In this presentation we are going to review many of the principles for abstracting CNS tumors. Many of the slides apply to non-malignant CNS tumors only as these are the type of brain tumors where the rules differ from the normal rules. This presentation will include a discussion on assigning the CS data items for the brain. You should follow along in your manuals where applicable.

Resource: CDC NPCR Brain Tumor Training Materials 2004
Because the brain and CNS control the functions of the human body, CNS tumors are a grave concern. As shown in this picture, the brain controls thought, feeling, and function, including knowledge and memory, as well as the senses of smell, sight, hearing, and touch. Any abnormal growth in the CNS can affect a person's ability to function.
Shown here are the major areas of the CNS sites. The intracranial sites are the brain (C71.0-C71.9), cerebral meninges (C70.1), cranial nerves and other intracranial parts of the CNS (C72.0-C72.9), the craniopharyngeal duct (C75.2), and the pineal (C75.3) and pituitary (C75.1) glands. The brain is the largest intracranial organ. The pituitary and pineal glands and the cranial nerves are found inside of the brain tissue. The brain is attached to the spinal cord.

The extracranial sites are the spinal cord, cauda equina, and spinal meninges. The spinal cord is part of the CNS even though it is not intracranial. The non-malignant tumors must originate in the brain and spinal cord, not in the skull or vertebrae, which are bone, to be reportable.
The skull or cranium is bone that covers the brain. Non-malignant tumors of tissues inside of the cranium, in other words, those that are intracranial, are reportable.

The foramen magnum is the opening (foramen= hole) in the base of the skull (occipital bone) which allows the nerves of the spinal cord to enter the skull.

The foramen magnum would be a site code of C71.7. You may want to add this to your ICD-O-3 manual.
The cerebrum is the largest part of the brain and contains two hemispheres. The right hemisphere controls the left side of the body, and the left hemisphere controls the right side of the body. Each hemisphere contains four lobes; the frontal lobe, the parietal lobe, the occipital lobe, and the temporal lobe.

- The frontal lobe controls cognitive ability, memory, behavior, and the ability to speak and write. Symptoms caused by a frontal lobe tumor include seizures, impaired judgment, personality changes, and short-term memory loss.

- The parietal lobe controls sensory discrimination and body orientation. Spatial disorders, seizures, language disturbances, and the inability to do arithmetic are symptoms of a parietal lobe tumor.

- The occipital lobe controls the understanding of visual images. Symptoms of a tumor in the occipital lobe include blindness in one direction and seizures.

- The temporal lobe controls hearing and the ability to understand the spoken word, and the most common symptom of a tumor in the temporal lobe is seizure.
**Broca’s area** – Code to left frontal lobe
The patient can understand words, but words are not properly formed. Speech is slow and slurred. Some patients can only say the same word over and over.

**Wernicke’s area** – Code to left temporal lobe
Patient can speak clearly, but the word order makes no sense. The words are all mixed up and the chart may mention - “word salad”.
The cerebellum is the second largest area of the brain. It plays a role in muscle coordination, walking, and speech. Symptoms of a tumor in the cerebellum may include but are not limited to swaying, difficulty with coordination and walking, and difficulty with speech.

The brain stem is at the bottom of the brain and connects the spinal cord to the cerebrum. The pons, midbrain, medulla oblongata, and reticular formation are part of the brain stem. The brain stem controls blood pressure, heart beat, breathing, consciousness, and eating and sleeping patterns. Brain stem tumor symptoms may include but are not limited to vomiting, one-sided facial muscle weakness, swallowing difficulty, double vision, and headache just after waking.
The ventricular system is divided into four cavities called ventricles which are connected by a series of holes (called foramen) and tubes called aqueducts or canals. The ventricles contain cerebrospinal fluid that is produced by the choroid plexus, and flows through the ventricles and the subarachnoid space of the meninges.

Two ventricles enclosed in the cerebral hemispheres are called the lateral ventricles. They are called lateral rather than first and second because they are paired, but they count as two of the four ventricles. The third and fourth are unique so they each have their own numbered names. The lateral ventricles each communicate with the third ventricle through a separate opening called the Foramen of Monro. Central neurocytoma and choroid plexus papilloma are rare central nervous system tumors typically found in the lateral ventricles. The third ventricle is in the bottom center of the brain and its walls are made up of the thalamus and hypothalamus. Tumors in this location include choroid glioma. The third ventricle connects with the fourth ventricle through a long tube called the Aqueduct of Sylvius. The lateral ventricles and the third ventricle are coded to C71.5 Ventricle NOS. Most of the fourth ventricle is between the pons and cerebellum. Medulloblastomas and ependymomas commonly occur in the fourth ventricle. Tumors of the fourth ventricle are coded to C71.7 Brain stem.

Marked expansion of the ventricles, called hydrocephalus, may occur when there is a blockage in the pathways through which the fluid normally travels. It may also arise from an overproduction of fluid or a difficulty in absorbing the fluid that is produced. Because the brain is enclosed within the bony skull, this extra fluid within the ventricular system will produce increased pressure symptoms: headaches, vomiting, drowsiness and in some cases, confusion. Rare tumors involving the choroid plexus within the ventricles may affect the production and absorption of the fluid. A similar condition may arise in the spinal cord as tumors block the flow of fluid down the central canal of the spinal cord; this is known as syringomyelia.
Cranial Nerves

The human body has 12 pairs of cranial nerves. Cranial nerves 3 through 12 are found in the brain stem. 3 and 4 are in the mid-brain, 5 through 8 are in the pons, and 9 through 12 are in the medulla oblongata.

- The olfactory nerve controls sense of smell
- The optic nerve controls vision
- The oculomotor nerve controls eye movement and pupil size
- The trochlear nerve controls eye movement
- The trigeminal nerve controls sensation in the face, nose, mouth, teeth, cornea, chewing, and facial expression
- The abducens nerve controls eye muscles
- The facial nerve controls facial expression, tears, and saliva taste
- The vestibulocochlear, also known as acoustic, nerve controls hearing and balance
- The glossopharyngeal nerve controls throat movement and sensation, and taste
- The vagus nerve controls sensation and muscles in the throat and windpipe
- The accessory nerve controls movement of the neck
- The hypoglossal nerve controls tongue movement and swallowing.

Symptoms may include but are not limited to problems in the functions described above for each nerve.
The meninges are three membranes that cover the brain and spinal cord, and protect the CNS. The dura mater is the tough outer membrane. The arachnoid is the middle web-like membrane. The pia mater is the inner-most membrane and is delicate and highly vascular. The subarachnoid space is between the arachnoid and pia mater and contains cerebrospinal fluid (CSF). Symptoms of tumors of the meninges are usually caused by compression and pressure, not by growth into brain tissue. Seizures are the most common symptom of such tumors.
Meninges – PAD the brain

- This is a file from the Wikimedia Commons. The description on its description page there is shown below. Commons is a freely licensed media file repository.
The tentorium is a flap of meninges that separates the cerebral hemispheres from the posterior fossa. The posterior fossa contains the cerebellum and brain stem. Intracranial tumors are often described by their location in relation to the tentorium. Supratentorial tumors are located above the tentorium in the cerebral hemispheres and include tumors in the parietal lobe. Infratentorial tumors are located below the tentorium in the cerebellum or brain stem and include tumors in the medulla oblongata, part of the brain stem.
The spinal cord begins in the medulla oblongata, which is part of the brain stem and is made up of nerve fibers. Meninges cover and protect the spinal cord. Symptoms of spinal cord tumors depend on the nerves involved and may include pain in the chest for thoracic tumors, and pain in the neck, arm, back, or leg with lumbar or cervical tumors.
Part I will review the types of CNS tumors that are considered reportable and issues related to casefinding.
The Benign Brain Tumors Cancer Registries Amendment Act, Public Law 107-260, refers to CNS tumors as “brain-related tumors”. Brain-related tumors are defined by Public Law 107-260 as follows:

“The term 'brain-related tumor' means a listed primary tumor (whether malignant or benign) occurring in any of the following sites: (I) the brain, meninges, spinal cord, cauda equina, a cranial nerve or nerves, or any other part of the central nervous system, (II) the pituitary gland, pineal gland, or craniopharyngeal duct.

**Listed** means listed in the ICD-O.

All cancer registry standard setters have adopted this definition for reportability of brain tumors.

The COC requires that clinically and pathologically diagnosed analytic (Class of Case 00-22) non-malignant primary intracranial and CNS tumors diagnosed on or after January 1, 2004, with an ICD-O-3 behavior code of 0 or 1 be accessioned, abstracted, and followed for the following primary sites: meninges (C70._), brain (C71._), spinal cord, cranial nerves, and other parts of the CNS (C72._), pituitary gland (C75.1), craniopharyngeal duct (C75.2), and pineal gland (C75.3).
Benign and borderline brain and CNS tumors are reportable for cases diagnosed

**January 1, 2004 forward**

Lifetime follow-up is also required on these tumors.

Remember that malignant brain tumors are mentioned here because they have a behavior code of /3 and have always been reportable. Also, remember that state requirements may differ from the COC requirements and these may have been reportable prior to 2004 in certain states.
Only selected sites with a non-malignant tumor are reportable. This slide lists the sites that are reportable for non-malignant tumors.

Remember that all malignant tumors will be reportable, so this slide is focusing on the reportable non-malignant sites – since not all sites with a non-malignant tumor would be reportable. This list is the same as listed in the law.
This slide lists the specific sites for the brain that are reportable for non-malignant tumors. The ICD-O-3 codes for brain are C71.0 through C71.9. Parts of the brain are the cerebrum, frontal lobe, temporal lobe, parietal lobe, occipital lobe, and the ventricle, cerebellum, brain stem, overlapping lesions of the brain, and Brain, NOS.
### Reportable Sites - Meninges

<table>
<thead>
<tr>
<th>Site</th>
<th>ICD-O-3 Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cerebral meninges</td>
<td>C70.0</td>
</tr>
<tr>
<td>Spinal meninges</td>
<td>C70.1</td>
</tr>
<tr>
<td>Meninges, NOS</td>
<td>C70.9</td>
</tr>
<tr>
<td>Spinal cord</td>
<td>C72.0</td>
</tr>
<tr>
<td>Cauda equina</td>
<td>C72.1</td>
</tr>
</tbody>
</table>

This slide lists the meninges and other CNS sites that reportable for non-malignant tumors. These sites are included in the definition of CNS tumors. The ICD-O-3 codes for meninges are C70.0 through C70.9. The ICD-O-3 code for spinal cord is C72.0, and for cauda equina is C72.1.
Tumors of the cranial nerves are also included in the definition of CNS tumors. The codes for cranial nerves are C72.2 through C72.5.
NON-malignant tumors of the spinal nerves and peripheral nerve roots are NOT reportable.
These other sites are also included in the definition of CNS tumors. The ICD-O-3 codes for these other parts of the CNS are C72.8 and C72.9. The ICD-O-3 code for pituitary gland is C75.1, for craniopharyngeal duct is C75.2, and for pineal gland is C75.3.

Again, for the sites described tumors with non-malignant and malignant behavior are reportable for cases diagnosed on or after January 1, 2004.
The pineal gland is the size of a pea and shaped like a tiny pine cone. It grows in size until about age 2 then only increases in weight. It produces a hormone that controls biological body rhythms including the aging process and sleeping cycles.

Hormones produced in the hypothalamus carry signals or messages to the pituitary gland which sends signals to the thyroid gland, adrenal glands, ovaries and testes. The pituitary gland produces thyroid hormone, estrogen, testosterone, growth hormone and many more. These hormones effects metabolism, blood pressure, and reproduction.
Reportable Sites - Cysts

Cysts and tumor-like lesions:

Reportable:
- Dermoid cysts (M9084)
- Granular cell tumors (M9580)
- Rathke pouch tumors (M9350)

Not reportable:
- Epidermoid cysts
- Colloid cysts
- Enterogenous cysts
- Neuroglial cysts
- Plasma cell granulomas
- Nasal glial heterotopias
- Rathke cleft cysts

The NPCR law defines a “brain-related tumor” as a primary tumor listed in the ICD-O. Therefore, only tumors listed in ICD-O are reportable. The WHO lists several cysts and tumor-like conditions. Of these, only three are reportable.

Dermoid cysts or tumors are congenital ectodermal inclusion cysts. All elements composing the tumors originate from the embryonic ectoderm. These are rare tumors, accounting for less than 0.5% of all intracranial tumors. Dermoid cysts may be found intracranially or in the spinal canal, mainly in or near the midline; some typical locations are frontobasal, parasellar, or in the posterior fossa. They are cystic masses, with a fibrous capsule lined with squamous epithelium and containing a thick fluid composed of cholesterol, keratin, and lipid metabolites, derived from decomposed epithelial cells. The cysts may rupture and fatty components of their contents may spread into the subarachnoid spaces and within the ventricles.

Granular cell tumors (GCTs) are benign neoplasms composed of proliferations of round or polygonal cells that contain eosinophilic granular cytoplasm. The most common locations are tongue and subcutaneous tissue, but a variety of other sites may be involved, including the CNS. Most CNS GCTs arise in the pituitary gland, but rare cases involving brain and leptomeninges have been described.

“Rathke pouch tumor” is a synonym for craniopharyngioma.

All of the other listings are NOT reportable because they do not have an ICD-O code. Including Rathke's cleft cyst.

Some controversy exists among pathologists regarding the need to include some CNS cystic lesions in the ICD-O. A concensus conference is being planned to discuss issues pertaining to CNS ICD-O codes. Some have suggested revising to the ICD-0 in a few years.
It can be difficult to determine if the primary site is intracranial or the skull (C41.0). Tumors that originate in the skull are not intracranial.

**Chondroma** (9220/0) is a benign tumor of cartilage cells. Complete an abstract if the primary tumor is in an intracranial site. Chondroma (M9220/0) must originate in a brain-related site to be reportable. If it arises in the SKULL (bone), it is NOT reportable (not intracranial).

**Chordoma** is a malignant tumor arising from the embryonic tissue and is a malignant tumor of cartilage cells.
Unusual and ambiguous terminology used in the diagnosis of CNS tumors makes it difficult to determine if a case is reportable, and if it is reportable, to determine site and histology.

If the final pathologic diagnosis is a CNS neoplasm or mass, an ICD-O-3 code must be listed in the manual for the mass or neoplasm to be reportable. If no ICD-O-3 code is listed for the mass or neoplasm, the case should not be reported.

If the only diagnosis is “hypodense mass” or “cystic neoplasm” this is not reportable even for CNS sites because there are no ICD-O-3 codes for this terminology.

If the ONLY diagnosis available is CNS “tumor” or “neoplasm”, this is reportable and is coded 8000/1.

A benign meningioma with the site listed as "skull" should be coded to the cerebral meninges. The meninges are between the skull and the intracranial tissues. A meningioma originates in the meninges and can invade the skull.
Casefinding

- Brain tumors may only be diagnosed clinically
- Expand your casefinding
  - Radiology
  - Gamma/cyber knife center
  - Additional codes for Disease Index
  - Diagnostic imaging, MRI
  - Neurology clinics
  - Autopsy
  - Treatment facilities

Cancer registries should first examine the sources used to identify malignant CNS tumors and expand the procedures to include those needed to identify non-malignant CNS tumors.

- Since surgery is often the treatment of choice for CNS tumors of all behaviors, pathology reports are an excellent casefinding source.
- Many patients with CNS tumors of all behaviors are treated with adjuvant or primary radiation therapy and a review of radiation oncology appointment logs can identify these cases of primary CNS tumors.
- Gamma or cyber knife is becoming a common treatment for non-malignant CNS tumors. If the hospital has a gamma or cyber knife center, logs and schedules should be reviewed as part of casefinding.
- Disease indices include diagnosis codes for both inpatients and outpatients. Cases coded with a diagnosis of non-malignant or malignant neoplasm of a CNS site should be reviewed.
- As mentioned, since surgery is often the treatment of choice for CNS tumors of all behaviors, inpatient and outpatient surgery logs should also be reviewed.
- Diagnostic imaging is often the first source of diagnosis for CNS tumors and a review of imaging reports is recommended; however, because so many diagnostic imaging procedures are performed, the work involved in reviewing all imaging reports is often not worth the effort for the number of cases identified. Your cancer committee will determine if radiology reports are used for casefinding. Many of these tumors are diagnosed by MRI.
- Logs or schedules from departments or clinics for radiation oncology, neurology, and medical oncology should be reviewed. In facilities with large neurology services, many cases may be identified through the neurology clinic schedules.
- Autopsy reports should be reviewed because occasionally a non-malignant intracranial tumor is identified only at autopsy.

Send a list of the required tumors to departments, including the pathology department.
### ICD-9-CM CASEFINDING CODE LIST

Go to seer.gov for a complete list

<table>
<thead>
<tr>
<th>ICD-9-CM Code^</th>
<th>Explanation of Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>140.0 – 208.9</td>
<td>Malignant Neoplasms</td>
</tr>
<tr>
<td>209.0 – 209.3</td>
<td>Neuroendocrine tumors (Effective date: 1/1/2009)</td>
</tr>
<tr>
<td>225.0 – 225.9</td>
<td>Benign neoplasm of brain and spinal cord neoplasm</td>
</tr>
<tr>
<td>227.3 – 227.4</td>
<td>Benign neoplasm of pituitary gland, pineal body, and other intracranial endocrine-related structures</td>
</tr>
<tr>
<td>227.9</td>
<td>Benign neoplasm; endocrine gland, site unspecified</td>
</tr>
<tr>
<td>228.02</td>
<td>Hemangioma; of intracranial structures</td>
</tr>
<tr>
<td>228.1</td>
<td>Lymphangioma, any site</td>
</tr>
<tr>
<td>237.0 – 237.9</td>
<td>Neoplasm of uncertain behavior [borderline] of endocrine glands and nervous system</td>
</tr>
<tr>
<td>239.6</td>
<td>Neoplasms of unspecified nature, brain</td>
</tr>
<tr>
<td>239.7</td>
<td>Neoplasms of unspecified nature; endocrine glands and other parts of nervous system</td>
</tr>
<tr>
<td>259.8</td>
<td>Other specified endocrine disorders</td>
</tr>
<tr>
<td>V10.0 – V10.9</td>
<td>Personal history of malignancy (screen for recurrences, subsequent primaries, and/or subsequent treatment)</td>
</tr>
</tbody>
</table>

The hospital disease index is one source of casefinding utilized by cancer registrars and central registry staff. Data are stored in the index by ICD-9-CM code. The ICD-9-CM codes shown on the slide should be added to casefinding lists to identify non-malignant CNS tumors through the hospital disease index. Changes to the ICD-9-CM codes are posted each year. The SEER website is currently posting a current copy of the ICD-9-CM casefinding list for each year.
PART II

Assigning the Primary Site, Histology and Grade

Part II will review issues with assigning the primary site, histology and grade as well as introduce some of the common types of brain tumors.
The WHO classification of tumors affecting the CNS divides tumors into Neuroepithelial and Other CNS neoplasms.

Neuroepithelial means derived from neurons and glia cells of the nervous system. The neuroepithelial tumors include astrocytomas, oligodendroglioma, ependymomas, pineal parenchymal tumors, and others.

The category of “Other” includes tumors derived from non-glial cells such as sellar tumors, hematopoetic tumors, germ cell tumors, meningiomas, and tumors of cranial nerves.
Many researchers divide brain tumors into two categories of glial and non-glial. Glia (glial tissue) is the supportive tissue of the brain made up of astrocytes and oligodendrocytes which are glial cells.

Glial tumors develop from glial tissue and may be benign, borderline, or malignant. They are assigned ICD-O-3 histology codes from the glioma series, codes 938 through 948.

Some histologies of glial origin are glioma, subependymoma, astrocytoma, glioblastoma, and medulloblastoma.

CNS tumors that develop in other tissues are defined as non-glial tumors and may be benign, borderline, or malignant. Some non-glial histologies are meningioma, germ cell tumor, and pituitary adenoma.
Astrocytic tumors are divided into:  

- **Non-infiltrating**  
  - **Exceptions to the non-infiltrating rule:**  
    - Juvenile astrocytoma M9421/3  
    - Pilocytic astrocytoma M9421/3  
    - Occurs usually in children or teens  
    - Physician may refer to these as a benign tumor  
    - Must code to invasive, behavior code /3  

- **Infiltrating**  
  - Astrocytoma  
  - Glioblastoma multiforme (M9440)  
  - Brain stem gliomas (M9380)

Astrocytic tumors are divided into non-infiltrating and infiltrating categories.

Non-infiltrating tumors include **Juvenile pilocytic** and **subependymal astrocytomases** which are often curable.

In the **Infiltrating category**, **well-differentiated mildly and moderately anaplastic astrocytomases** are less often curable.

For **anaplastic astrocytomases** of higher grade, the cure rate is low with standard local treatment.

The cure rate for **glioblastoma multiforme**, is very also low with standard local treatment.

**Brain stem gliomas** have a relatively poor prognosis that is correlated with histology (when biopsies are performed), location, and extent of tumor. The overall median survival time of patients with brain stem gliomas in studies has been 44 to 74 weeks. The best results have been attained with hyperfractionated radiation therapy.

**Pilocytic astrocytoma** starts in the dura and extends to the bone. There are **many exceptions to the primary site rule**. Continue to code to behavior 3 for consistent historical data. This correction should have already been made to your ICD-O-3 manual. It was included in the errata for the ICD-O-3.
Pineal region tumors:

Parenchymal tumors
- Pineocytoma (M9361/1)
- Pineoblastoma (M9362/3)

Germ cell tumors:
- Germinoma (M9064/3) – most common
- Embryonal carcinoma (M9070/3)
- Choriocarcinoma (M9100/3) - rare
- Teratoma (M9080/0,1,or 3)

**Pineocytomas** are slow growing and carry variable prognoses for cure. **Pineoblastomas** are more rapidly growing and have a poorer prognosis. **Pineal astrocytomas** vary in prognosis depending on the degree of anaplasia. Higher grades have a poorer prognosis.

**Germ cell tumors** include germinoma, embryonal carcinoma, choriocarcinoma, and teratoma. The prognosis and treatment depends on the histology, location, presence and amount of biological markers, and surgical resectability.

**Germinomas** are the most common tumors of germ cell origin, representing approximately 0.5–2% of primary intracranial tumors. They are midline tumors and common sites include the pineal region, suprasellar cistern, and posterior third ventricle. Suprasellar germinomas may represent a primary localization or metastatic disease from pineal lesions. Synchronous occurrence in the pineal gland and suprasellar cistern is frequent.

**Embryonal cell carcinomas** and choriocarcinomas are rare, highly malignant germ cell tumors of the pineal region that are devoid of specific characteristics on imaging studies compared to other germ cell tumors. Prior to biopsy, the diagnosis may be suspected clinically on the basis of cerebral spinal fluid analysis of tumor markers. Embryonal cell carcinomas express both AFP and the beta subunit of HCG. Choriocarcinomas express the beta subunit of -HCG but not AFP.

**Teratomas** arise from multipotential cells that produce tissues consisting of a mixture of two or more embryological layers (ectoderm, mesoderm, and endoderm). They can be benign (typical, or mature or immature) or malignant (formerly called teratocarcinomas). Teratomas are the second most common pineal region tumor, accounting for 15% of pineal masses. Male predominance ranges from 2:1 to 8:1.
### Non-Glial Tumors

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>Behavior Code</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Meningioma</td>
<td>Benign or malignant</td>
<td></td>
</tr>
<tr>
<td>Choroid plexus tumors (C71_, M9390)</td>
<td>Behavior can be 0, 1, or 3</td>
<td>Part of the ventricle which makes the spinal fluid</td>
</tr>
<tr>
<td>Choroid plexus papilloma</td>
<td>Behavior can be 0, 1, or 3</td>
<td></td>
</tr>
<tr>
<td>Choroid plexus carcinoma</td>
<td>Only behavior 3</td>
<td></td>
</tr>
</tbody>
</table>

**Benign meningiomas** are usually curable when they are resectable. The prognosis for patients with **malignant meningioma** is poorer than for the more well-differentiated meningiomas because complete resections are less common and the proliferative capacity is greater.

The choroid plexus can be the site of different types of tumors, including those that are primary or secondary, and benign or malignant. The most common primary tumors are choroid plexus papilloma, choroid plexus carcinoma, and choroid plexus meningioma. Radiological diagnosis is based on location within the ventricles, mainly lateral ventricles in the trigonal region but also the fourth and, more rarely, the third ventricle. MRI shows a thin rim of cerebral spinal fluid around the tumor; sometimes, however, the tumor becomes so large that makes it is difficult to determine its primary intraventricular origin.

The choroid plexus is glandular, and **PART OF THE VENTRICLE**, which makes **THE SPINAL FLUID**. These may be primary or secondary, benign or malignant.
Craniopharyngiomas are tumors usually located in the sellar and parasellar region, deriving from squamous epithelial resting along the involuted hypophyseal Rathke’s duct. These tumors account for approximately 3–5% of all intracranial tumors and show no sex predominance. They have a bimodal age distribution; more than half occur in childhood or adolescence, with a peak incidence between 5 and 10 years of age; there is a second smaller peak in the sixth decade. These tumors are often curable.

Chordomas are most common in people in their 20s and 30s. These tumors develop from the remains of a spine-like structure that forms and then dissolves in the fetus. Although these tumors are often slow-growing, they can metastasize or recur even after treatment. They are usually treated with a combination of surgery and radiation.

Schwannomas come from the cells that form a protective sheath around the body’s nerve fibers. They are usually benign and are surgically removed when possible. One of the more common forms of this tumor affects the eighth cranial nerve, which contains nerve cells that are important for balance and hearing. Also known as "vestibular schwannomas" or "acoustic neuromas," these tumors may grow on one or both sides of the brain and are potentially curable with surgery or stereotactic radiosurgery. Acoustic neuromas account for about 7% of all skull tumors. Early symptoms may include loss of hearing, ringing in the ears, dizziness and vertigo. When the condition is detected early, doctors may order an MRI scan and conduct hearing tests, which may include a special technique to test nerve impulses as they travel to the brain. When tumors are small, they can be removed through microsurgical procedures, avoiding damage to the facial nerve. For larger tumors, extensive surgery many be needed.
Vascular tumors are rare, non-cancerous tumors that arise from the blood vessels of the brain and spinal cord. The most common vascular tumors are the hemangioblastomas, often linked in a small number of people to a genetic disorder called von Hippel-Lindau disease. Hemangioblastomas do not usually spread, and surgery can offer a cure.
Arteriovenous malformations
Vascular Tumors (AVM)

- Check with the managing physician for the type of AVM
- No ICD-O-3 code for AVM, NOS
- AVM NOS Not reportable

Examples:
- 9120/0 Hemangioma, NOS
- 9121/0 Cavernous hemangioma
- 9150/0 Hemangiopericytoma, benign
- 9150/1 Hemangiopericytoma, NOS
- 9161/1 Hemangioblastoma
Primary cerebral lymphomas are thought to arise from indigenous brain histiocytes (microglia) or from rare lymphocytes that are normally present in the meninges and around vessels. Most often, they affect immunosuppressed individuals, such as patients with AIDS, but may also develop in people with intact immune systems. Their high incidence in patients with AIDS and frequency in non immunosupressed people, have made primary cerebral lymphomas a relatively common brain tumor. The brain, especially subarachnoid space, is also a frequent site of metastasis of systemic lymphoma and leukemia. Grossly, cerebral lymphomas are single or multiple, poorly defined tumors with necrosis, similar to glioblastomas. Meningeal spread is very common, and some cerebral lymphomas arise in the subarachnoid space. Cerebral lymphomas, like their extracerebral high-grade counterparts, are highly malignant.
CNS tumors occur in both adults and children, but the histologies and location are different.

Brain tumors are classified according to histology, but tumor location and extent of spread are important factors that affect treatment and prognosis in both children and adults. Immunohistochemical analysis, cytogenetic and molecular genetic findings, and measures of mitotic activity are increasingly used in tumor diagnosis and classification.

Primary brain tumors are a diverse group of diseases that, together, constitute the most common solid tumors of childhood. CNS tumors are the most common solid tumors of childhood and occur more often in children aged 7 years and younger than in older children.
Coding Issues
Coding Issues

- If the ONLY diagnosis available is CNS tumor, or neoplasm report the case with histology 8000/1

  Example:
  Patient admitted with a tumor of the frontal lobe. No other information is available
  Code: 8000/1

- Do not code 8010 (carcinoma NOS) for brain
- A CNS tumor cannot be in situ. There is no epithelial tissue.
A patient had a left fibrous meningioma (9532/0) with invasion of the skull.

- **Benign meningiomas often penetrate the bone**

Do not abstract this as malignant because of the invasion. You must have documentation of malignancy to be invasive.

A meningioma can originate in the meninges & can invade the skull. Site code is the meninges.
Coding Issues

• **PNET** tumors – reportable brain
  
• Primitive neuroectodermal tumor (9364/3) which originate from leftover cells in the fetus

• **PPNET** Peripheral primitive neuroectodermal tumor - bone or soft tissue and coded as Ewing sarcoma
When the exact primary site for intracranial schwannoma (9560/0) is not documented in the record

Code to cranial nerves NOS (C72.5)
Grade for CNS Tumors

Sixth digit of ICD-O-3 histology code
- Describes tumor differentiation or grade.
- Is not usually specified for CNS tumors.
- Is always assigned code 9 for non-malignant CNS tumors.
- Not the same as WHO grade.

The sixth digit of the ICD-O-3 morphology code describes the histologic grade or differentiation of the tumor. The ICD-O-3 grade or differentiation is not always described by pathologists for CNS tumors. When it is not described, code 9 (not determined, not stated, or not applicable) should be assigned.

Some histologies include differentiation in the terms. When this is the case, the differentiation can be coded. Also, other terms for grade may be used such as with low grade astrocytomas (M9400/3). These should be coded according to FORDS. In this case, low grade is coded as a grade I-II or code 2.

The ICD-O-3 grade or differentiation code for non-malignant CNS tumors is always code 9.

Other grading systems used to describe CNS tumors are WHO grade, Kernohan grade, and St. Anne-Mayo grade. These grades are not the same as the ICD-O-3 grade or differentiation, and are not recorded in the sixth digit histology code data field for grade. The grade is used by the clinician to plan treatment and predict prognosis. WHO grade is recorded in the Collaborative Stage Site-specific fields for Brain.
World Health Organization’s tumor grading system

**WHO grade:**
*Code only in Collaborative Stage*
*Site-Specific Factor.*
*MUST be documented as the WHO grade.*
Part III will review issues with coding laterality and sequence number.
The brain is not a paired organ but laterality should be collected on both non-malignant and malignant CNS tumors.

Researchers, including epidemiologists, have requested the collection of laterality, because the location of certain tumors might help in determining causation. Certain investigations, such as that involving cell phone usage, would benefit from having this information routinely available. Also, non-treatment-related factors, such as location of tumor by hemisphere, can be predictive factors for cognitive outcome.
Laterality must be coded for:

- Benign
- Borderline
- AND
- Malignant tumors

Laterality will be used to determine if multiple non-malignant CNS tumors are counted as multiple primary tumors. It is NOT used to determine if multiple malignant tumors of the same CNS site are multiple primary tumors.
Sequence Numbers
Benign and Borderline ONLY

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

Codes 60–88 indicate neoplasms of non-malignant behavior (Behavior equals 0 or 1).

Try not to use 99-unknown.

If you will remember from the Cancer Registry Operations course, the rule for sequence number was that you only included it in the count if it was reportable at the time of diagnosis. For example, refractory anemia did not become reportable until 1/1/2001 with the implementation of the ICD-O-3. Therefore, if a patient was diagnosed with refractory anemia in 1995 and then colon cancer in 2010, the sequence number for the colon cancer will be 00. The refractory anemia is not counted because it was diagnosed prior to it becoming reportable in 2001.

This rule is a little different for non-malignant CNS tumors. Remember that non-malignant CNS tumors did not become reportable (according to the COC and federal law) until 1/1/2004. However, the rule for assigning the sequence number states that for non-malignant CNS tumors, you are to count ALL non-malignant CNS tumors over the lifetime of the patient, even if diagnosed prior to 2004.

The range for non-malignant tumors is 60-88. Try to avoid code 99. If no mention of a prior CNS tumor, assume there are none.
Sequence Example

A patient was admitted in 2010 with a benign meningioma.

History stated the patient had a benign left frontal tumor in 1979.

Abstract the 2010 tumor.
   Sequence number is 62- 2\textsuperscript{nd} benign tumor over the patient’s lifetime

Record in the text information about the first primary tumor.
Part IV will discuss the rules and instructions for assigning the CS data items for the brain schema.
Reading Assignments

- As each data item is being discussed, you should stop and read the information in CSv2 Part II and CSv2 Part I, Section 2 for that data item including the associated notes, codes and definitions.
- Also, review the AJCC chapter for Brain and Spinal Cord (Chapter 56)

It is important that you follow along and make notes in your manual. In addition to reading the slides and the instructor’s notes, it is important that you stop and read the related sections in your manual as not every point will be discussed in detail.

There is no TNM staging for any of these primary sites, but there is a chapter for brain and spinal cord in the seventh edition of the *AJCC Cancer Staging Manual*. 
There are 3 different CS schemas that apply to CNS tumors. Be sure that you are using the correct schema based on the primary site. Remember that lymphoma of the brain will use the lymphoma CS schema.

Each schema applies to BOTH malignant AND non-malignant tumors. There are special codes in the schema for non-malignant tumors.

We will only be discussing the schema for the brain and meninges in this presentation.
Collaborative Stage

<table>
<thead>
<tr>
<th>Coding Required</th>
<th>Coded to 8’s or 9’s</th>
</tr>
</thead>
<tbody>
<tr>
<td>• CS Tumor Size</td>
<td>• CS TS/Ext-Eval (9)</td>
</tr>
<tr>
<td>• CS Extension</td>
<td>• CS Lymph Nodes (988)</td>
</tr>
<tr>
<td>• CS Mets at Dx</td>
<td>• CS Reg Nodes Eval (9)</td>
</tr>
<tr>
<td>• SSF 1 - 8</td>
<td>• Reg LN Pos (99)</td>
</tr>
<tr>
<td></td>
<td>• Reg LN Exam (99)</td>
</tr>
<tr>
<td></td>
<td>• CS Mets Eval (9)</td>
</tr>
<tr>
<td></td>
<td>• SSF 9-25 (988)</td>
</tr>
</tbody>
</table>

There are four CS data fields that need to be coded for CNS tumors, plus 8 SSF’s. The rest are either all 8’s, all 9’s, or 988.

Note: There are only 2 SSF’s for the Intracranial Gland.

The CS Tumor Size data item follows the general instructions and will not be discussed here.
An important factor in assigning the CS Extension data item is whether the tumor is above or below the tentorium.

The tentorium is a fold of dura matter that covers the cerebellum and supports the occipital lobes of the cerebrum. Whether the tumor is supratentorial or infratentorial is important for assigning the collaborative stage CS Extension data item.
Supratentorial Sites

- Cerebrum
- Frontal, temporal, parietal, and occipital lobes
- Meninges of cerebrum
- Ventricle, NOS
- Lateral ventricle
- 3rd ventricle
- Corpus callosum - white matter that connects left and right hemispheres
- Tapetum - main body of the fibers between the corpus callosum
- Anterior cranial fossa
- Middle cranial fossa
- Suprasellar

Note 1:
- C71.0 is SUPRAtentorial.
- C71.1-C71.5 are SUPRAtentorial.
- The following subsites coded to C71.8 are SUPRAtentorial: corpus callosum, tapetum.
- The following sites coded to C71.9 are SUPRAtentorial: anterior cranial fossa, middle cranial fossa, suprasellar.
Infratentorial Sites

- Cerebral subsites
- Hypothalamus - links the nervous system to the endocrine system
- Pallium - layer of neurons in grey matter
- Thalamus - between cerebral cortex and midbrain
- Cerebellum
- Meninges of cerebellum
- Brain Stem
- 4th ventricle
- Posterior cranial fossa

C71.6-C71.7 are INFRAtentorial.

The following subsites coded to C71.0 are INFRAtentorial: hypothalamus, pallium, thalamus.
The following subsites coded to C71.9 are INFRAtentorial: posterior cranial fossa.
If the behavior code is /0 (benign) or /1 (borderline), then CS Extension must be 050, even if they invade surrounding structures. Code 050 is ALWAYS assigned to non-malignant tumors.

All of the other codes are used for malignant tumors only.
Assign code 100 for Supratentorial tumors that are confined to the:
• CEREBRAL HEMISPHERE (cerebrum) or
• MENINGES of CEREBRAL HEMISPHERE on one side:
  • Frontal lobe
  • Occipital lobe
  • Parietal lobe
  • Temporal lobe

Code 110 is for Infratentorial tumors confined to:
• CEREBELLM or
• MENINGES of CEREBELLM on one side:
  • Vermis
  • Lateral lobes
  • Median lobe of cerebellum

Code 200 is for an Infratentorial tumor involving both the cerebellum and brain stem on one side.
Code 300 is for tumors confined to the ventricles. Or, the tumor invades or encroaches upon ventricular system.
Code 400 is for tumors that cross the midline, or they involve the contralateral hemisphere. Code 400 is also for tumors that involve the corpus callosum (including splenium).

The *corpus callosum* is a wide, flat bundle of axons beneath the cortex at the longitudinal fissure. It connects the left and right cerebral hemispheres and facilitates interhemispheric communication.

Code 500 is for supratentorial tumors that extend infratentorially to involve cerebellum or brain stem.

And, Code 510 is for Infratentorial tumors that extend supratentorially to involve cerebrum (cerebral hemisphere).

To choose between codes 500 and 510, it is important to determine where the tumor started and did it grow infratentorially or supratentorially.

Code 600 is used when the tumor invades any one of the following:
- Bone (skull)
- Major blood vessel(s)
- Meninges (dura)
- Nerves, NOS
- Cranial nerves
- Spinal cord/canal
Code 700 is used where there are circulating cells in the:
- cerebral spinal fluid (CSF)
- Nasal cavity
- Nasopharynx
- Posterior pharynx
- Outside central nervous system (CNS)

Use code 750 when there is direct extension from the brain or meninges to any one of the following:
- Nasal cavity
- Nasopharynx
- Posterior pharynx

Use code 800 when there is further contiguous extension to any site not included in the lower codes.
For non-malignant tumors, CS Mets at Dx must be 00 as it is not possible for these tumors to metastasize.
The collaborative stage data fields include fields for site-specific factors which assist in validating the extent of disease at diagnosis and recording other prognostic information. SSF 1 is used to record the WHO grade. The WHO grade describes tumor aggressiveness for CNS tumors and is used by clinicians to estimate prognosis.

The codes in the data field are:
- Code 010  Grade 1
- Code 020  Grade II
- Code 030  Grade III
- Code 040  Grade IV
- Code 999  WHO grade unknown or not documented.

The WHO grade may be found on the pathology report. If it is not recorded, code 999 should be used.

WHO grade should be coded in CS Site-Specific Factor 1. WHO grade should not be coded in the sixth digit histology data field. If the differentiation is not stated (well differentiated, moderately differentiated, etc.) then assign the 6th digit as 9.
WHO Grade

**WHO Grade I:**
- Slow growing
- Benign or borderline tumors
- Patients have long-term survival

**WHO Grade II**
- Relatively slow growing
- Sometimes recur as higher grade tumors
- May be benign, borderline or malignant

**WHO Grade III**
- Malignant tumors
- Often recur as higher grade tumors

**WHO Grade IV**
- Highly malignant and aggressive

If different WHO grades are reported, use the rule “grade up” and code the highest grade (worst prognosis).

If the pathology report does NOT state a WHO grade but another diagnostic test such as an MRI does, you can use the WHO grade from the diagnostic test.
CS Site-Specific Factors 2-8

- Review the details provided in Part I, Section 2
- Review the notes provided before each SSF

As these are new SSF’s required with CSv2, there is no additional information that can be added to the instructions and information provided in the manual.
Download a copy of the book from CDC
www.cdc.gov

This guide can be a great resource when abstracting CNS tumors. Remember that this guide was written before CSv2 and the MP/H rules. You should use the current coding and staging manuals for assigning data items. However, there are many other areas of information provided in this guide that can be informative, including a glossary of terms and several anatomy pictures.
This concludes this presentation.

PLEASE RETURN TO THE COURSE CONTENT.