Hemangioblastoma 9161 (/1) • Capillary hemangioblastoma	

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Non –Malignant CNS Multiple Primary Rules

Rule M2	Abstract a **single primary** when there is a single tumor.

Example: Patient has a MRI with a diagnosis of a benign /0 tumor. The pathology report from the subsequent surgery shows a borderline /1 tumor. This is a single tumor and **a single primary**. (It is the same tumor as seen on the MRI; better information is provided by the pathology report.)

Rule M3	Abstract a **single primary** (the malignant) when a single tumor meets the following two criteria: 1. The original diagnosis was non-malignant /0 or /1 AND • First course treatment was active surveillance (no tumor resection). Diagnosis was: Clinical, Radiographic, Stereotactic biopsy 2. Subsequent resection pathology is malignant /3
Rule M4	Abstract a **single primary** when a single benign tumor /0 transforms to an uncertain/borderline tumor /1. **Timing is irrelevant**. ***Example 2:*** A meningioma **9530/0 transforms** to an atypical meningioma **9539/1.**
Rule M5	Abstract a **single primary** when a neoplasm is originally diagnosed as low-grade glioma and subsequently recurs in residual tumor with a more specific histology

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Non –Malignant CNS Multiple Primary Rules

Rule M6	Abstract multiple primaries when a malignant tumor /3 occurs after a non-malignant tumor **C71.1 9380/1 Seq 60 and C71.1 9401/3 Seq 00**
Rule M7	Abstract a single primary when the patient has bilateral: • **Acoustic neuromas/ vestibular schwannomas 9560/0** • **Optic gliomas/pilocytic astrocytomas 9421/1**
Rule M8	Abstract multiple primaries when multiple tumors are present in any of the following sites: *brain C710-C719, C721, C700-C701 meninges, spinal meninges C720 spinal cord*
Rule M9	Abstract **multiple primaries** when separate, non-contiguous tumors are **two** or more different **subtypes/variants** in Column 2, **Table 5** in the Equivalent Terms and Definitions. **Timing is irrelevant**
Rule M10	Abstract a single primary when two or more separate/non-contiguous meningiomas arise in the cranial meninges – laterality is irrelevant

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Non –Malignant CNS Multiple Primary Rules

Rule M11	Abstract a single primary when there are separate/non-contiguous tumors in the brain (multicentric/multifocal) with the same histology XXXX
Rule M12	Abstract a single primary when separate/non-contiguous tumors are on the same row in **Table 5** in the Equivalent Terms and Definitions. Timing is irrelevant
Rule M13	Abstract a single primary when separate, non-contiguous tumors are Glioma NOS and a subtype/variant of Glioma NOS. **Low-grade glioma 9380 (/1)**
Rule M14	Abstract multiple primaries when separate/non-contiguous tumors are on different rows in **Table 5** in the Equivalent Terms and Definitions. Timing is irrelevant
Rule M15	Abstract a single primary when the tumors do not meet any of the above criteria

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In Summary

- Facts About Brain and CNS tumors
- Review of Anatomy
- Coding WHO Grade
- Reportability
- NPCR DQE Audit Findings
- STRs Histology and Multiple Primaries

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Resources

- SEER Appendix C – Site Specific Coding
- FCDS DAM 2026
- STORE Manual 2026
- Solid Tumor Rules (Malignant and Non-Malignant)
- SEER Program Coding and Staging Manual 2026
- ICD-0-3 Updates
- CAPs guidelines

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THANK YOU

Questions?

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References

- Genetic and Rare Diseases Information Center https://rarediseases.info.nih.gov/diseases/6902/li-fraumeni-syndrome
- National Brain Tumor Society https://braintumor.org/
- American Cancer Society
- Central Brain Tumor Registry of United States https://cbtrus.org/reports/
- National Comprehensive_Cancer Network https://www.nccn.org/guidelines/category_1

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