Brain and CNS Tumors

FCDS 2011/2012 Educational Webcast Series

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Updated for 2012 Requirements and CSv02.03.02

Presentation Outline

- Overview
- Anatomy of the Human Brain
- Multiple Primary and Histology Coding Rules
- Collaborative Stage Data Collection System (CSv2)
- C.S. Site Specific Factors
- Treatment Options

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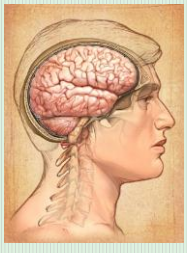
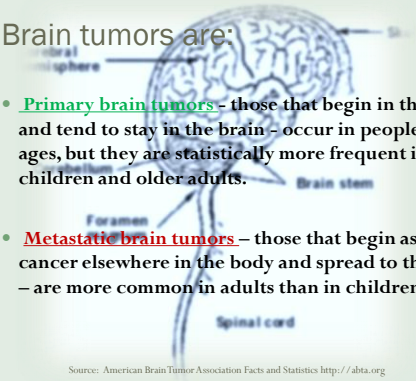
Overview



Brain tumors are:

- **Primary brain tumors – those that begin in the brain and tend to stay in the brain – occur in people of all ages, but they are statistically more frequent in children and older adults.**
- **Metastatic brain tumors – those that begin as a cancer elsewhere in the body and spread to the brain – are more common in adults than in children.**



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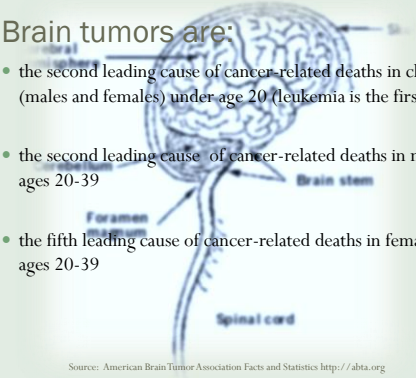
Source: American Brain Tumor Association Facts and Statistics http://abta.org

Brain tumors are:

- the second leading cause of cancer-related deaths in children (males and females) under age 20 (leukemia is the first)
- the second leading cause of cancer-related deaths in males ages 20-39
- the fifth leading cause of cancer-related deaths in females ages 20-39



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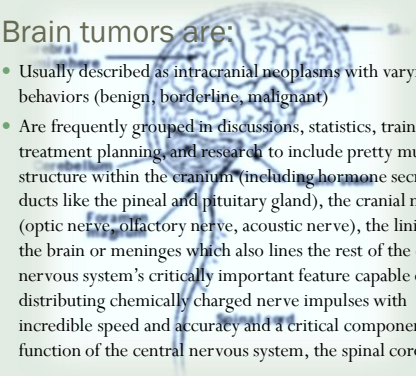
Source: American Brain Tumor Association Facts and Statistics http://abta.org

Brain tumors are:

- Usually described as intracranial neoplasms with varying behaviors (benign, borderline, malignant)
- Are frequently grouped in discussions, statistics, training, treatment planning, and research to include pretty much any structure within the cranium (including hormone secreting ducts like the pineal and pituitary gland), the cranial nerves (optic nerve, olfactory nerve, acoustic nerve), the lining of the brain or meninges which also lines the rest of the central nervous system's critically important feature capable of distributing chemically charged nerve impulses with incredible speed and accuracy and a critical component of the function of the central nervous system, the spinal cord.



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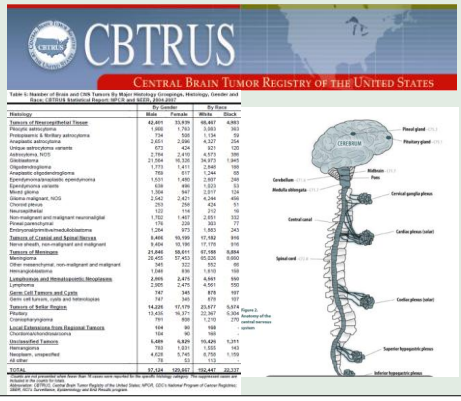
Intracranial vs. Extra-cranial

Brain tumors are classified as either intra- or extra-cranial and both produce clinical/symptomatic effects that are similar in terms of mass effect, hemorrhage, seizure activity, and edema

Intracranial Sites	Extracranial Sites
• Brain	• Spinal cord
• Cerebral meninges	• Cauda equina
• Cranial nerves and other intracranial parts of the CNS	• Spinal meninges
• Craniopharyngeal duct	
• Pineal gland	
• Pituitary gland	

ALL Brain Tumors are Reportable

- Public Law 107-260, the Benign Brain Tumor Cancer Registries Amendment Act, [PDF-185KB] requires programs participating in the National Program for Cancer Registries (NPCR) to collect data on benign and borderline tumors of the central nervous system in addition to the previously required data on malignant tumors.
- In addition to NPCR, the National Cancer Institute's (NCI) Surveillance, Epidemiology and End Results (SEER) program and the American College of Surgeons' (ACoS) Commission on Cancer began requiring that these tumors be reported, starting with cases diagnosed on January 1, 2004.



Brain and CNS Tumors – All Ages

- 2011 estimates in the United States
  - 64,540 new cancer cases

This includes:

Malignant brain tumors	(24,070)
Non-malignant brain tumors	(40,470)

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Source: American Brain Tumor Association Facts and Statistics http://abta.org

Brain and CNS Tumors - Children

- Approximately 4,150 children younger than age 20 will be diagnosed with primary brain tumors in 2011
  - 2,960 will be less than 15 years of age
  - 1,190 will be between the ages of 15 and 19
- Giomas represent a high percentage of childhood tumors
  - 55% of all tumors and 71% of malignant tumors in children age 0-14
  - 39% if all tumors and 74% of malignant tumors in children age 15-19

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Source: American Brain Tumor Association Facts and Statistics http://abta.org

Brain and CNS Tumors

Table 21: Primary Brain and Other Nervous System Tumors, Estimated Number of Cases [a,b] Overall and by Behavior by State, 2011; Primary Malignant Brain and Other Nervous System Tumors, Estimated Number of Deaths [a,b] by State, 2010

	2011 Estimated New Cases			2010 Estimated Deaths
STATE	All	Malignant	Non-Malignant	Malignant
Alabama	990	360	640	210
Alaska	130	50	80	-
Arizona	1,460	570	890	280
Arkansas	620	230	390	150
California	7,260	2,700	4,560	1,490
Colorado	1,040	400	640	210
Connecticut	770	290	480	150
Delaware	200	70	120	-
District of Columbia	110	30	80	-
Florida	4,560	1,700	2,870	800
Georgia	1,920	690	1,230	340
Hawaii	260	80	180	-
Idaho	320	130	190	80
Illinois	2,610	970	1,640	470
Indiana	1,330	510	820	340
Iowa	660	250	400	170
Kansas	590	220	360	140
Kentucky	910	350	560	180
Louisiana	890	320	580	210
Maine	320	120	190	80
Maryland	1,200	420	780	210
Massachusetts	1,430	540	890	280
Michigan	2,160	810	1,350	500

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Source: American Brain Tumor Association Facts and Statistics http://abta.org

Figure 13. Most Common Brain and CNS Tumors by Age

CBTRUS Statistical Report: NPCR and SEER Data from 2004-2007

Age (yr)	Most Common Histology	Second Most Common Histology
0-4	Embryonal/medulloblastoma	Pilocytic astrocytoma
5-9	Pilocytic astrocytoma	Malignant glioma, NOS
10-14	Pilocytic astrocytoma	Neuronal/glial
15-19	Pituitary	Pilocytic astrocytoma
20-34	Pituitary	Meningioma
35-44	Meningioma	Pituitary
45-54	Meningioma	Glioblastoma
55-64	Meningioma	Glioblastoma
65-74	Meningioma	Glioblastoma
75-84	Meningioma	Glioblastoma
85+	Meningioma	Neoplasm, unspecified

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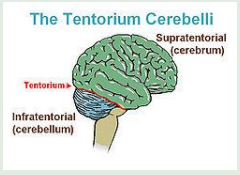
Childhood Brain Tumors

Tentorium - extension of the dura mater separating the cerebellum from the occipital lobes

- 50% of childhood brain and CNS tumors are infratentorial, originating below the tentorium
- 20+% of childhood CNS tumors are located in the sellar or suprasellar region around the sella turcica (the bone that contains the pituitary gland)
- Remainder of tumors occur in spinal cord, brain stem, cranial nerves, etc.



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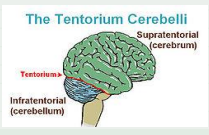
Childhood Brain Tumors

Supratentorial - childhood	Infratentorial - childhood
• Craniopharyngiomas.	• Cerebellar astrocytomas (usually high-grade).
• Diencephalic and hypothalamic gliomas.	• Medulloblastomas (primitive neuroectodermal tumors).
• Germ cell tumors.	• Ependymomas (low-grade or anaplastic).
• Low-grade astrocytomas.	• Brain stem gliomas (high-grade or low-grade).
• Anaplastic astrocytomas.	• Atypical teratoid tumors
• Glioblastoma multiforme.	
• Mixed gliomas.	
• Oligodendrogliomas.	
• Primitive neuroectodermal tumors.	
• Low-grade or anaplastic ependymomas.	
• Meningiomas.	
• Choroid plexus tumors.	



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Pilocytic Astrocytoma

- Synonyms include:
  - Juvenile pilocytic astrocytoma
  - Cystic cerebellar astrocytoma
  - Juvenile pilomyxoid astrocytoma
- Characteristics:
  - Usually slow growing, well-circumscribed neoplasm
  - Associated with the formation of a single (or multiple) cyst(s)
  - Arise in cerebellum near brainstem
  - Other common sites include hypothalamic region and optic chiasm
  - May occur in cerebral hemispheres and spinal cord
  - Associated with neurofibromatosis Type 1 (NF1)
  - 10 year survival greater than 90% with total removal
  - Not associated with recurrence with total removal
  - WHO Grade I - benign

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Pilocytic Astrocytoma

- HOWEVER, when the *ICD-O-3* was published, the behavior code for pilocytic astrocytoma downgraded from /3 (malignant behavior) to 1 (borderline behavior) as it still appears in the *ICD-O-3* reference sitting on your desktop.
- Registrars in the United States were in 2000 and continue to be instructed by our national standard setting agencies to assign the behavior code /3 to these tumors despite the WHO downgrade.
- Rationale: To ensure complete reporting and data consistency, registrars should continue to assign the malignant behavior code (3) to pilocytic astrocytoma. This is the standard for all U.S. registries in all programs.
- Confusing to researchers and public health studies since we reference ICD-O as our primary coding reference and ICD-O-3 has never published the U.S. change and does not assign a malignant behavior to this type of astrocytoma.

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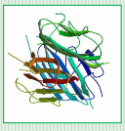
Causes and Risk Factors

No single risk factor accounting for the majority of brain tumors has been identified even though many environmental and genetic factors are being studied



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Causes and Risk Factors

ENVIRONMENTAL

- Many studies have examined a wide spectrum of environmental factors as a cause for brain tumors. Of the long list of factors studied, only **exposure to ionizing radiation** has consistently been shown to put one at increased risk for developing a brain tumor.

GENETIC

- There are a few **rare genetic syndromes** that involve brain tumors.
  - NF1 (NF1 gene)
  - NF2 (NF2 gene)
  - Turcots (APC gene)
  - Gorlins (PTCH gene)
  - Tuberous sclerosis (TSC1 and TSC2 genes)
  - Li-Fraumeni syndrome (TP53 gene)

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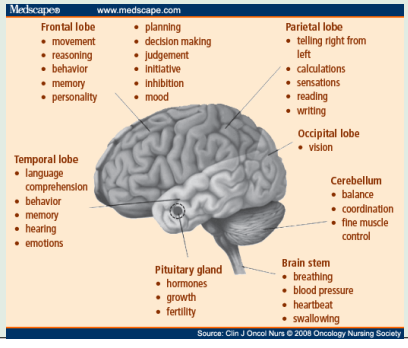
Range of tumors and symptoms

- There are over 120 different types of brain/CNS tumors.
- 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 very much on the size and location of the tumor. General symptoms include persistent headaches which tend to be worse with activity, at night or early in the morning, convulsions, vomiting, subtle changes in personality, memory, mental ability, drowsiness, lethargy.

20 SEER Training Modules

Range of tumors and symptoms



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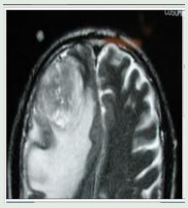
Range of tumors and symptoms

- Symptoms are often 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. Symptoms of a tumor located here may include difficulty saying correct words while still capable of understanding what is being said.
  - If the tumor is located in the frontal lobe which controls intellectual function, thought process, behavior and memory, those activities may be affected.
- Similarity to closed head injury victims (motorcycle crash).

Midline Shift and Mass Effect

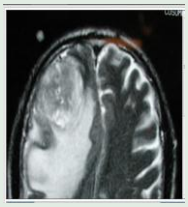
- The bony cranium protects the brain from outside impacts to the head. When swelling occurs in the brain, there isn't much "give".
- The swelling results in intracranial pressure and can cause a number of effects that begin to impact quality of life and comfort for the patient.
- The easiest way to describe midline shift is to bring to mind siting in a movie theater. As soon as the person to one side of you puts his elbow onto the shared armrest between you, you tend to shift away.



Source: Medscape

Midline Shift and Mass Effect

- Midline is a central boundary separating the left and right hemispheres.
- Midline Shift – Tumor crosses the brain to shift across the center line
- Mass Effect is – Edema or swelling causes the brain to shift across center line
- Both create new symptoms at cross-over
  - Depends on the size and location of he tumor and level of spread
  - Edema caused by many things
  - Either cause pushes midline out of alignment



Source: Medscape

The Brain is Incapable of Feeling Pain

- Surgeons are able to cut living brains without fear of hurting their patients
- However, symptoms from tumors and their effect within the cranial cavity on various functions of the brain is a different story, altogether.
- Much is dependent upon tumor location and infiltration

Source: National Geographic, couretsy of Fred Hossler/Getty Images

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Benign/Borderline/Malignant ???

BENIGN TUMORS	MALIGNANT TUMORS
• slow growing	• usually rapidly growing
• distinct borders	• invasive
• rarely spread	• life-threatening

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Source: American Brain Tumor Association Facts and Statistics http://abta.org

Survival Trends

- SEER data from 1995-2007 5-year relative survival
  - Males 34%
  - Females 38%
- Children age 0-19 have the highest 5-year relative survival rate 72%
- The survival rate diminishes as age increases, down to 5% for persons age 75 and older

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Table 23: One-, Two-, Three-, Four-, Five-, and 10-Year Relative Survival Rates[a,b] For Selected Malignant Brain and Central Nervous System Tumors, SEER 17 Registries, 1995-2007

Histology	# Cases	1-Yr	2-Yr	3-Yr	4-Yr	5-Yr	10-Yr
Pilocytic astrocytoma	2,294	97.79%	96.80%	95.90%	95.04%	94.40%	92.10%
Protoplasmic & fibrillary astrocytoma	626	74.30%	60.97%	55.29%	51.09%	48.10%	36.35%
Anaplastic astrocytoma	2,846	60.32%	42.74%	34.70%	30.50%	27.36%	21.87%
Astrocytoma, NOS	3,280	70.01%	60.14%	54.59%	50.95%	48.16%	39.10%
Glioblastoma	19,797	34.60%	12.63%	7.31%	5.40%	4.75%	2.80%
Oligodendroglioma	2,453	94.17%	89.92%	86.25%	82.77%	79.48%	63.58%
Anaplastic oligodendroglioma	970	79.91%	66.07%	59.55%	53.90%	49.40%	34.95%
Ependymoma/anaplastic ependymoma	1,748	94.00%	89.23%	85.81%	83.41%	82.41%	76.22%
Mixed glioma	1,296	87.52%	74.86%	67.56%	61.74%	57.32%	46.37%
Glioma malignant, NOS	2,790	60.40%	49.07%	46.30%	44.25%	43.27%	39.54%
Neuroepithelial	118	54.10%	42.58%	41.98%	36.34%	33.02%	28.74%
Malignant neuronal/glial, Neuronal and mixed	526	88.48%	79.55%	76.46%	71.69%	70.66%	58.93%
Embryonal/primitive/ medulloblastoma	1,865	82.42%	71.72%	66.77%	63.76%	61.71%	55.05%
Lymphoma	3,421	47.48%	38.69%	34.23%	31.01%	28.52%	21.61%
Total: All Brain and CNS[c]	47,692	57.21%	43.34%	38.99%	36.74%	35.47%	31.73%

[a]The cohort analysis of survival rates was utilized for calculating the survival estimates presented in this table. Long-term cohort-based survival estimates reflect the survival experience of individuals diagnosed over the time period, and they may not necessarily reflect the long-term survival outlook of newly diagnosed cases.
[b]Rates are an estimate of the percentage of patients alive at one, two, three, four, five, and ten year, respectively.
[c]Includes histologies not listed in this table.

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Tumor-Specific Statistics

Meningioma	34% of all primary brain tumors
Glioma	31% of all primary brain tumors
(80% of all malignant brain tumors)	
Glioblastoma	17% of all primary brain tumors
(54% of all gliomas)	
Astrocytoma	7% of all primary brain tumors
Oligodendroglioma	2% of all primary brain tumors
Ependymoma	1% of all primary brain tumors
Pituitary tumors	13% of all primary brain tumors
Nerve sheath tumors	9% of all primary brain tumors (ie: acoustic neuromas, schwannoma, malignant peripheral nerve sheath tumor)
Medulloblastoma/embryonal/and other tumors of primitive (developmental) nerve origin	3% of all primary brain tumors
Lymphoma	2-3% of all primary brain tumors

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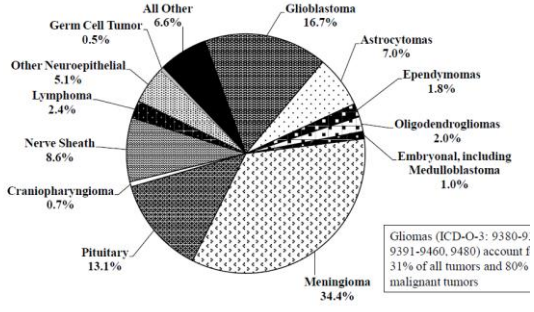
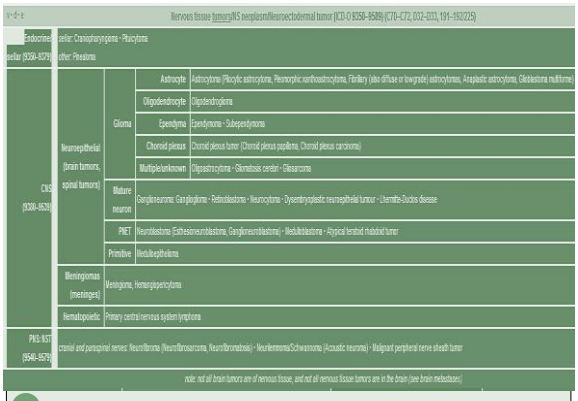
Figure 5. Distribution of All Primary Brain and CNS Tumors by Histology (N=226,791)

CBTRUS Statistical Report: NPCR and SEER Data from 2004-2007





Nervous tissue tumors/NS neoplasm/Neuroectodermal tumor (ICD-O 9350–9589) (C70–C72, D32–D33, 191–192.2))				
Endocrine (sellar (9350–9379))	sellar: Craniopharyngioma - Pituicytoma other: Pinealoma			
CNS (9380–9626)	Neuroepithelial (brain tumors, spinal tumors)	Glioma	Astrocyte	Astrocytoma (Pilocytic astrocytoma, Pleomorphic xanthoastrocytoma, Fibrillary (also diffuse or lowgrade) astrocytomas, Anaplastic astrocytoma, Glioblastoma multiforme)
			Oligodendrocyte	Oligodendroglioma
			Ependyma	Ependymoma - Subependymoma
			Choroid plexus	Choroid plexus tumor (Choroid plexus papilloma, Choroid plexus carcinoma)
			Multiple/unknown	Oligoastrocytoma - Gliomatosis cerebri - Gliosarcoma
		Mature neuron	Ganglioneuroma: Gangliocytoma - Retinoblastoma - Neurocytoma - Dysembryoplastic neuroepithelial tumor - Lhermitte-Duclos disease	
		PNET	Neuroblastoma (Esthesioneuroblastoma, Ganglioneuroblastoma) - Medulloblastoma - Atypical teratoid rhabdoid tumor	
		Primitive	Medulloepithelioma	
	Meningiomas (meninges)	Meningioma, Hemangiopericytoma		
	Hematopoietic	Primary central nervous system lymphoma		
PNS (9540–9579)	cranial and paraspinal nerves: Neurofibroma (Neurofibrosarcoma, Neurofibromatosis) - Neurilemmoma/Schwannoma (Acoustic neuroma) - Malignant peripheral nerve sheath tumor			
note: not all brain tumors are of nervous tissue, and not all nervous tissue tumors are in the brain (see brain metastases)				

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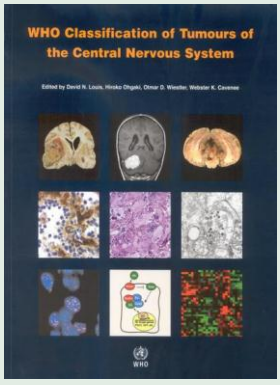
Source: wikipedia.org and ccrcal.org



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WHO Classification Groups

- Tumors of Neuroepithelial Tissue
- Tumors of Cranial and Paraspinal Nerves
- Tumors of Meninges
- Lymphomas and Hematopoietic Malignancies
- Germ Cell Tumors
- Tumors of the Sellar Region
- Metastatic Tumors

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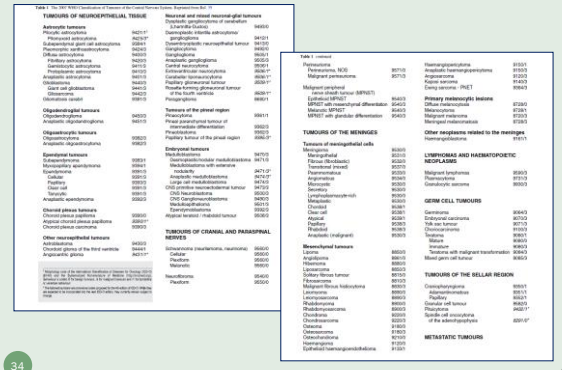

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WHO Grade

- Four categories of tumor
  - Grade I slow growing, non-malignant, associated with long-term survival – benign tumors
  - Grade II relatively slow-growing, sometimes recur as higher grade tumors, can be malignant or non-malignant (borderline malignant)
  - Grade III malignant and often recur as higher grade tumors
  - Grade IV reproduce rapidly and are very aggressive malignant tumors
- WHO grade is not recorded as part of the histology
- WHO grade is used by the clinician to plan treatment and predict prognosis

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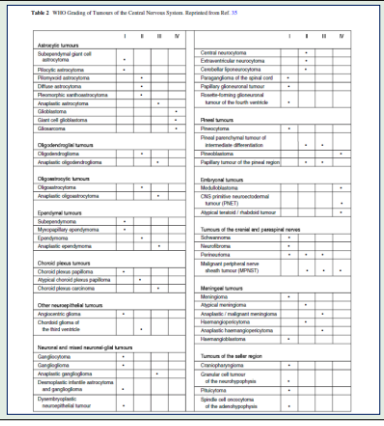

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ANATOMY OF THE HUMAN BRAIN

Source: National Geographic, couretsy of Fred Hossler/Getty Images

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THE HUMAN BRAIN

- The brain is the largest intracranial organ
- The brain is a 3-pound mass of jelly-like fats and tissues
- It is the most complex of all known living structures
- The skull or cranium is bone that covers the brain
- Up to one trillion nerve cells working together coordinate the physical actions and mental processes (voluntary and involuntary) that set humans apart from all other species

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Source: CDC Data Collection of Primary CNS Tumors, NPCR Training Materials 2004

- The CNS includes both intracranial sites (inside the cranium) and extra-cranial sites (outside the cranium).
  - The pituitary gland, craniopharyngeal duct and pineal gland are found inside, alongside brain tissue
  - Cranial nerves directly link to brain tissue
  - The spinal cord is part of the CNS though not intracranial
- Any tumor that originates in the brain, spinal cord, the cranial nerves, one of the glands/ducts within the cranium (pineal, pituitary, craniopharyngeal), or the lining of the cranium (meninges) is reportable regardless of behavior (benign, borderline, or malignant).

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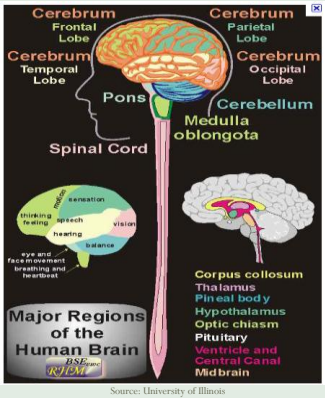
Source: CDC Data Collection of Primary CNS Tumors, NPCR Training Materials 2004



Source: University of Illinois

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ICD-O Topography Codes (Anatomic Site)

- Brain (C71.0 - C71.9)
- Cerebrum (C71.0)
- Frontal lobe (C71.1)
- Temporal lobe (C71.2)
- Parietal lobe (C71.3)
- Occipital lobe (C71.4)
- Ventricle (C71.5)
- Cerebellum (C71.6)
- Brain stem (C71.7)
- Overlapping lesion of the brain (C71.8)
- Brain NOS (C71.9)
- Meninges (C70.0 - C70.9)
  - Cerebral meninges (C70.0)
  - Spinal meninges (C70.1)
  - Meninges NOS (C70.9)
- Spinal cord (C72.0)
- Cauda equina (C72.1)
- Cranial nerves (C72.2 - C72.5)
  - Olfactory nerve (C72.2)
  - Optic nerve (C72.3)
  - Acoustic nerve (C72.4)
  - Cranial nerve NOS (C72.5)
- Other CNS (C72.8, C72.9)
- Pituitary gland (C75.1)
- Craniopharyngeal duct (C75.2)
- Pineal gland (C75.3)

41

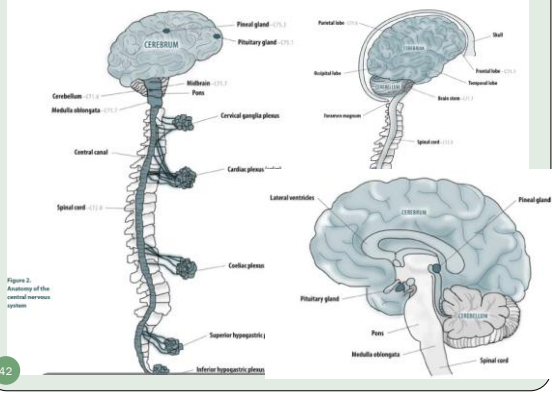
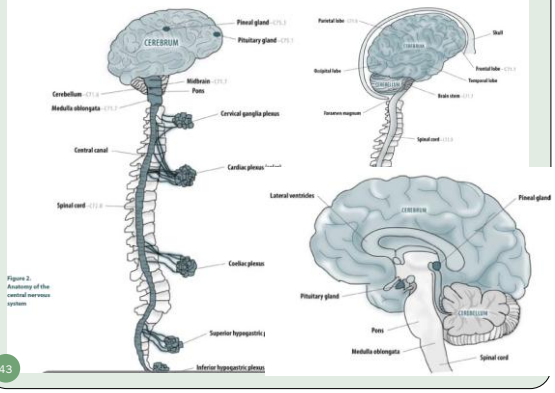


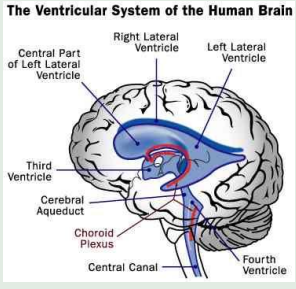
Figure 2. Anatomy of the central nervous system

42



43

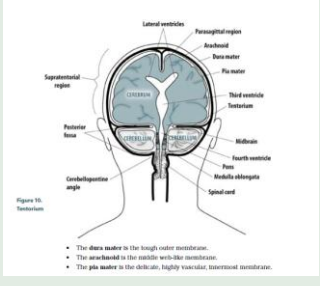
Ventricular System of the Brain

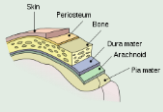


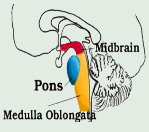
Source: solarnavigator.net/human_brain

44

Meninges and Brain Stem

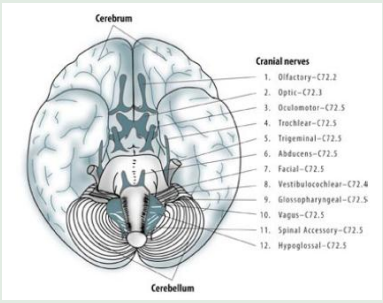






45

Cranial Nerves



46

Cranial Nerve Functions

Cranial Nerve:	Major Functions:
I Olfactory	smell
II Optic	vision
III Oculomotor	eyelid and eyeball movement
IV Trochlear	turns eye downward and laterally, controls superior oblique muscles
V Trigeminal	chewing, face & mouth touch & pain
VI Abducens	turns eye laterally
VII Facial	facial expressions, taste, tears, saliva
VIII Vestibulocochlear	Also referred to as Auditory Nerve: hearing, equilibrium sensation
IX Glossopharyngeal	Taste, senses carotid blood pressure
X Vagus	aortic blood pressure, heart rate, stimulates digestive organs, taste
XI Spinal Accessory	controls trapezius & sternocleidomastoid muscles, controls swallowing
XII Hypoglossal	controls tongue movements

47

Characteristics of Brain Tumors

- Start in the brain and grow steadily there.
- Very rarely spread to other organs through the bloodstream.
- Are named for the cells from which they arise, each having a certain function essential to normal physiological functioning of the brain. For example:
  - Gliomas arise from glial cells which support the CNS.
    - Astrocytomas arise from astrocytes
    - Ependymomas arise from ependymal cells which line the ventricles (fluid filled spaces within the brain) or central canal of the spinal cord.
    - Oligodendrogliomas arise from oligodentdrocyte cells which make up the fatty substance called myolin that covers nerves like electrical insulation.
    - Brain Stem Gliomas arise in the lowest part of the brain.

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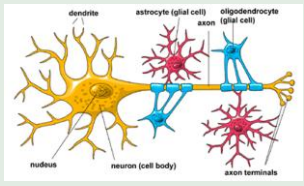
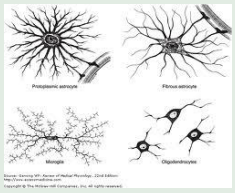
Characteristics of Brain Tumors





Source: medicalgeek.com/indian-post-graduate-exams

49

Histologic Type - Glioma

- Most common category of primary brain tumors. They begin in glial cells (supporting cells of the CNS)
- Often spread into surrounding brain tissue along nerve fibers invading the spaces between nearby normal brain cells. Some invade the surrounding brain more than others.
- Difficulty obtaining complete surgical removal. MRI scans show the largest part of the glioma, but cannot reliably show areas of the brain where tumor cells have invaded. Aggressive efforts to remove small numbers of tumor cells within the brain could cause loss of neurologic function.
- When it is not possible to remove the entire glioma, post-op radiation therapy and chemotherapy may be advised.
- Even with maximum safe resection followed by radiation and chemotherapy, gliomas can grow back.

50

Glioma – 3 Main Histologic Types

1. Astrocytoma: In adults most often arise in the cerebrum. In children they occur in the brain stem, cerebrum and cerebellum. Rarely in brain stem in adults. Felt to be most aggressive of brain tumors.
   - Grade I and II astrocytomas are low-grade astrocytomas.
   - Grade III astrocytoma is an "anaplastic astrocytoma".
   - Grade IV astrocytoma is a "glioblastoma multiforme".

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Glioma – 3 Main Histologic Types

2. Oligodroglioma: Rare tumor that usually occurs in the cerebrum, grows slowly and usually does not spread into surrounding brain tissue like astrocytoma does. Most common in middle-aged adults.

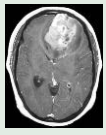
3. Ependymoma: Most commonly arise in children and young adults. They are also seen with neurofibromatosis Type II. (which we will discuss in a bit)

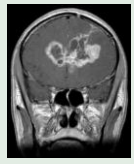
52

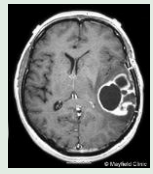
Glioma – Other Subtypes

There are other subtypes of gliomas, each with their own specific characteristics and modes of growth.

- Brain Stem Glioma
- Juvenile Pilocytic Astrocytoma
- Pleomorphic Xanthoastrocytoma
- Subependymoma
- Ganglioglioma







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Glioma Tumor Markers

Table 5 Current Molecular Biomarkers in Glioma

Biomarker	Molecular Compartment	Purpose	Analytic Validity Demonstrated	Level of Evidence	NCCN Category of Evidence	References
Markers With Accepted Clinical Utility						
1p/19q codeletion (unbalanced translocation)	Tumor DNA	Diagnostic (oligodendroglioma)	FISH, aCGH, LOH, MPLA	IA	1	Smith et al.[46]
IDH mutation (*IDH1* c. 395 G>A p.R132H (*IDH2*)	Tumor DNA, tumor protein	Positive is favorably prognostic; also a diagnostic marker	IHC, DNA sequencing	IIB		Houillier et al.[49] Dubbink et al.[51]
MGMT methylation	Tumor DNA	Prognostic, predictive (benefit for chemotherapy), pharmacodynamic (pseudorecurrence)	MS-PCR, MS-pyrosequencing, MS-MPLA	IIB		Hegi et al.[61] Gilbert et al.[215]
Markers With Emerging Evidence						
BRAF fusion (pilocytic astrocytoma)	Tumor DNA	Diagnostic (pilocytic astrocytoma)	LDI-PCR, 5′ RACE, FISH	IIB		Jeuken and Wesseling,[216] Jones et al.[59]
CIMP (CpG island methylator phenotype)	Tumor DNA	Positive is favorably prognostic	Gene expression microarray, pyrosequencing	IIB		Noushmehr et al.[65] Gilbert et al.[215]

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Non-Glial Tumors

- Medulloblastoma: Usually arises in the cerebrum, is the most common brain tumor in children, and is sometimes called a "primitive neuroectodermal tumor" or PNET.
- Meningioma: Arises from the meninges which are the outside coverings of the brain between the skull and the brain itself. It usually presses on the brain, but does not invade it and often grows slowly.

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Non-Glial Tumors

- Schwannoma: Arises from Schwann cells present in certain nerves, including those that control balance and hearing.
- A common site is the vestibular nerve which carries signals from the inner ear to the brain stem.
- Tumors in this location are called "acoustic neuromas" (a.k.a. vestibular schwannoma), and occur most often in adults.

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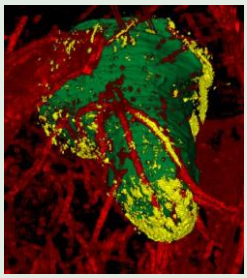
Non-Glial Tumors

- Craniopharyngioma: Grows at the base of the brain, arises from the tissue connecting the brain and the pituitary gland and occurs in both adults and children.
- Pituitary Adenoma: Arises from the pituitary gland and may cause compression of the optic nerves causing vision problems. Some produce excessive amounts of hormones that can disrupt the body's metabolism.

57

Observing Migration of Glioma Cells



Source: Case Western Reserve University School of Medicine, public release 8/25/11

58

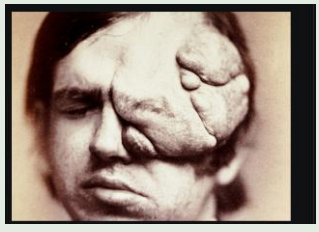
Neurofibromatosis

- The neurofibromatoses (NF) are a group of genetic disorders which cause tumors to grow along nerves and can also affect the development of non-nervous tissues such as bones and skin.
- Neurofibromatosis Type I (NF-I), also known as Peripheral NF and historically as von Recklinghausen Disease
  - Occurs in 1:4,000 births
  - Multiple cafe-au-lait spots (not reportable)
  - Many, many neurofibromas on or under the skin (not reportable)
  - Enlargement and deformation of bones and curvature of the spine
  - Tumors may develop in brain, on cranial nerves, or the spinal cord

59

Neurofibromatosis Foundation

NF Type I: First documented photo 1871



60

Source Credit: Dr. Stanley B. Burns
http://www.cbsnews.com/2300-204_162-10007019-6.html#ixzz1clEzAchI

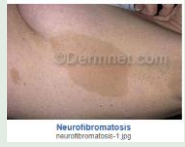
Other Manifestions of NF Type I

Lisch nodules on the eye

- Melanocytic hemartomas

Café-au-lait spots on skin

- Discolored birth marks



61

Medscape Source: Dermnet.com; Dermatologic Manifestations of NFType I

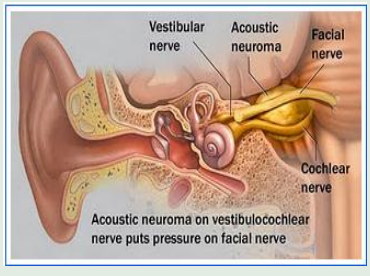
Neurofibromatosis Type II

- Neurofibromatosis Type II (NFII), also known as Multiple Inherited Schwannomas, Meningiomas and Ependymomas (MISME) or Bilateral Acoustic Neurofibromatosis (BAN).
- Is a genetically inherited diseasecaused by mutations of the "Merlin" gene, which appears to influences the form and movement of cells
- Primary manifestation is a development of non-malignant brain tumors in the region of the cranial nerves, frequently bilaterally. The eighth cranial nerve is the auditory-vestibular nerve which transmits sensory information from the inner ear to the brain and is commonly affected.

62

Source: California Ear Institute

Acoustic Neuroma/Schwannoma



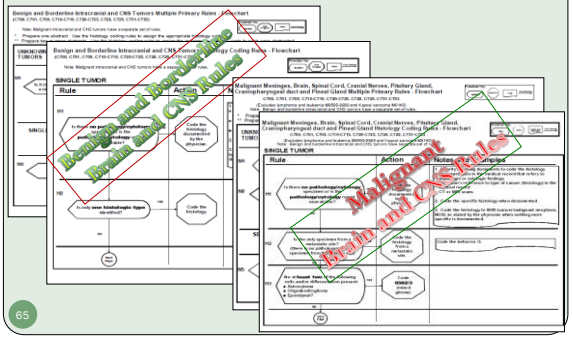
63

Source: http://thrivingwithneurofibromatosis.blogspot.com

Multiple Primary Rules
Histology Coding Rules

64

Different Rules for Benign and Malignant



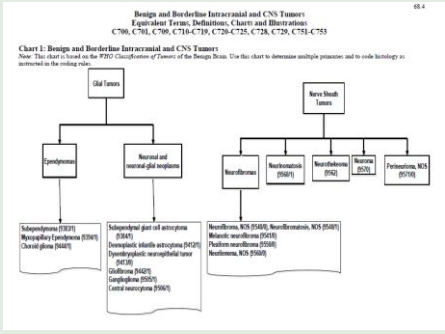
65

Sequence Numbering for Brain Tumors

- **Malignant primary brain and CNS tumors** are assigned **Sequence Codes in the range 00-35**
  - Sequence Chronologically 00-35
  - Only count malignant tumors in the sequence
  - If only one malignant tumor occurs, it is coded 00
  - If subsequent (multiple) primary malignant and/or in situ neoplasms, the sequence number for the first tumor begins at 01, the sequence number for the second primary tumor is 02, and so forth.
- **Non-malignant primary brain and CNS tumors** are assigned **Sequence Codes in the range 60-87**.
  - Sequence Chronologically 60-87
  - Only count benign/borderline or reportable by agreement neoplasms in the sequence
  - If only one non-malignant tumor occurs, it is coded 60.
  - If subsequent (multiple) non-malignant neoplasms are diagnosed, the first tumor should be sequenced as 61, the second 62 and so forth.

66

Benign and Borderline Tumor Rules



67

Benign and Borderline Tumor Rules

- When multiple tumors are present registrars should identify and document specific characteristics for MPH Rules Text
  - Date of Diagnosis (Timing is not used to determine number of abstracts or primary neoplasms to abstract)
  - Method and Details of Diagnosis (some are never resected)
  - Location of Tumor
  - Laterality
  - Histologic Type – refer to Chart 1
  - Tumor Behavior
  - Multiple Meningioma's (meningiomatosis)
  - Neurofibromatosis Characteristics (when applicable)

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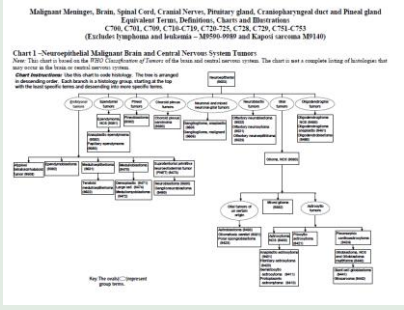
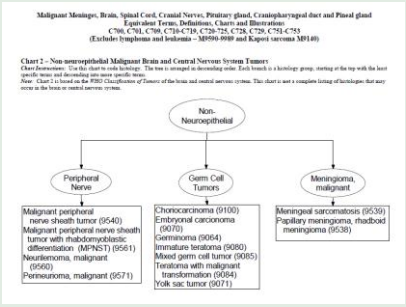
Malignant Tumor Rules



69

Malignant Tumor Rules



70

Malignant Tumor Rules

- When multiple tumors are present registrars should identify and document specific characteristics for MPH Rules Text
  - Date of Diagnosis (Timing is not used to determine number of abstracts or primary neoplasms to abstract)
  - Method and Details of Diagnosis (most attempt resection)
  - Location of Tumor (not spread or invasion – but bulk of tumor)
  - Histologic Type – refer to Chart 1 and/or Chart 2
  - Tumor Behavior
  - Variations or Combinations of One or More Glial Tumors Over Lifetime – astrocytoma, glioblastoma, ependymoma, or oligodendroglioma
    - Special rules for determining # abstracts
    - Special rules for determining whether or not is mixed glioma
  - Note: Recurrence, progression, or any reappearance of histologies on the same branch in Chart 1 or Chart 2 is always the same disease process.

71

Report /Sequence All Tumors Over Lifetime

REMINDER: Sequence numbers for malignant neoplasms and for benign, borderline, and other reportable-by-agreement cases are assigned over a lifetime.

- Therefore,

IF A PATIENT WAS DIAGNOSED WITH A NON-MALIGNANT CNS NEOPLASM ***BEFORE REPORTING WAS REQUIRED (January 1, 2004)***,

THE NEW (SECOND) NEOPLASM SHOULD BE ASSIGNED SEQUENCE NUMBER 62 AND THE FIRST NEOPLASM (Seq 61) IS REPORTABLE AS A HISTORICAL CASE TO FCDS.

(An abstract/accession is not be required by CoC or SEER but is by FCDS)

- Any benign and/or borderline brain or CNS tumor(s) diagnosed before January 1, 2004 ARE REPORTABLE TO FCDS as historical cases when accompanied by another reportable primary.

72

FCDS Data Acquisition Manual and CDC Data Collection of Primary Central Nervous System Tumors

73

COLLABORATIVE STAGE DATA COLLECTION SYSTEM

Brain

Brain and Cerebral Meninges

C70.0, C71.0-C71.9

- C70.0 Cerebral meninges
- C71.0 Cerebrum
- C71.1 Frontal lobe
- C71.2 Temporal lobe
- C71.3 Parietal lobe
- C71.4 Occipital lobe
- C71.5 Ventricle, NOS
- C71.6 Cerebellum, NOS
- C71.7 Brain stem
- C71.8 Overlapping lesio
- C71.9 Brain, NOS
- Note 1: This schema is c 7th edition. The AJCC ca
- Note 2: AJCC does not

CNSOther

Other Parts of Central Nervous System

C70.1, C70.9, C72.0-C72.5, C72.8-C72.9

- C70.1 Spinal meninges
- C70.9 Meninges, NOS
- C72.0 Spinal cord
- C72.1 Cauda equina
- C72.2 Olfactory nerve
- C72.3 Optic nerve
- C72.4 Acoustic nerve
- C72.5 Cranial nerve, N
- C72.8 Overlapping lesi
- C72.9 Nervous system,
- Note 1: This schema is seventh editions.

IntracranialGland

Pituitary Gland, Craniopharyngeal Duct, and Pineal Gland

C75.1, C75.2, C75.3

- C75.1 Pituitary gland
- C75.2 Craniopharyngeal duct
- C75.3 Pineal Gland
- Note: AJCC does not define TNM staging for this schema.

74

Brain

Brain and Cerebral Meninges

C70.0, C71.0-C71.9

CS Tumor Size
CS Extension
CS Tumor Size/Ext Eval = **9**
CS Lymph Nodes
CS Lymph Nodes Eval = **9**
Regional Nodes Positive = **99**
Regional Nodes Examined = **99**
CS Mets at DX
CS Mets Eval = **9**
CS Site-Specific Factor 1
World Health Organization (WHO) Grade Classification
CS Site-Specific Factor 2
KI-67/MIB-1 Labeling Index (LI) Brain
CS Site-Specific Factor 3
Functional Neurologic Status - Karnofsky Performance Scale (KPS)
CS Site-Specific Factor 4
Methylation of O6-Methylguanine-Methyltransferase (MGMT)
CS Site-Specific Factor 5
Chromosome 1p: Loss of Heterozygosity (LOH)
CS Site-Specific Factor 6
Chromosome 19q: Loss of Heterozygosity (LOH)

CS Site-Specific Factor 7
Surgical Resection
CS Site-Specific Factor 8
Unifocal vs Multifocal Tumor
CS Site-Specific Factor 9 = **988**
CS Site-Specific Factor 10 = **988**
CS Site-Specific Factor 11 = **988**
CS Site-Specific Factor 12 = **988**
CS Site-Specific Factor 13 = **988**
CS Site-Specific Factor 14 = **988**
CS Site-Specific Factor 15 = **988**
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CS Site-Specific Factor 18 = **988**
CS Site-Specific Factor 19 = **988**
CS Site-Specific Factor 20 = **988**
CS Site-Specific Factor 21 = **988**
CS Site-Specific Factor 22 = **988**
CS Site-Specific Factor 23 = **988**
CS Site-Specific Factor 24 = **988**
CS Site-Specific Factor 25 = **988**
Histology Inclusion Table AJCC 7th ed.
Histology Exclusion Table AJCC 6th ed. = **NA**
AJCC TNM 7 Stage = **NA**
AJCC TNM 6 Stage = **NA**
Summary Stage

Revision Info List of Schemas

75

CS Extension

- Note 1: The tentorium cerebelli is an extension of the dura mater that separates the cerebellum from the inferior portion of the occipital lobes. The location of the tumor above or below the tentorium can help in determining the type of tumor, also most adult brain tumors are supratentorial, and most pediatric brain tumors are infratentorial. In the following list, note that ICD-O-3 codes C71.0 and C71.9 include both supratentorial and infratentorial subsites.
  Supratentorial sites by ICD-O-3 codes:
  C71.0, except hypothalamus, pallium, thalamus
  C71.1-C71.5
  C71.8: Corpus callosum, tapetum
  C71.9: Anterior cranial fossa, middle cranial fossa, suprasellar
  Infratentorial sites by ICD-O-3 codes:
  C71.0: Hypothalamus, pallium, thalamus
  C71.6-C71.7
  C71.9: Posterior cranial fossa

Code	Description	TNM 7 Map	TNM 6 Map	SS77 Map	SS2000 Map
050	Benign or borderline brain tumor	NA	NA	NA	NA
100	Supratentorial tumor confined to: Cerebral hemisphere (cerebrum) or Meninges of cerebral hemisphere (one side): Frontal lobe Occipital lobe Parietal lobe Temporal lobe	NA	NA	L	L
110	Infratentorial tumor confined to cerebellum or meninges of cerebellum (one side or midline): Vermis Lateral lobes Median lobe of cerebellum	NA	NA	L	L
120	Infratentorial tumor confined to: Brain stem or meninges of brain stem (one side): Medulla oblongata Midbrain (mesencephalon)	NA	NA	L	L

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Brain

CS Mets at DX

- Note 1: According to AJCC, p. 594: "Tumors affecting the central nervous system rarely develop extraneural metastases, probably because of inherent biologic characteristics of these tumors, and also because the brain does not have a well-developed drainage system. . . . Certain tumors do spread through cerebrospinal fluid (CSF) pathways, and such spread has a major impact on survival."

Code	Description	TNM 7 Map	TNM 6 Map	SS77 Map	SS2000 Map
00	No distant metastasis	NA	NA	NONE	NONE
10	OBSOLETE DATA RETAINED AND REVIEWED V0203 See cdodes 20, 30, and 50 Distant metastases	NA	NA	D	D
20	Metastasis within CNS and CSF pathways "Drop" metastases	NA	NA	D	D
30	Metastasis outside the CNS Extra-neural metastasis	NA	NA	D	D
50	30 + 20 Metastasis within and outside CNS	NA	NA	D	D
85	OBSOLETE DATA CONVERTED V0203 See code 20 "Drop" metastases	ERROR	ERROR	ERROR	ERROR
99	Unknown; distant metastasis not stated Distant metastasis cannot be assessed Not documented in patient record	NA	NA	U	U

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IntracranialGland

Pituitary Gland, Craniopharyngeal Duct, and Pineal Gland

C75.1, C75.2, C75.3

- C75.1 Pituitary gland
- C75.2 Craniopharyngeal duct
- C75.3 Pineal Gland
- Note: AJCC does not define TNM staging for this schema.

CS Tumor Size
CS Extension
CS Tumor Size/Ext Eval = **9**
OBSOLETE - CS Lymph Nodes
CS Lymph Nodes Eval = **9**
Regional Nodes Positive = **99**
Regional Nodes Examined = **99**
CS Mets at DX
CS Mets Eval = **9**
CS Site-Specific Factor 1
World Health Organization (WHO) Grade Classification
CS Site-Specific Factor 2
Ki-67/MIB-1 Labeling Index (LI): Brain
CS Site-Specific Factor 3 = **988**
CS Site-Specific Factor 4 = **988**
CS Site-Specific Factor 5 = **988**
CS Site-Specific Factor 6 = **988**
CS Site-Specific Factor 7 = **988**
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CS Site-Specific Factor 21 = **988**
CS Site-Specific Factor 22 = **988**
CS Site-Specific Factor 23 = **988**
CS Site-Specific Factor 24 = **988**
CS Site-Specific Factor 25 = **988**

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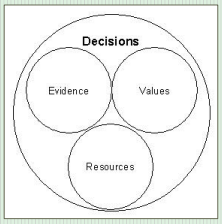
Treatment



Surgical Option(s)

- Decisions regarding aggressiveness of surgery for primary brain lesions are complex and depend on the:
  - Age and performance status of the patient
  - Proximity to "eloquent" areas of the brain
  - Feasibility of decreasing the mass effect with aggressive surgery
  - Resectability of the tumor (including the number and location of lesions)
  - In patients with recurrent disease, the time since the last surgery
- Surgical options include:
  - Stereotactic biopsy
  - Open biopsy or debulking procedure
  - Subtotal resection
  - Maximal safe resection

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Craniotomy

Any bony opening that is cut into the skull.

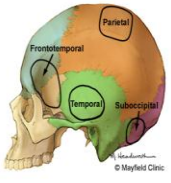


Figure 1. Craniotomies are often named for the bone being removed. Some common craniotomies include frontotemporal, parietal, temporal, and suboccipital.

81

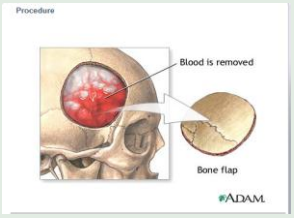
Source: Mayfield Clinic

Craniotomy Procedure

A section of the skull, (called a bone flap) is removed to access the brain underneath.

Typically the bone flap is replaced.

If the flap is not replaced, the procedure is called a craniectomy



82

Source: MedlinePlus/US National Library of Medicine, NIH

Surgeon has drawn the cutline circle around the tumor location

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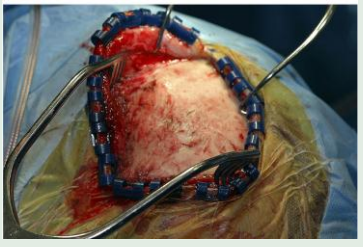
Source: The Alien-a set on Flickr www.flickr.com/photos/woodcreeper/sets/598206

Surgeon has cut the scalp and pulled it back to expose the skull over the tumor



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Source: The Alien-a set on Flickr www.flickr.com/photos/woodcreeper/sets/598206

The skull is removed revealing the dura layer under which is the brain and tumor

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Source: The Alien-a set on Flickr www.flickr.com/photos/woodcreeper/sets/598206

Here you see the circular cut through the dura layer with the brain and tumor exposed.

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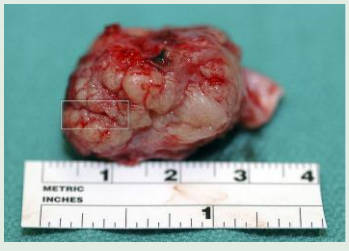
Source: The Alien-a set on Flickr www.flickr.com/photos/woodcreeper/sets/598206

Meningioma Resected



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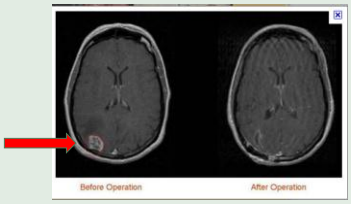
Source: The Alien-a set on Flickr www.flickr.com/photos/woodcreeper/sets/598206

Pre- and Post-Operative Imaging

Pre-op tumor is outlined in red

Post-operative MRI shows complete resection of the tumor



Source: Desert Spine and Neurosurgical Institute

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Surgery Codes

APPENDIX B: Site-Specific Surgery Codes 283

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

(Except for M-9727, 9733, 9741-9742, 9764-9809, 9832, 9840-9931, 9945-9946, 9950-9967, and 9975-9992)

Do not code laminectomies for spinal cord primaries.

Codes

00 None; no surgery of primary site; autopsy ONLY

10 Tumor destruction, NOS

No specimen sent to pathology from surgical event 10.

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.

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Surgery Codes

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 can not be coded as 20-30.

55 Gross total resection of lobe of brain (lobectomy)

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

Specimen sent to pathology from surgical events 20–55.

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Radiation Therapies

- Primary XRT for brain tumors includes tumor volume/margins
- Tumor volume is defined by pre- and post-operative imaging
- Standard fractionated external beam radiation is most common
- Hypofractionation (daily dose given in smaller increments with 4 or 6 hours between treatments) is an emerging option
- Whole brain XRT and stereotactic radiosurgery for brain mets

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Source: NCCN

Tumor Volume

- The larger the brain tumor, the more desirable "fractionation" (e.g. multiple smaller treatments, rather than one big one)
- Tumor size can determine schedule for fractionation and dose/tx
- Why: The "shell" of normal tissue outside the tumor volume will receive some part of the dose. For larger tumors, this "shell" volume increases rapidly as a function of tumor diameter
- Why: Fractionation spares this "shell" of normal tissue much more effectively than the single "shot" techniques

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Source: Johns Hpkins Medicine

Stereotactic Radiosurgery (SRS)

- Despite name, SRS is an XRT treatment, not a surgical procedure
- Acoustic neuroma, pituitary tumors, spinal cord tumors and brain metastasis are candidates for this technique
- Special equipment focuses up to 200 beams of radiation on tumor
- Although each beam has very little effect on the brain tissue it passes through, a strong dose of radiation is delivered to the site where all the beams meet.
- Results in minimal damage to healthy tissues surrounding target.

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Zdpirce" Mayo Clinic

Gamma Knife Perfexion	CyberKnife
	
Designed exclusively for non-invasive brain surgery with 192 cobalt-60 beams that converge and focus on the treatment target	Uses a single-source linear accelerator; not exclusive to brain surgery
Radiological accuracy better than 0.3mm	1 mm accuracy; dose **outside** the target area is 2 to 6 times greater than with the Gamma Knife
Rigid immobilization to prevent head movement using a lightweight stereotactic head frame fixed to the outer skull. Provides exact MR and CT correlation from planning to treatment delivery in 3D.	Non-rigid immobilization reduces head movement by using a thermoplastic face mask that is shrink-wrapped to the table during treatment. Provides relative MR and CT correlation from planning to treatment delivery in 3D. The CyberKnife is inherently less accurate because head positioning is optically guided, not head-frame based.
Treatment delivered during one session	Single or multiple treatments, possibly over a period of days
Target is confirmed 10 times per second	Target is confirmed once every 10 seconds

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Source: San Diego Gamma Knife Center

Chemotherapy

- Chemotherapy is not an effective initial treatment for low-grade brain tumors. Why? Because most standard chemo agents have a hard time passing into the brain because of how the brain protects itself (the blood-brain barrier)
- Not all types of brain tumors respond to chemotherapy
- In general, chemotherapy for brain tumors is usually administered following surgery or radiation therapy
- Participation in clinical trials should be encouraged

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Blood Brain Barrier

- Composed of special cells that make up brain's blood vessels
- Selectively prevents substances from entering the blood and brain, only allowing essential molecules such as amino acids, oxygen, glucose and water through
- Adenosine, a molecule produced by the body, seems to modulate the entry of large molecules into the brain
- When adenosine receptors are activated on cells that comprise the blood-brain barrier, a gateway into the barrier can be established

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Science Daily Source: September 13, 2001

Approved Chemotherapy Agents

- Carmustine (BCNU) – IV or dissolvable wafers placed surgically
- Temozolomide (Temodar) – oral
- Lomustine (CCNU) – oral
- Carboplatin
- Cisplatin
- Etoposide
- Irinotecan
- Vincristine
- Procarbazine (Matulane) – oral
- Methotrexate - oral, by injection or intrathecally

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NCCN Treatment Guidelines

Infiltrative Low-Grade Glioma

- Best management strategy has yet to be defined
- Small tumor samples can provide a lower histologic grade
- Rationale: Needle biopsies are often performed when lesions are in deep or critical regions of the brain, but can be misleading because gliomas often have varying degrees of cellularity, mitosis, or necrosis from one region to another
- General recommendation is to first attempt as complete an excision of tumor as possible (based on postsurgical MRI verification) without compromising function
- No consensus exists regarding proper timing of postoperative external beam radiation
- Chemotherapy is not a traditional upfront treatment modality

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Infiltrative Low-Grade Glioma

- When possible, maximal safe resection
- If gross total resection is achieved, some patients may be observed without adjuvant therapy. However, close follow-up is essential as over half of patients will eventually progress
- These tumors behave aggressively in patients over 40 years old
  - Adjuvant radiation or chemotherapy is recommended
- If stereotactic biopsy, open biopsy, or other subtotal excision was done, immediate fractionated external beam RT or chemotherapy should be given

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Anaplastic Glioma and Glioblastoma

- Whenever possible, major tumor removal should be performed
- If glioblastoma is confirmed, options include radiation, chemotherapy, best supportive care, chemoradiation only if carmustine wafer was implanted
- If high-grade glioma is confirmed, BCNU wafer is an option
  - In patients with good Karnofsky score (70 or above) options include fractionated external beam radiation therapy, chemotherapy or chemoradiation in the context of a clinical trial
  - In patients with poor Karnofsky score (below 70) management may include radiation, chemotherapy or best supportive care

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Intracranial Ependymoma

- Whenever possible, maximal safe resection should be attempted
- Adjuvant treatment depends on the extent of surgical resection, histology and staging by cranial spinal MRI and CSF cytology
- CSF dissemination occurs in up to 15% of intracranial ependymomas
- If MRI spine /CSF reveal disease, craniospinal radiation is mandatory
- If gross total resection with negative spinal MRI and CSF, adjuvant regional fractionated EBRT or observation may be considered

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Medulloblastoma and PNET (supratentorial)

- MRI is the gold standard to assess PNET
- Maximal safe resection is recommended when possible
- Average Risk Patients: craniospinal radiation alone or concurrent chemoradiation followed by chemotherapy are both options
- High Risk Patients: patients with large cell or anaplastic medulloblastoma, supratentorial PNET, disease dissemination, unresectable tumors, or residual tumors over 1.5cm post-surgery are high risk and should undergo radiation followed by chemotherapy

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Primary CNS Lymphoma

- Treatment to be initiated as immediately following diagnosis
- Treatment options depend on patient overall health and age
- For healthier patients a high-dose methotrexate regimen
- RT after systemic treatment depends on the responsiveness of the disease to the chemotherapy
- However, one or both may increase neurotoxicity, especially in patients older than 60 years of age

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Primary Spinal Cord Tumors

- MRI is the gold standard for diagnosis of spinal cord lesions
- Asymptomatic patients may be observed or resected
- Symptomatic patients should undergo some form of surgery
- Maximal safe resection should be attempted
- Post-operative adjuvant radiation is not recommended
- However, if symptoms persist after incomplete resection or biopsy, radiation should be administered

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Meningioma

- Meningiomas are typically diagnosed by CT or MRI imaging
- Biopsy may be considered for confirmation
- Options stratified by presence/absence of symptoms and tumor size
- Most asymptomatic patients with small tumors (<30mm) may just be observed. If neurological impairment is imminent, surgery (if accessible) or radiotherapy (EBRT OR SRS) is feasible
- Asymptomatic tumors >30mm can be either resected or observed

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Meningioma

- Symptomatic disease requires active treatment by surgery if possible
- Non-surgical candidates should undergo radiation
- All patients with surgically resected grade III meningiomas (even after gross total resection) should receive adjuvant radiation for local control regardless of tumor size and symptom status

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Additional Resources

- **NCCN Evidence Based Treatment Guidelines**, nccn.org, 2011
- **Collaborative Stage Data Collection System**, AJCC, 2010
- **Multiple Primary and Histology Coding Rules**, SEER 2007
- **The 2007 WHO Classification of Tumours of the Central Nervous System,** David N. Louis, Hiroko Ohgaki, Otmar D. Wiestler, Webster K. Cavenee, Peter C. Burger, Anne Jouvet, Bernd W. Scheithauer and Paul Kleihues, World Health Organization, Lyon, France, 2007
- **Data collection of primary central nervous system tumors. National Program of Cancer Registries Training Materials.** Department of Health and Human Services, Centers for Disease Control and Prevention. Atlanta, Georgia, 2004.

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QUESTIONS ???

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