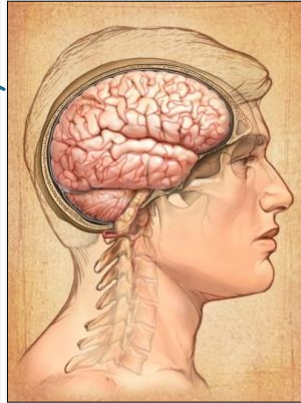


Overview



Brain tumors are:

- Primary “brain” tumors - those that begin in the brain or central nervous system (or its supporting tissues) 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.

Brain tumors are:

- Usually described as intracranial neoplasms with varying behaviors (benign, borderline, malignant – ref. ICD-O-3)
- Include most identifiable structures within the cranium including the brain itself, small hormone-secreting ducts like the pineal and pituitary glands, the cranial nerves (primarily the optic nerve, olfactory nerve, acoustic nerve), the outer protective lining of the brain (meninges), and the spinal cord.
- Metastatic neoplasms are excluded
- Certain benign “tumors” are excluded
- No malignant neoplasms are excluded
- Benign bone tumors are excluded

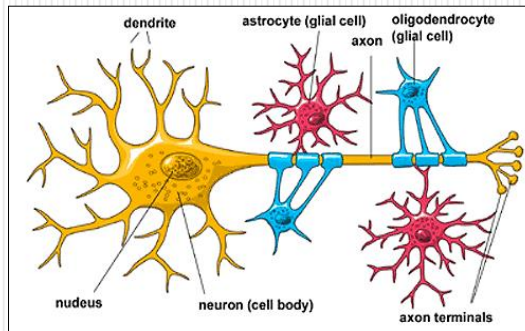
5

Brain Tumor Characteristics

- Start in the brain and grow steadily there.
- Very rarely spread to other organs through the bloodstream.
- Are named for the anatomic location of tumor and/or the cells from which they arise, each having a certain function essential to normal physiological functioning of the brain.
- For example:
 - Brain Stem Gliomas arise in the lowest part of the brain.
 - Meningiomas arise in the meninges.
 - Gliomas arise from glial cells that support the CNS.
 - Astrocytomas arise from astrocytes
 - Ependymomas arise from ependymal cells which line the ventricles
 - Oligodendrogliomas arise from oligodendrocyte cells which make up the fatty substance called myelin that covers nerves like electrical insulation.

6

Characteristics of Brain Tumors



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

7

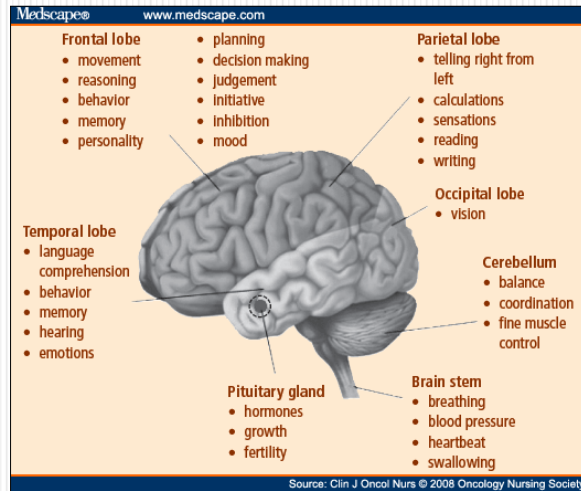
Range of Tumors and Symptoms

- 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.

8

SEER Training Modules

Brain Anatomy and Function



9

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. 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).

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SEER Training Modules

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.

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ALL Brain Tumors are Reportable

- **BUT** – Do Not Report Benign/Borderline Tumors that were Diagnosed BEFORE 1/1/2004 – NOT EVEN HISTORICAL
- This creates an ERROR for FCDS at Call for Data
- FCDS then has to Delete the Case and Adjust Sequences

12

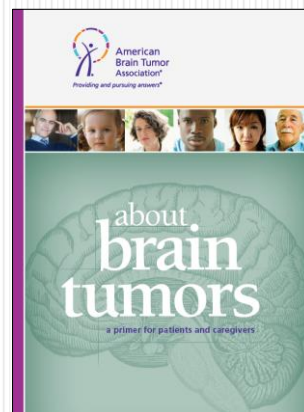
ICD-O Topography Codes (Anatomic Site)

- | | |
|--|--|
| <ul style="list-style-type: none"> • 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) | <ul style="list-style-type: none"> • Meninges (C70.0 - C70.9) <ul style="list-style-type: none"> ◦ 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) <ul style="list-style-type: none"> ◦ 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) |
|--|--|

13

Brain Tumor Characteristics

- Benign – Borderline – Malignant
- Patient Age
- Tumor Location
- Tumor Histologic Type
- WHO Grade of Primary Tumor



American Brain Tumor Association – <http://www.abta.org>

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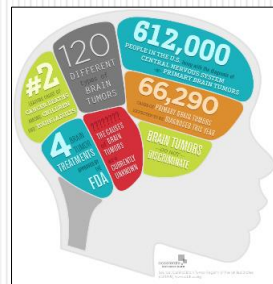
Tumor Behavior

- **Benign Tumors** • *Slow growing* • *Distinct borders* • *Rarely spread*
- **Malignant Tumors** • *Usually rapid growing* • *Invasive* • *Life threatening*
- **Borderline Malignant Tumors** • *Rare* • *Likely to Recur following Surgical Resection* • *May become Life threatening*

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Common Histologic Types

- Astrocytoma, Grade I (Juvenile Pilocytic Astrocytoma)
- Astrocytoma, Grade II (Low Grade Pilocytic Astrocytoma)
- Astrocytoma, Grade III (Anaplastic Astrocytoma)
- Glioblastoma (Glioblastoma Multiforme/ Astrocytoma, Grade IV)
- Ependymoma
- Medulloblastoma
- Meningioma
- Oligodendroglioma
- Oligoastrocytoma
- Pituitary Adenoma



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Figure 13. Most Common Brain and CNS Tumors by Age

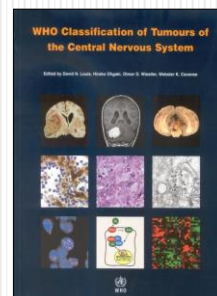
CBTRUS Statistical Report: NPCR and SEER Data from 2004-2007

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/glia
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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WHO Classification Groups

- Tumors of Neuroepithelial Tissue
- Tumors of Cranial and Paraspinal Nerves
- Tumors of Meninges
- Tumors of Uncertain histogenesis
- Lymphomas and Hematopoietic Malignancies
- Germ Cell Tumors
- Cysts and Tumor-Like Lesions
- Tumors of the Sellar Region
- Metastatic Tumors



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Table 1 The 2007 WHO Classification of Tumours of the Central Nervous System. Reprinted from Ref. 35

TUMOURS OF NEUROEPITHELIAL TISSUE**Astrocytic tumours**

Piloicytic astrocytoma	9421/1*
Piloxyoid astrocytoma	9425/2*
Subependymal giant cell astrocytoma	9384/1
Plasmocytic xanthoastrocytoma	9424/3
Diffuse astrocytoma	9400/3
Fibrillary astrocytoma	9400/3
Gemistocytic astrocytoma	9411/3
Protoplasmic astrocytoma	9410/3
Anaplastic astrocytoma	9401/3
Glioblastoma	9400/3
Giant cell glioblastoma	9441/3
Gliosarcoma	9440/3
Gliomatosis cerebri	9391/3

Oligodendroglial tumours

Oligodendroglioma	9420/3
Anaplastic oligodendroglioma	9451/3

Oligoastrocytic tumours

Oligoastrocytoma	9382/3
Anaplastic oligoastrocytoma	9392/3

Ependymal tumours

Subependymoma	9393/1
Myxopapillary ependymoma	9394/1
Ependymoma	9391/3
Cellular	9391/3
Papillary	9392/3
Clear cell	9391/3
Tanycytic	9391/3
Anaplastic ependymoma	9392/3

Choroid plexus tumours

Choroid plexus papilloma	9390/0
Atypical choroid plexus papilloma	9390/1*
Choroid plexus carcinoma	9390/3

Other neuroepithelial tumours

Astroblastoma	9430/3
Choroid glioma of the third ventricle	9444/1
Angioepithelial glioma	9451/1*

*Topography code of the International Classification of Diseases for Oncology (2002) (ICD-O), and the International Classification of Diseases for Oncology (2002) (ICD-O-2) denotes a code 0 for benign tumours, 1 for malignant tumours and 1 for borderline or uncertain tumours.

*The tumour is not a primary, but originates in the wall of the ventricle. It is expected to be incorporated into the next ICD-O edition, the current version adapted to merge.

Neuronal and mixed neuronal-glia tumours

Dysplastic gangliocytoma of cerebellum (Lissencephalic)	9493/0
Desmoplastic infantile astrocytoma/ganglioglioma	9412/1
Dysembryoplastic neuroepithelial tumour	9413/0
Gangliocytoma	9400/0
Ganglioglioma	9501/1
Anaplastic ganglioglioma	9501/3
Central neurocytoma	9501/1
Extraventricular neurocytoma	9501/1*
Cerebellar liponeurocytoma	9501/1*
Papillary glioneuronal tumour	9501/1*
Rosette-forming glioneuronal tumour of the fourth ventricle	9501/1*
Paraganglioma	8680/1

Tumours of the pineal region

Pineocytoma	9361/1
Pinoid pineocytoma tumour of intermediate differentiation	9362/3
Pineoblastoma	9362/3
Papillary tumour of the pineal region	9395/3*

Embryonal tumours

Medulloblastoma	9470/3
Desmoplastic nodular medulloblastoma	9471/3
Medulloblastoma with extensive nodularity	9471/3*
Anaplastic medulloblastoma	9474/3*
Large cell medulloblastoma	9474/3
CHS primitive neuroectodermal tumour	9473/3
CHS Neuroblastoma	9500/3
CHS Ganglioneuroblastoma	9480/3
Medulloepithelioma	9501/3
Ependymoblastoma	9390/3
Atypical teratoid / rhabdoid tumour	9508/3

TUMOURS OF CRANIAL AND PARASPINAL NERVES

Schwannoma (neurilemma, neurinoma)	9500/0
Cellular	9500/0
Plexiform	9500/0
Melanotic	9500/0
Neurofibroma	9540/0
Plexiform	9550/0

Table 1 continued

Petrous tumor	9571/0	Haemangiopericytoma	9150/1
Petroneuritis, NOS	9571/0	Anaplastic haemangiopericytoma	9150/3
Malignant petrous tumor	9571/3	Angiosarcoma	9120/3
		Kaposi sarcoma	9140/3
		Ewing sarcoma - PNET	9364/3

TUMOURS OF THE MENINGES

Tumours of meningeothelial cells	9530/0
Meningioma	9531/0
Meningothelial	9531/0
Fibrous (fibroblastic)	9531/0
Transitional (mixed)	9531/0
Psammomatous	9531/0
Angiomatous	9531/0
Microcystic	9531/0
Secretory	9531/0
Lymphoplasmacytic-rich	9531/0
Meningeal	9531/0
Chordoid	9531/0
Clear cell	9531/0
Atypical	9531/0
Papillary	9531/0
Rhabdoid	9531/0
Anaplastic (malignant)	9531/0

Mesenchymal tumours

Lipoma	8850/0
Angiolipoma	8851/0
Hibernoma	8850/0
Liposarcoma	8850/3
Solitary fibrous tumour	8815/0
Fibrosarcoma	8810/3
Malignant fibrous histiocytoma	8850/3
Leiomyosarcoma	8890/3
Rhabdomyoma	8900/0
Rhabdomyosarcoma	8900/3
Chondroma	9220/0
Chondrosarcoma	9220/3
Osteoma	9180/0
Osteosarcoma	9180/3
Osteochondroma	9210/0
Haemangioma	9120/0
Epithelioid haemangioendothelioma	9133/1

Primary melanocytic lesions

Diffuse melanocytosis	8728/0
Melanocytoma	8728/1
Malignant melanoma	8720/3
Meningeal melanomatosis	8728/3

Other neoplasms related to the meninges

Haemangioblastoma	9161/1
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LYMPHOMAS AND HAEMATOPOIETIC NEOPLASMS

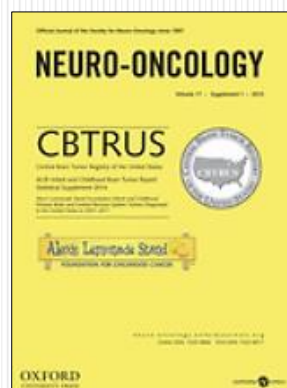
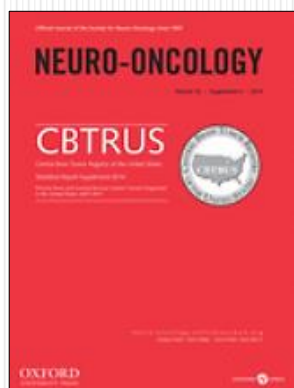
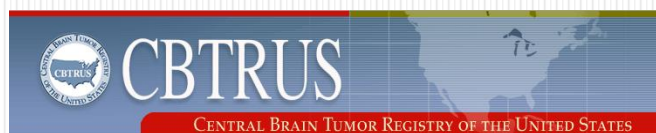
Malignant lymphomas	9590/3
Plasmacytoma	9731/3
Granulocytic sarcoma	9590/3

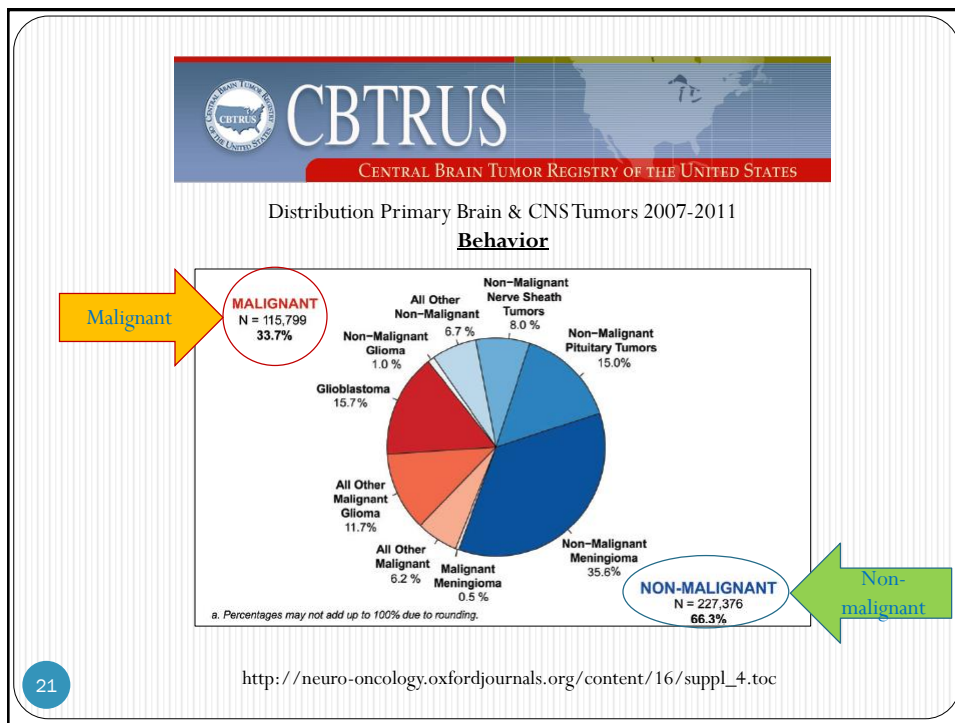
GERM CELL TUMOURS

Germioma	9064/3
Embryonal carcinoma	9070/3
Yolk sac tumour	9071/3
Choriocarcinoma	9100/3
Teratoma	9080/1
Mature	9080/0
Immature	9080/3
Teratoma with malignant transformation	9084/3
Mixed germ cell tumour	9085/3

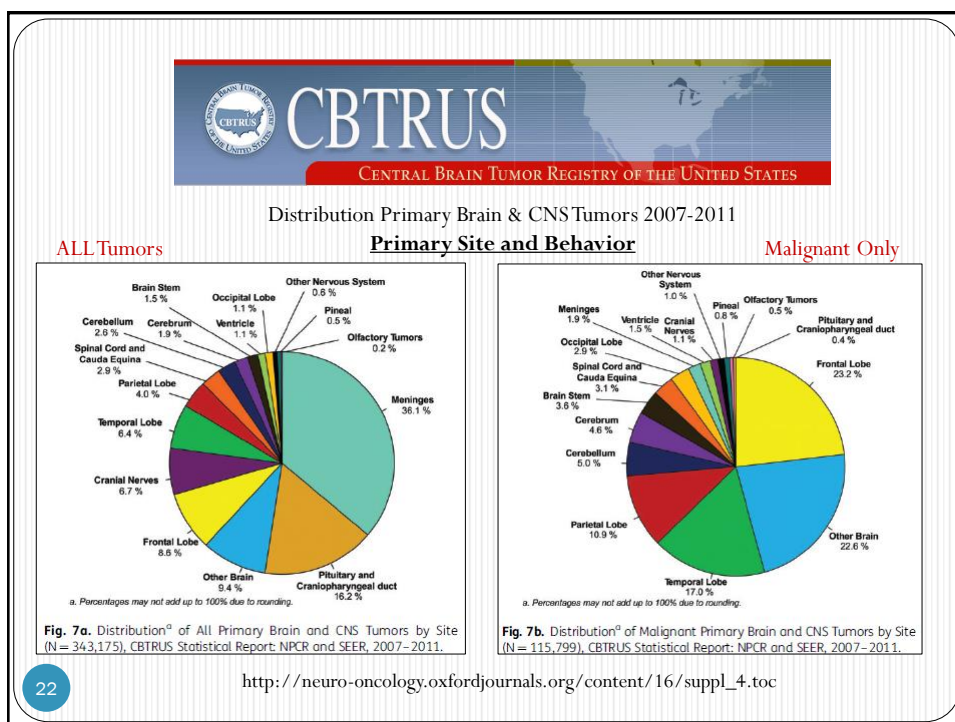
TUMOURS OF THE SELLAR REGION

Craniopharyngioma	9550/1
Adenomatous	9551/1
Papillary	9552/1
Granular cell tumour	9550/0
Pituitary	9430/1*
Spindle cell oncocytoma of the adenohypophysis	8291/0*

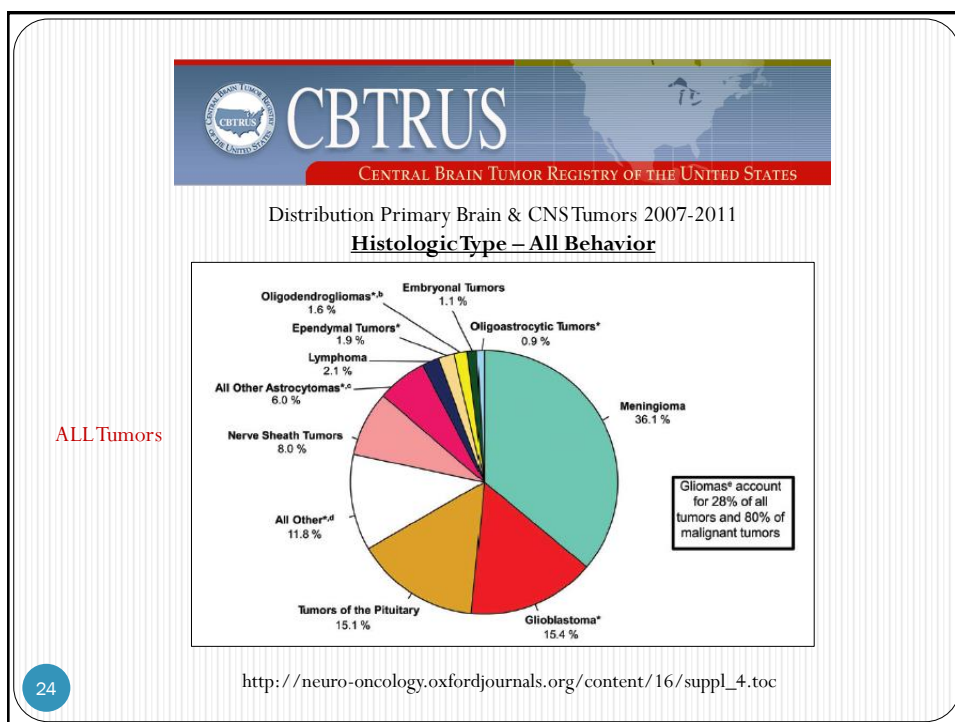
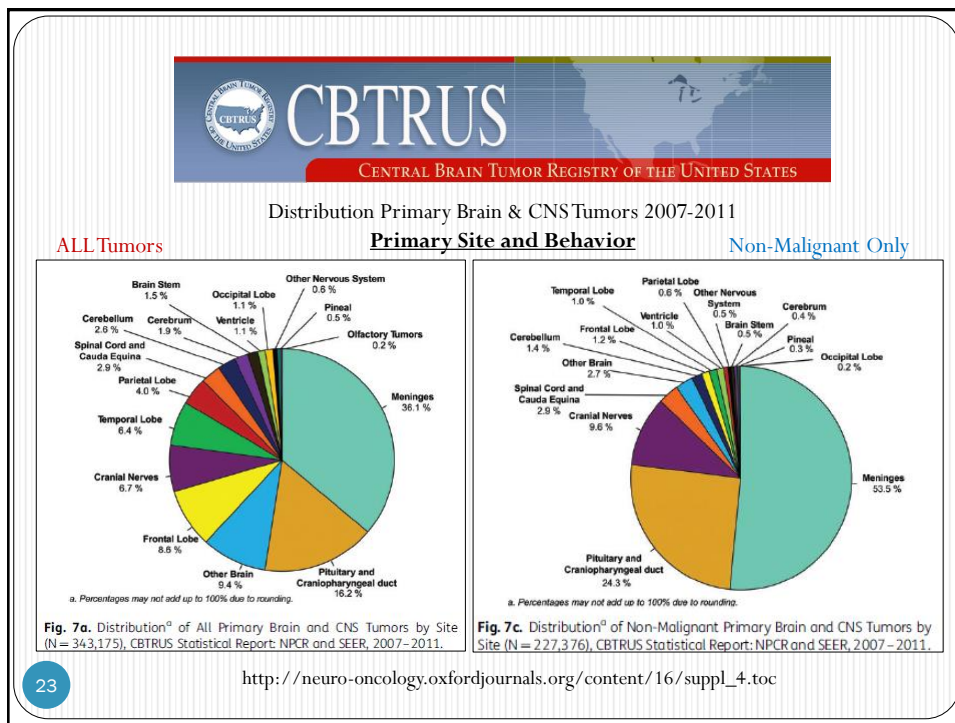
METASTATIC TUMOURS

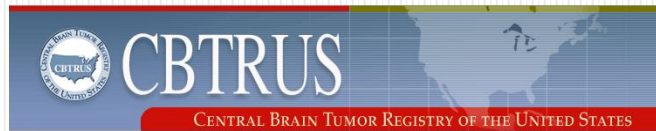


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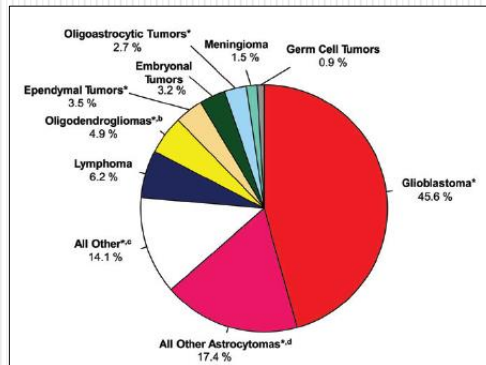




Distribution Primary Brain & CNS Tumors 2007-2011

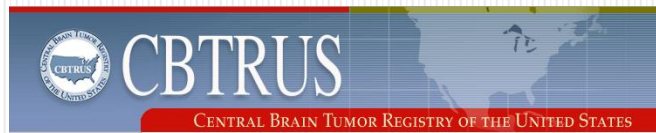
Behavior and Histologic Type

Malignant
Only



http://neuro-oncology.oxfordjournals.org/content/16/suppl_4.toc

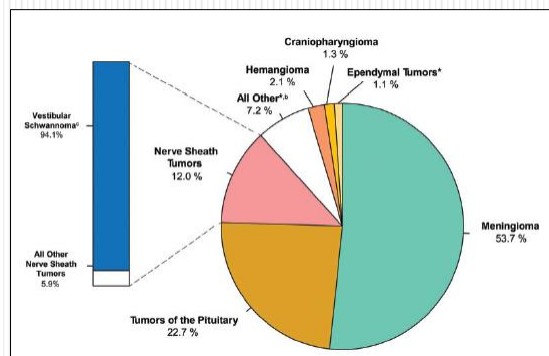
25



Distribution Primary Brain & CNS Tumors 2007-2011

Behavior and Histologic Type

Non-
Malignant
Only



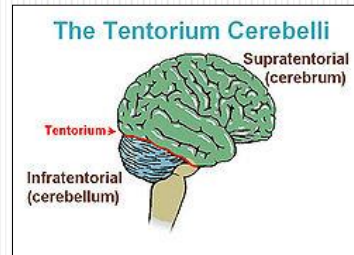
http://neuro-oncology.oxfordjournals.org/content/16/suppl_4.toc

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Childhood Brain Tumors

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

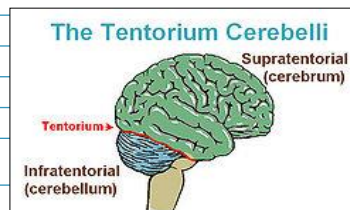
- 50% of childhood brain and CNS tumors are infratentorial, originating below the tentorium
- 20+% of childhood CNS tumors are located in the sellar or suprasellar region around the sella turcica (the bone that contains the pituitary gland)
- Remainder of tumors occur in spinal cord, brain stem, cranial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



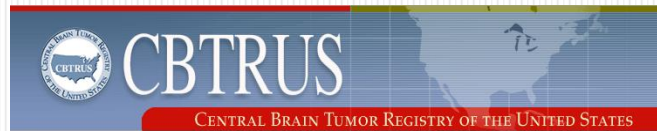
27

Childhood Brain Tumors

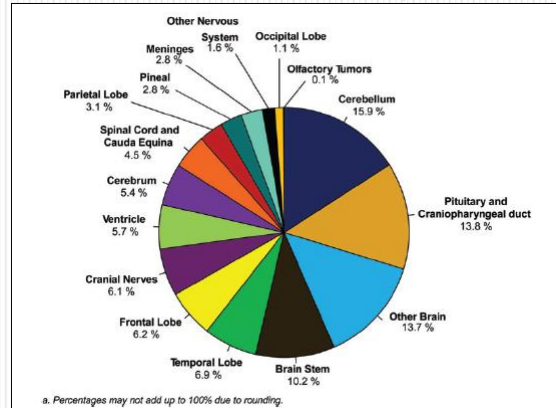
Supratentorial - childhood	Infratentorial - childhood
• Craniopharyngiomas.	• Cerebellar astrocytomas (usually high-grade).
• Diencephalic and hypothalamic gliomas.	• Medulloblastomas (primitive neuroectodermal tumors).
• Germ cell tumors.	• Ependymomas (low-grade or anaplastic).
• Low-grade astrocytomas.	• 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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Distribution Primary Brain & CNS Tumors 2007-2011
Children and Adolescents by Site

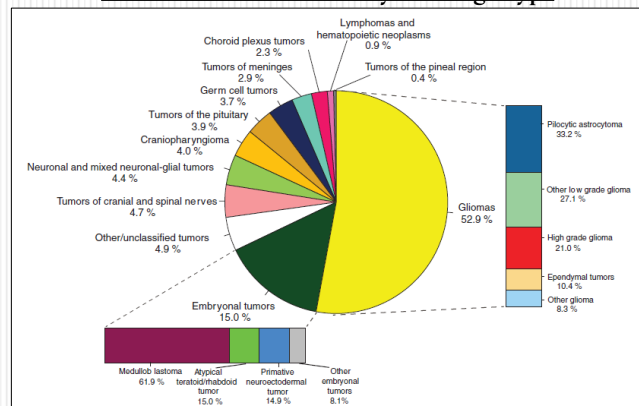


http://neuro-oncology.oxfordjournals.org/content/16/suppl_4.toc

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Distribution Primary Brain & CNS Tumors 2007-2011
Children and Adolescents by Histologic Type



http://neuro-oncology.oxfordjournals.org/content/16/suppl_10?etoc

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WHO Grade for Brain/CNS Tumors

- WHO Tumor Grades – Grade I, II, III, and IV
- Higher the grade – the more malignant the tumor
- A tumor can contain more than one grade of cell
- Always record and code the highest tumor grade noted
- Record WHO Grade in SSF1 NOT in “Grade/Differentiation”

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<http://www.abta.org/brain-tumor-information/diagnosis/grading-staging.html>

WHO Grade for Brain/CNS Tumors

LOW GRADE Neoplasms

- **Grade I:** least malignant tumors associated with long-term survival. They grow slowly and have an almost normal appearance when viewed through a microscope. Surgery alone may be an effective treatment for this grade tumor.
- **Grade II:** tumors are slow-growing and look slightly abnormal under a microscope. Some can spread into nearby normal tissue and recur, sometimes as a higher grade tumor.

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<http://www.abta.org/brain-tumor-information/diagnosis/grading-staging.html>

WHO Grade for Brain/CNS Tumors

- **Grade III:** These tumors are, by definition, malignant although there is not always a big difference between grade II and grade III tumors. The cells of a grade III tumor are actively reproducing abnormal cells, which grow into nearby normal brain tissue. These tumors tend to recur, often as a grade IV.
- **Grade IV:** The most malignant tumors. Tumors reproduce rapidly, can have a bizarre appearance when viewed under the microscope, and easily grow into nearby normal brain tissue. These tumors form new blood vessels so they can maintain their rapid growth. They also have areas of dead cells in their centers.

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<http://www.abta.org/brain-tumor-information/diagnosis/grading-staging.html>

WHO Grade for Brain/CNS Tumors

WORLD HEALTH ORGANIZATION (WHO) GRADING SYSTEM

Grade I Tumor

- Slow-growing cells
- Almost normal appearance under a microscope
- Least malignant
- Usually associated with long-term survival

Grade II Tumor

- Relatively slow-growing cells
- Slightly abnormal appearance under a microscope
- Can invade adjacent normal tissue
- Can recur as a higher grade tumor

Grade III Tumor

- Actively reproducing abnormal cells
- Abnormal appearance under a microscope
- Infiltrate adjacent normal brain tissue
- Tumor tends to recur, often as a higher grade

Grade IV Tumor

- Abnormal cells which reproduce rapidly
- Very abnormal appearance under a microscope
- Form new blood vessels to maintain rapid growth
- Areas of dead cells (necrosis) in center

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<http://www.abta.org/brain-tumor-information/diagnosis/grading-staging.html>

Name	Grade	Description
Pilocytic astrocytoma	I	Common in children and young adults and in people with neurofibromatosis type 1 and rarely causes death
Diffuse astrocytoma	II	Also known as low-grade diffuse astrocytoma, it is common in young adults and in people with Li-Fraumeni syndrome. It commonly forms in the cerebrum but can form in any part of the brain. It grows slowly, often spreading into nearby tissues. It can progress to become an anaplastic astrocytoma or a glioblastoma
Anaplastic astrocytoma	III	Also known as malignant astrocytoma or high-grade astrocytoma, this kind of tumor is more often found in younger adults. It forms in the cerebrum. It grows quickly and spreads into nearby tissues. An anaplastic astrocytoma may transform and become a glioblastoma
Glioblastoma	IV	Also known as glioblastoma multiforme. About 50% of the gliomas are glioblastomas. It grows and spreads rapidly and has the highest degree of malignancy but poor prognosis

Adapted from American Brain Tumor Association (www.abta.org)

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Common pediatric brain tumors	Grade
Astrocytic tumors	
Subependymal giant cell astrocytoma	I
Pilocytic astrocytoma	
Pilomyxoid astrocytoma	II
Diffuse astrocytoma	
Pleomorphic xanthoastrocytoma	
Anaplastic astrocytoma	III
Glioblastoma	IV
Giant cell glioblastoma	
Gliosarcoma	
Ependymoma	
Subependymoma	I
Mixopapillary ependymoma	
Ependymoma	II
Anaplastic ependymoma	III
Embryonal tumor	
Medulloblastoma	IV
Central nervous system primitive neuroectodermal tumor	
Atypical teratoid/rhabdoid tumor	

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Table 2 WHO Grading of Tumours of the Central Nervous System. Reprinted from Ref. 35

	I	II	III	IV		I	II	III	IV
Astrocytic tumours									
Subependymal giant cell astrocytoma	*				Central neurocytoma		*		
Pilocytic astrocytoma	*				Extraventricular neurocytoma		*		
Piloxyoid astrocytoma		*			Cerebellar liponeurocytoma		*		
Diffuse astrocytoma		*			Paraganglioma of the spinal cord	*			
Pleomorphic xanthoastrocytoma		*			Papillary glioneuronal tumour	*			
Anaplastic astrocytoma			*		Rosette-forming glioneuronal tumour of the fourth ventricle	*			
Glioblastoma				*	Pineal tumours				
Giant cell glioblastoma				*	Pineocytoma	*			
Gliosarcoma				*	Pineal parenchymal tumour of intermediate differentiation		*	*	
Oligodendroglial tumours					Pineoblastoma				*
Oligodendroglioma		*			Papillary tumour of the pineal region	*	*		
Anaplastic oligodendroglioma			*		Embryonal tumours				
Oligoastrocytic tumours					Medulloblastoma				*
Oligoastrocytoma		*			CNS primitive neuroectodermal tumour (PNET)				*
Anaplastic oligoastrocytoma			*		Atypical teratoid / rhabdoid tumour				*
Ependymal tumours					Tumours of the cranial and paraspinous nerves				
Subependymoma	*				Schwannoma	*			
Myxopapillary ependymoma	*				Neurofibroma	*			
Ependymoma		*			Perineurioma	*	*	*	
Anaplastic ependymoma			*		Malignant peripheral nerve sheath tumour (MPNST)		*	*	*
Choroid plexus tumours					Meningeal tumours				
Choroid plexus papilloma	*				Meningioma	*			
Atypical choroid plexus papilloma		*			Atypical meningioma		*		
Choroid plexus carcinoma			*		Anaplastic / malignant meningioma			*	
Other neuroepithelial tumours					Haemangiopericytoma		*	*	
Angiocentric glioma	*				Anaplastic haemangiopericytoma		*	*	
Choroid glioma of the third ventricle		*			Haemangioblastoma	*			
Neuronal and mixed neuronal-glial tumours					Tumours of the sellar region				
Gangliocytoma	*				Craniohypopharyngioma	*			
Ganglioglioma	*				Granular cell tumour of the neurohypophysis	*			
Anaplastic ganglioglioma		*			Pituitary tumour	*			
Desmoplastic infantile astrocytoma and ganglioglioma	*				Spindle cell oncocytoma of the adenohypophysis	*			
Dysembryoplastic neuroepithelial tumour	*								

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Survival

Type of Tumor	5-Year Relative Survival Rate		
	Age		
	20-44	45-54	55-64
Low-grade (diffuse) astrocytoma	65%	43%	21%
Anaplastic astrocytoma	49%	29%	10%
Glioblastoma	17%	6%	4%
Oligodendroglioma	85%	79%	64%
Anaplastic oligodendroglioma	67%	55%	38%
Ependymoma/anaplastic ependymoma	91%	86%	85%
Meningioma	92%	77%	67%

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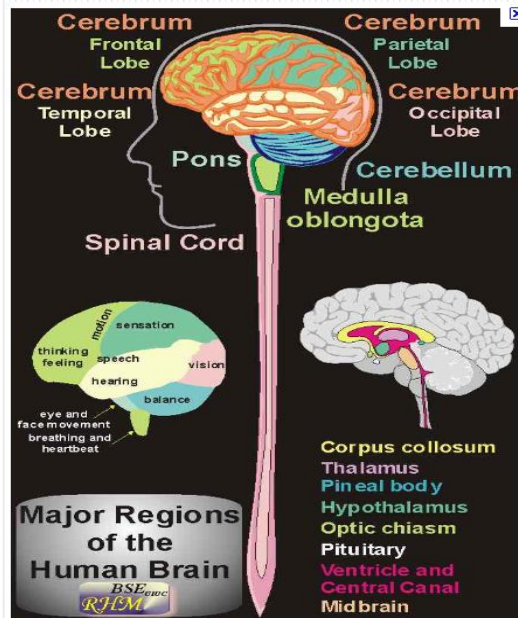
<http://www.cancer.org/cancer/brain-and-spinal-cord-tumors-in-adults-survival-rates>

ANATOMY OF THE HUMAN BRAIN



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

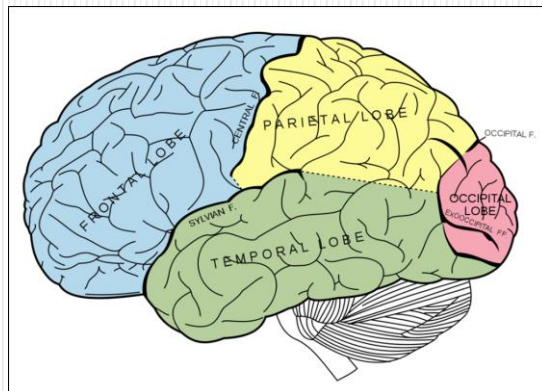
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40

Source: University of Illinois

Brain Lobes and Fissures



41

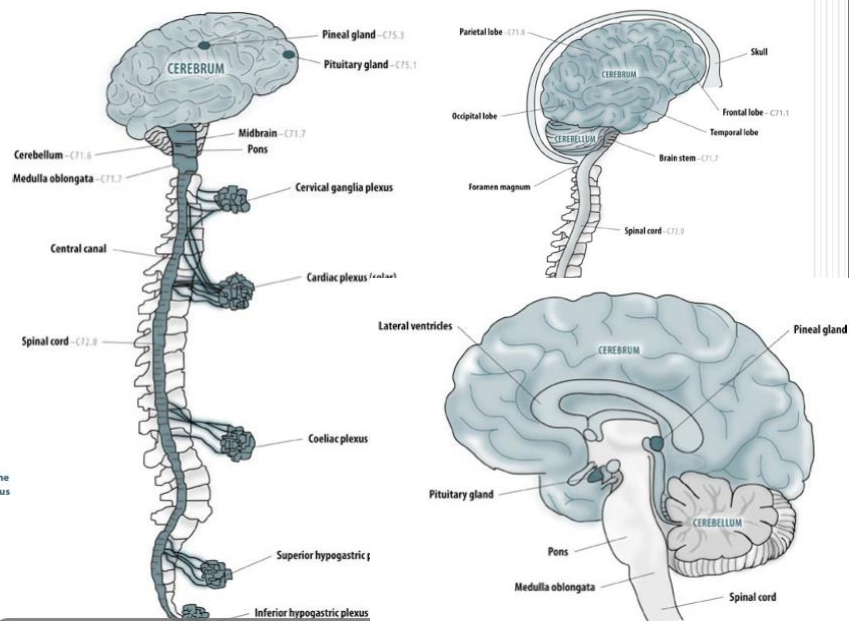
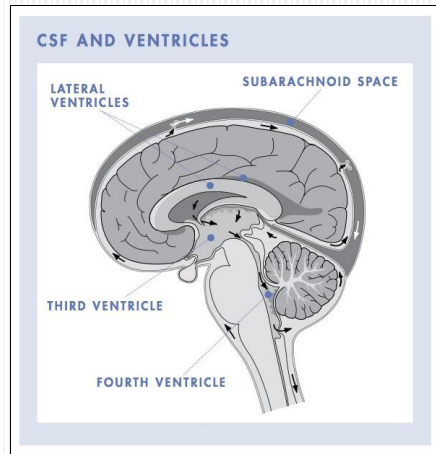


Figure 2.
Anatomy of the
central nervous
system

42

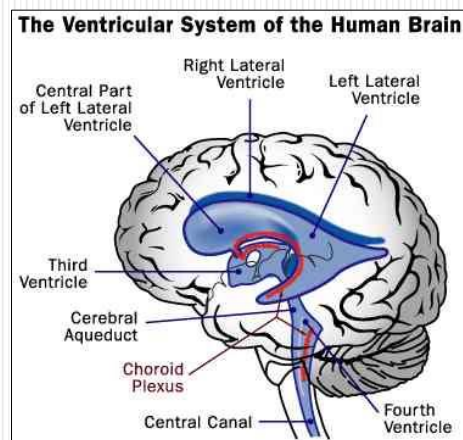
Ventricular System of the Brain



43

<http://www.abta.org/brain-tumor-information/brain-anatomy/structure.html>

Ventricular System of the Brain

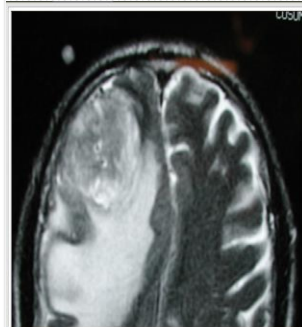


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Source: solarnavigator.net/human_brain

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 sitting 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.

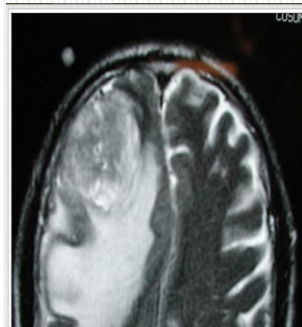


Source: Medscape

45

Midline Shift and Mass Effect

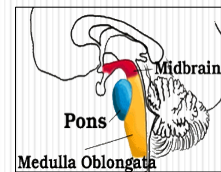
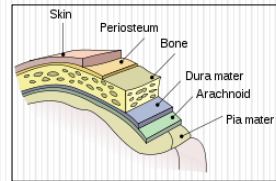
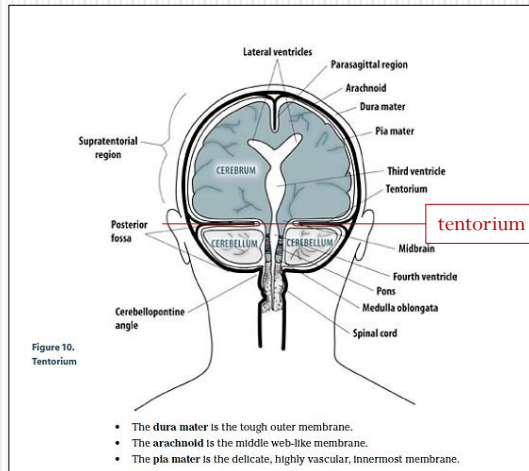
- Midline is a central boundary separating the left and right hemispheres.
- Midline Shift – Tumor crosses the brain to shift across the center line
- Mass Effect is – Edema or swelling causes the brain to shift across center line
- Both create new symptoms at cross-over
 - Depends on the size and location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



Source: Medscape

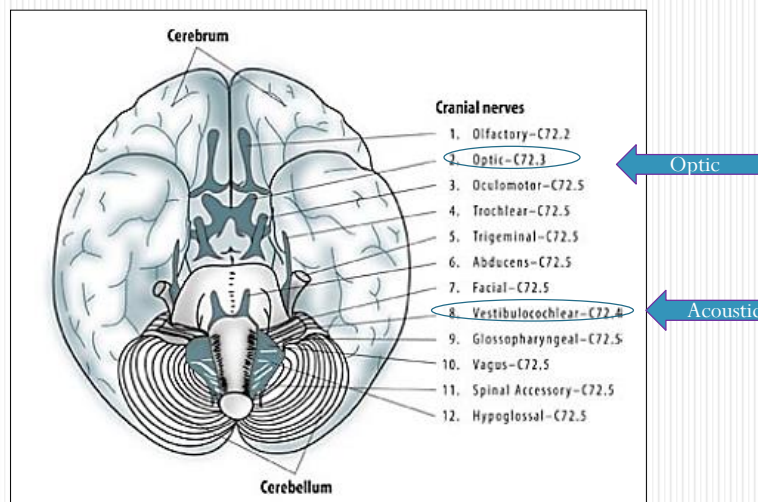
46

Meninges and Brain Stem



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Cranial Nerves



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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

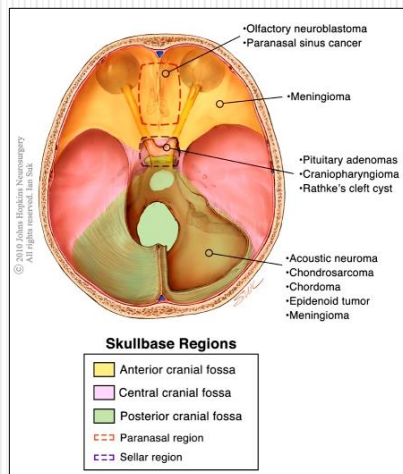
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Sinus, Olfactory, Base of Skull Tumors

- Cancer Registries treat many of these as Head & Neck Neoplasms
- Some are intra-cranial but many are not intra-cranial or CNS
- Primary Site of Tumor Critical for Stage, Treatment, and Prognosis
- Histology is used to identify which are abstracted as Brain/CNS
- Highly specialized surgical procedures – location of tumor
- Some Prognostic Factors Overlap

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Sinus, Olfactory, Base of Skull Tumors



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<http://www.hopkinsmedicine.org/sebin/d/w/skull-base-ian-suk-sml.jpg>

Histologic Type - Glioma

- Most common category of primary brain tumors. They begin in glial cells (supporting cells of the CNS) – can be Grade I-IV not just III-IV.
- 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.

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Glioma – 3 Main Histologic SubTypes

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 SubTypes

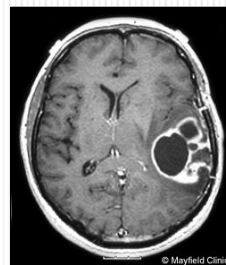
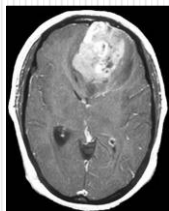
2. Oligodendroglioma: Rare tumor that usually occurs in the cerebrum, grows slowly and usually does not spread into surrounding brain tissue like astrocytoma does. Most common in middle-aged adults.
3. Ependymoma: Most commonly arise in children and young adults. They are also seen with neurofibromatosis Type II. (which we will discuss in a bit)

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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. ⁵¹ 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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Glioma Chromosome Alterations

Table 5 Common chromosomal alterations in gliomas.

Chromosomal region	Type of alteration	Candidate glioma genes
1p36.31-pter	Gains and deletions	Not known
1p36.22-p36.31	Gains and deletions	Not known
1p34.2-p36.1	Gains and deletions	Not known
1q32	Gains	<i>RIPK5</i> , <i>MDM4</i> , <i>PIK3C2B</i> and others
4q	Deletions	<i>NEK1</i> , <i>NIMA</i>
7p11.2-p12	Amplifications or gains	<i>EGFR</i>
9p21-p24	Deletions	<i>CDKN2</i>
10q23	Deletions	<i>PTEN</i>
10q25-q26	Deletions	<i>MGMT</i>
11p	Deletions	Between <i>CDKN1C</i> and <i>RRAS2</i>
12q13.3-q15	Amplifications	<i>MDM2</i> , <i>CDK4</i> and others
13p11-p13 and 13q14-q34	Loss	<i>RB1</i>
19q13	Loss	<i>GLTSCR1</i> , <i>GLTSCR2</i> , <i>LIG1</i> , <i>PSCD2</i> and many others
22q11.21-q12.2	Loss	28 genes, including <i>IN11</i>
22q13.1-q13.3	Loss	Not known

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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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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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Non-Glial Tumors

- Schwannoma: Arises from Schwann cells present in certain nerves, including those that control balance and hearing. May be called “neuroma”.
- 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.

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Roswell Park Cancer Institute

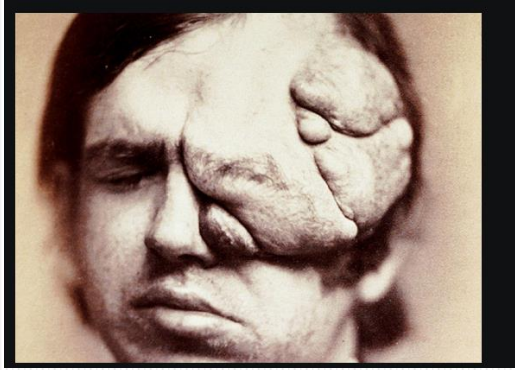
Neurofibromatosis

- 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

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Neurofibromatosis Foundation

NF Type I: First documented photo 1871



Source Credit: Dr. Stanley B. Burns

http://www.cbsnews.com/2300-204_162-10007019-6.html#ixzz1clEzAchl

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Other Manifestations of NF Type I

Lisch nodules on the eye

- Melanocytic hamartomas



Neurofibromatosis
lisch-nodules.jpg

Café-au-lait spots on skin

- Discolored birth marks



Neurofibromatosis
neurofibromatosis-1.jpg

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Medscape Source: Dermnet.com; Dermatologic Manifestations of NF Type I

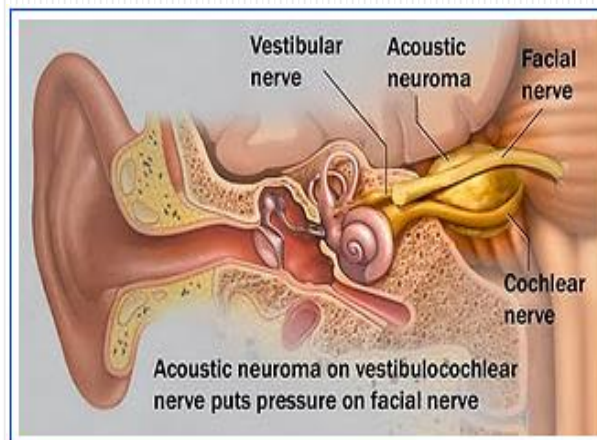
Neurofibromatosis Type II

- Neurofibromatosis Type II (NFII), also known as Multiple Inherited Schwannomas, Meningiomas and Ependymomas (MISME) or Bilateral Acoustic Neurofibromatosis (BAN).
- Is a genetically inherited disease caused by mutations of the "Merlin" gene, which appears to influence 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.

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Source: California Ear Institute

Acoustic Neuroma/Schwannoma



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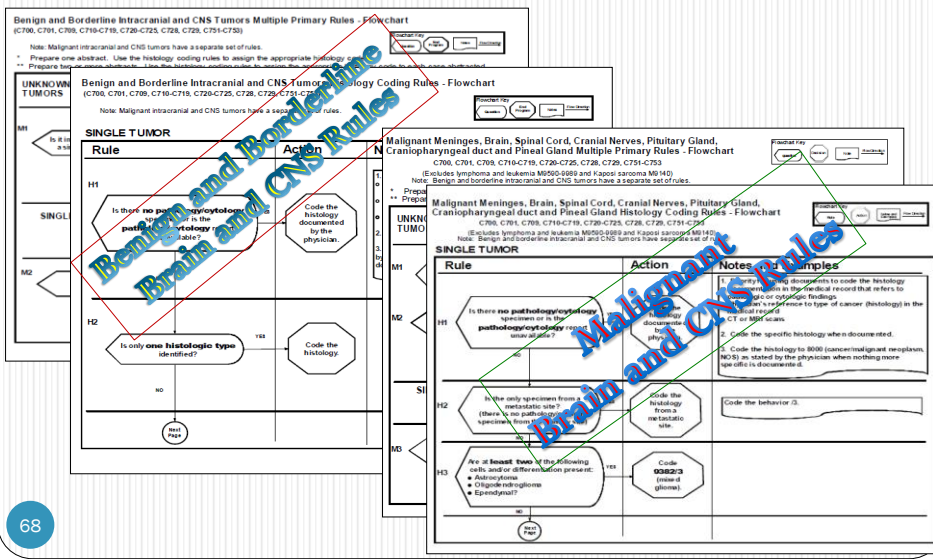
Source: <http://thrivingwithneurofibromatosis.blogspot.com>

Multiple Primary Rules Histology Coding Rules



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Different Rules for Benign and Malignant



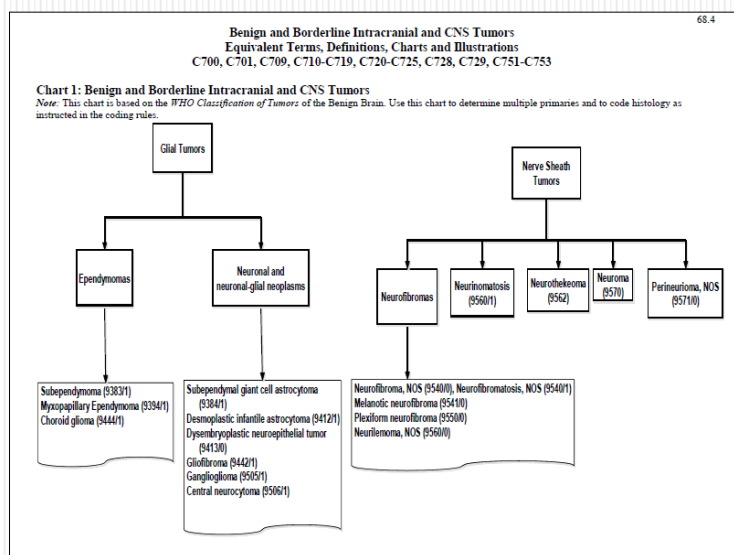
68

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.

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Benign and Borderline Tumor Rules



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Malignant Meninges, Brain, Spinal Cord, Cranial Nerves, Pituitary gland, Craniopharyngeal duct and Pineal gland
Equivalent Terms, Definitions, Charts and Illustrations
C700, C701, C709, C710-C719, C720-725, C728, C729, C751-C753
(Excludes lymphoma and leukemia – M9500-9989 and Kaposi sarcoma M9140)

Chart 1 –Neuroepithelial Malignant Brain and Central Nervous System Tumors
*Note: This chart is based on the *WHO Classification of Tumors of the brain and central nervous system*. The chart is not a complete listing of histologies that may occur in the brain or central nervous system.*

Chart Instructions: Use this chart to code histology. The tree is arranged in descending order. Each branch is a histology group, starting at the top with the least specific terms and descending into more specific terms.

```

graph TD
    NE[Neuroepithelial tumors (9300)] --> ET[Embryonal tumors]
    NE --> EMT[Ependymal tumors]
    NE --> PT[Pineal tumors]
    NE --> CPT[Choroid plexus tumors]
    NE --> NMT[Neuronal and mixed neuronal-glial tumors]
    NE --> NT[Neuroblast tumors]
    NE --> OT[Olig tumors]
    NE --> OGT[Oligodendroglial tumors]

    EMT --> EMT1[Ependymoma, NOS (9301)]
    EMT1 --> EMT2[Anaplastic ependymoma (9302)]
    EMT2 --> EMT3[Facillary ependymoma (9303)]

    PT --&retinoblastoma (9304)
    PT --> PT2[Pinealoblastoma (9305)]
    PT2 --> PT3[Choroid plexus papilloma (9306)]
    PT3 --> PT4[Choroid plexus carcinoma (9307)]

    NMT --> NMT1[Neuroblastoma, unspecified (9308)]
    NMT1 --> NMT2[Olfactory neuroblastoma (9309)]
    NMT2 --> NMT3[Olfactory neuroepithelioma (9310)]
    NMT3 --> NMT4[Neuroblastoma, malignant (9311)]

    NT --> NT1[Embryonal neuroblastoma (9312)]
    NT1 --> NT2[Embryonal neuroepithelioma (9313)]
    NT2 --> NT3[Embryonal neuroblastoma (9314)]

    OT --> OT1[Oligoma, NOS (9315)]
    OT1 --> OT2[Oligodendroglioma, NOS (9316)]
    OT2 --> OT3[Oligodendroglioma, anaplastic (9317)]
    OT3 --> OT4[Oligodendroblastoma (9318)]

    OGT --> OGT1[Oligodendroglioma, NOS (9319)]
    OGT1 --> OGT2[Oligodendroglioma, anaplastic (9320)]
    OGT2 --> OGT3[Oligodendroblastoma (9321)]

    EMT3 --> EMT3a[Atypical teratoid/rhabdoid tumor (9322)]
    EMT3 --> EMT3b[Medulloepithelioma (9323)]
    EMT3b --> EMT3b1[Medulloepithelioma, NOS (9324)]
    EMT3b1 --> EMT3b2[Teratoid medulloepithelioma (9325)]
    EMT3b2 --> EMT3b3[Teratoid medulloepithelioma (9326)]

    EMT3 --> EMT3c[Neuroepithelioma (9327)]
    EMT3c --> EMT3c1[Neuroepithelioma, NOS (9328)]
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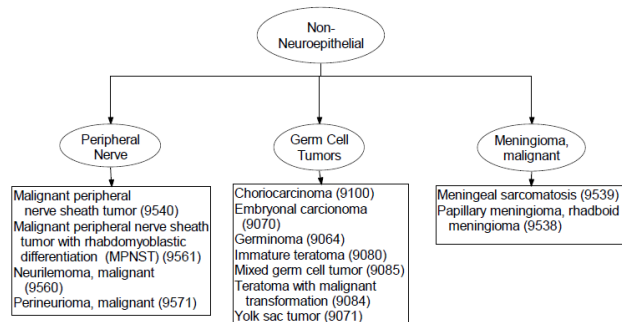
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    EMT3d61 --> EMT3d62[Supratentorial primitive neuroectodermal tumor (PNET) (9393)]
    EMT3d62 --> EMT3d63[Supratentorial primitive neuroectodermal tumor (PNET) (9394)]
    EMT3d63 --> EMT3d64[Supratentorial primitive neuroectodermal tumor (PNET) (9395)]
    EMT3d64 --> EMT3d65[Supratentorial primitive neuroectodermal tumor (PNET) (9396)]
    EMT3d65 --> EMT3d66[Supratentorial primitive neuroectodermal tumor (PNET) (9397)]
    EMT3d66 --> EMT3d67[Supratentorial primitive neuroectodermal tumor (PNET) (9398)]
    EMT3d67 --> EMT3d68[Supratentorial primitive neuroectodermal tumor (PNET) (9399)]
    EMT3d68 --> EMT3d69[Supratentorial primitive neuroectodermal tumor (PNET) (9400)]
    EMT3d69 --> E
```

Malignant Tumor Rules

Malignant Meninges, Brain, Spinal Cord, Cranial Nerves, Pituitary gland, Craniopharyngeal duct and Pineal gland
Equivalent Terms, Definitions, Charts and Illustrations
C700, C701, C709, C710-C719, C720-725, C728, C729, C751-C753
(Excludes lymphoma and leukemia – M9500-9989 and Kaposi sarcoma M9140)

Chart 2 – Non-neuroepithelial Malignant Brain and Central Nervous System Tumors

Chart Instructions: Use this chart to code histology. The tree is arranged in descending order. Each branch is a histology group, starting at the top with the least specific terms and descending into more specific terms.
Note: Chart 2 is based on the *WHO Classification of Tumors of the brain and central nervous system*. This chart is not a complete listing of histologies that may occur in the brain or central nervous system.



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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.

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Do NOT Report Benign Tumors Dx'd < 2004

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

- HOWEVER,

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

THE FIRST NEOPLASM IS NOT REPORTABLE TO FCDS EVEN AS A HISTORICAL CASE
THE NEW (SECOND) NEOPLASM SHOULD BE ASSIGNED SEQUENCE NUMBER 60

DO NOT REPORT THE BENIGN NEOPLASM IF DIAGNOSED BEFORE 1/1/2004

THIS HAS BEEN A SOURCE OF CONFUSION – PLEASE REVIEW AND SAVE

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

Staging Brain and CNS Neoplasms



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AJCC TNM, 7th edition

“Attempts at developing a TNM-based classification and staging system for tumors of the central nervous system have not been successful.” Basic TNM Concepts are not applicable to brain and CNS sites. “It continues to be the recommendation of the ASJCC CNS Tumor Task Force that a formal classification and staging system not be attempted.”

AJCC Stage = 88 (Not Applicable)

Includes: ANY Benign
or ANY Malignant Neoplasm

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AJCC TNM, 7th edition

Factors Felt to be of Prognostic and/or Clinical Significant Include:

- Tumor Histology
- Location of Tumor
- Unifocal or Multifocal
- WHO Grade of Tumor
- Patient Age at Diagnosis
- Functional Neurologic Status *KPS)
- Primary or Recurrent Tumor
- Extent of Resection
- Metastatic Spread Proliferative Fraction (Ki-67, M1 B-1)
- Gene Deletions (1p, 19q)
- MGMT Methylation

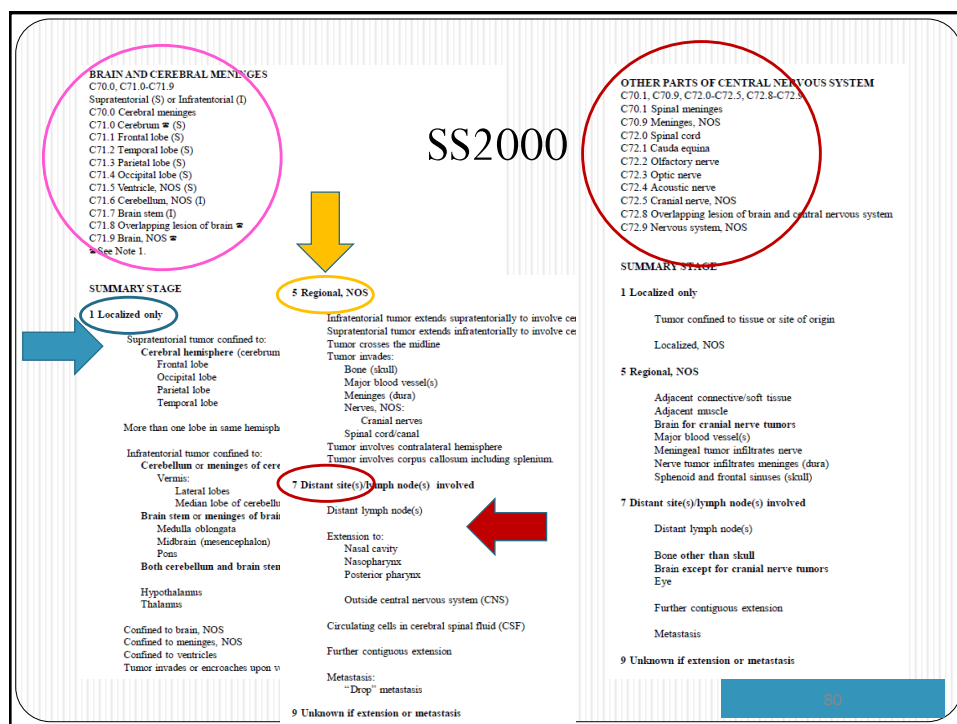
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SEER Summary Stage 2000

To obtain a FREE electronic copy of the SS2000 Manual:

<http://seer.cancer.gov/tools/ssm/SSSM2000-122012.pdf>

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Steps to Assign SS2000

Three summary stage groups can be ruled out quickly:
in situ, distant, and localized

FIRST

In situ

1. **Rule out in situ stage disease.** Carcinomas and melanomas are the only types of cancer that can be classified as in situ. Only carcinomas have a basement membrane. Sarcomas are never described as in situ. A pathologist must examine the primary organ and state that the tumor is in situ. If the cancer is anything except a carcinoma or melanoma, it cannot be in situ.
2. If there is any evidence of invasion (or extension to), nodal involvement or metastatic spread, the case is not in situ even if the pathology report so states. This is a common error in staging cervical cancer where the path report states that the cancer is "in situ with microinvasion"—such a case would be staged as localized.

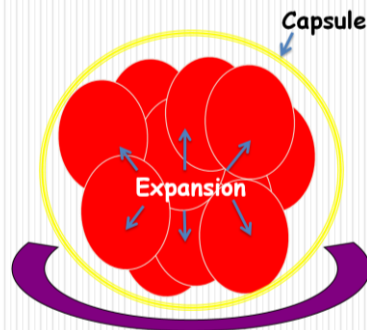
In Situ Stage is Not Applicable for Brain/CNS Tumors

In Situ Stage is Not the Same as Benign or Borderline Behavior

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SS2000 for Benign/Borderline Tumors

= 8



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Steps to Assign SS2000

Three summary stage groups can be ruled out quickly:
in situ, distant, and localized

SECOND

Distant

3. **Rule out distant disease** If metastases can be documented, there is no need to spend a great deal of time identifying local or regional spread. If distant metastases are recorded on x-ray or needle biopsy, the stage is already determined and the patient does not need to undergo a lot of other tests.
4. Hematopoietic diseases, such as leukemia and multiple myeloma, are considered disseminated or distant at time of diagnosis.
5. Rule out distant spread by reading the operative report for comments about seeding, implants, liver nodules, or other indications of metastases. Read diagnostic reports for references to distant disease.
6. If nodes, organs, or adjacent tissues are not specifically mentioned in the description of the various categories, attempt to cross-reference the term you have with those outlined. If there is no match, assume the site in question represents distant disease.

Brain and CNS these are usually CSF Involvement – cells in fluid
On rare occasion you may see “drop metastasis” – code as Distant Stage

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Steps to Assign SS2000

Three summary stage groups can be ruled out quickly:
in situ, distant, and localized

THIRD

Localized

7. **Rule out that the cancer is “confined to the organ of origin.”** In order for a lesion to be classified as localized, it must not extend beyond the outer limits of the organ and there must be no evidence of metastases anywhere else.
8. Terms such as “blood vessel invasion” or “perineural lymphatic invasion” do not necessarily indicate that the cancer has spread beyond the primary organ. If tumor at the primary site has invaded lymph or blood vessels, there is the potential for malignant cells to be transported throughout the body. Step 1 (invasion), has occurred, but not necessarily steps 2 (transport of cancer cells) and 3 (growth at the secondary site). The case may still be localized.
9. Vascular invasion within the primary is not a determining factor in changing the stage unless there is definite evidence of tumor at distant sites.

Most common Summary Stage – unless tumor crosses midline

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Steps to Assign SS2000

Three summary stage groups can be ruled out quickly:
in situ, distant, and localized
FINAL (if needed)

Regional

- 10. If in situ, local and distant categories have been ruled out, the stage is regional.
- 11. For carcinomas, if there are lymph nodes involved with the tumor, the stage is at least regional.
- 12. For tissues, structures, and lymph nodes, assume ipsilateral unless stated to be contralateral or bilateral.

Unknown if Extension or Metastasis

- 13. If there is not enough information in the record to categorize a case, it must be recorded as unstageable.

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Additional Resources

- **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 Jouvett, Bernd W. Scheithauer and Paul Kleihues, World Health Organization, Lyon, France, 2007
- **Central Brain Tumor Registry of the United States (CBTRUS)**, 2015
- **NCCN Evidence Based Treatment Guidelines**, nccn.org, 2015
- **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.
- **Multiple Primary and Histology Coding Rules**, SEER 2007
- **AJCC Cancer Staging Manual**, 7th ed., AJCC, 2010
- **SEER Summary Staging Manual 2000**

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QUESTIONS ??

