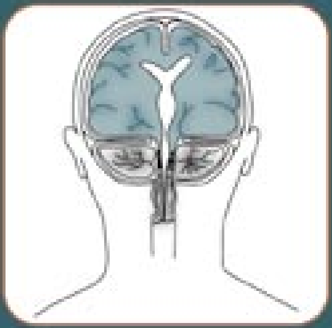
CEREBRUM
MICROSURGERY
RADIATION THERAPY
FRONTAL LOBE
FOURTH VENTRICLE
CEREBRAL AQUEDUCT
FORAMEN MAGNUM
ACOUSTIC NERVE
SKULL
OCCIPITAL LOBE
PATHOLOGY REPORTS
LATERAL VENTRICLES
CEREBELLUM
BRAIN STEM
SPINAL CORD
CAUDA EQUINA
PONS
THIRD VENTRICLE
OLFACTORY NERVE
FORAMEN OF MONRO
TEMPORAL LOBE
PITUITARY GLAND
CRANIOPHARYNGEAL DUCT
PINEAL GLAND
PARIETAL LOBE
IMAGING REPORTS
BENIGN NEOPLASM
SPINAL MENINGIOMA
CRANIAL NERVES
NEUROFIBROMATOSIS
RADIATION ONCOLOGY
AUTOPSY REPORTS
DISEASE INDEXES
BENIGN
MEDULLA OBLONGATA
CENTRAL CANAL
MIDBRAIN
MALIGNANT

CDC
Cancer

Data Collection of

PRIMARY CENTRAL NERVOUS SYSTEM TUMORS





National Program of Cancer Registries Training Materials

2004

U.S. DEPARTMENT OF HEALTH AND HUMAN SERVICES
CENTERS FOR DISEASE CONTROL AND PREVENTION
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This technical training guide is for health professionals who report data on brain and central nervous system tumors (malignant, benign, and borderline). Portions of this guide are based on data collection rules adopted by the North American Association of Central Cancer Registries Uniform Data Standards Committee on June 23, 2003. A PowerPoint version of this information is also available at http://www.cdc.gov/cancer/npcr/training/ppt.htm. The Centers for Disease Control and Prevention published this guide in collaboration with the following partners:

National Cancer Institute
Surveillance, Epidemiology, and End Results Program

North American Association of Central Cancer Registries

National Coordinating Council for Cancer Surveillance
Brain Tumor Working Group

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Letter from the Chief of CDC's Cancer Surveillance Branch

Dear Colleagues,

I am pleased to present you with this training guide, *Data Collection of Primary Central Nervous System Tumors*, from the CDC's National Program of Cancer Registries (NPCR). Our goal is to help you collect consistent, complete, and timely data on central nervous system (CNS) tumors, both malignant and nonmalignant. Such data are critical to our nation's efforts to improve treatment and quality of life for people with central nervous system tumors. Researchers, clinicians, and others rely on the data you collect to study risk factors linked with CNS tumors, identify screening efforts that can be improved, monitor tumor treatments, and learn more about survivorship for people with CNS tumors.

Over the past decade, the NPCR has strengthened not only tumor surveillance but also the reporting of many types of cancers in the United States. New registries have been established, many registries have been enhanced, and the quality of data has improved. While this represents excellent progress, many challenges remain. We depend on you, our partners, to help us further strengthen tumor surveillance in this country. By working together, we can achieve our goal of collecting complete, high-quality data in all states and make a difference in the lives of thousands of people with CNS tumors.

Thank you for the excellent work you do, and I look forward to seeing continued progress toward complete, high-quality data.

Sincerely,

Phyllis A. Wingo, Ph.D., M.S.
Chief, Cancer Surveillance Branch
Division of Cancer Prevention and Control
National Center for Chronic Disease Prevention and Health Promotion
Centers for Disease Control and Prevention

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National Program of Cancer Registries Training Materials

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Part I
Introduction

Brain and other central nervous system (CNS) tumors affect people of all ages. These tumors can run in families, or they can result from developmental abnormalities. In the United States, many cancer registries have collected data on all brain and CNS tumors—not just those that are malignant, but also benign and borderline tumors. In January 2004, all registries were required to collect data on both malignant and nonmalignant CNS tumors. Some people might ask: Why do we need to collect data on nonmalignant CNS tumors? The history that follows conveys why such surveillance is needed and how the various organizations that set data standards have worked together to promote consistency in training the health professionals who report brain tumor data (see Appendices A–E for detailed background materials).

As reported in the Brain Tumor Working Group's (BTWG) 1998 report,[1] more than 28,000 new cases of primary malignant and benign brain tumors were diagnosed nationwide in 1995. Approximately 12,000 people died of invasive brain tumors, and 947 died of benign brain tumors during that year. Another 131 deaths were due to tumors of uncertain behavior, and 2,788 deaths were due to tumors of unspecified behavior reported for those sites.

History

In the early 1900s, neurosurgeon Harvey Cushing observed that some brain tumors were malignant because of their histology, and some were malignant because of their location. By this he meant that in the early 1900s, some tumors were not resectable because of their location and would result in the death of the patient because of mass effects on vital areas of the brain.

Over the past 100 years—with advances in microsurgery, radiation therapy, and earlier diagnosis—Dr. Cushing's maxim still stands, although to a greatly diminished extent. Brain tumors, whether benign or malignant, produce clinical effects that are quite similar in terms of mass effect, hemorrhage, seizure activity, and edema. Although benign brain tumors are rare, patients with these tumors bear an underappreciated financial and health burden. Benign brain tumors can rupture and cause serious trauma to the brain. In rare cases (for example, optic nerve gliomas), the nonmalignant tumor transforms into a malignant tumor.

Before January 2004, central cancer registries and hospital cancer programs* were required to collect data on only malignant CNS tumors, in accordance with the data collection standards set by the National Program of Cancer Registries (NPCR), National Cancer Institute's (NCI) Surveillance, Epidemiology, and End Results (SEER) Program, and the Commission on Cancer (CoC) of the American College of Surgeons.

The clinicians treating patients with CNS tumors and the researchers studying these tumors felt that it was just as important to study nonmalignant tumors as it was to study malignant tumors. Clinicians and researchers urged the cancer surveillance community to include nonmalignant CNS tumors in cancer registries so that the data would be available for research.

In July 1992, the Central Brain Tumor Registry of the United States (CBTRUS) was established. Its mission was to report population-based incidence data on all primary CNS tumors, regardless of tumor behavior. At that time, 15 state cancer registries collected data on benign, borderline, and malignant primary CNS tumors, and many shared aggregate data with CBTRUS. The need for national population-based incidence data on all CNS tumors was presented to the National Coordinating Council on Cancer Surveillance (NCCCS), which is composed of stakeholders in the cancer surveillance community and provides an arena for identifying issues related to the collection and use of cancer data.

In response, the NCCCS formed the BTWG in 1996. The group was charged with examining all issues related to cancer registries' collection of data on nonmalignant CNS tumors. In September 1998, the BTWG forwarded a report with four recommendations to NCCCS:†

- **Recommendation 1.** Registries use the following standard definition in order to collect precise data on all primary intracranial and CNS tumors: Primary intracranial and CNS tumors are all primary tumors occurring in the following sites, irrespective of histologic type or behavior—brain, meninges, spinal cord, cauda equina, cranial nerves and other parts of the CNS, pituitary gland, pineal gland, and craniopharyngeal duct.

- **Recommendation 2.** A standard site and histology definition is developed for tabulating estimates of CNS tumors to allow comparability of data across

* Hospital programs approved by the Commission on Cancer of the American College of Surgeons.
† The recommendations in their entirety are found in the BTWG report to the NCCCS (Appendix E).

registries. Pathologists, the North American Association of Central Cancer Registries (NAACCR), the CoC, the SEER Program, the NPCR, and the International Agency for Research on Cancer need to be involved in developing this standard definition.

- **Recommendation 3.** All registries—hospital- and population-based—collect data on CNS tumors. This effort will necessitate a change in the CoC requirements and will increase costs to the hospital-based programs. Federal funding should be allocated to supplement the additional transition and ongoing data collection costs that will be incurred by central registries. Before additional data collection is implemented, a pilot study should be conducted in multiple states to assess the procedures and quality control functions needed as well as the costs of collecting data on these tumors.

- **Recommendation 4.** The appropriate government and professional organizations develop and implement special training programs and curricula for the central registry, hospital registries, and laboratory personnel, and develop computerized edit-checking procedures. Training for reporting and tabulating primary intracranial and CNS tumors should be offered on a regular basis.

In November 2000, at the annual meeting of the Society for Neuro-Oncology, a consensus conference was convened. Conference attendees agreed with the site definition in Recommendation 1 of the BTWG report. They also agreed to the development of a standard site and histology definition based on the SEER site and histology validation list. They discussed differences in definition of CNS tumors between the surveillance community and the neuropathology clinical community. For example, the clinicians wanted lymphomas of the brain included with brain tumors. Brain lymphomas have always been collected by registries, but the incidence is tabulated with site category lymphoma, not brain. All attendees recognized the importance of continuing the dialogue between the clinical community and the surveillance community.

In 2001, the NCCCS met and accepted Recommendations 1 and 2 as being completed. They reconvened the BTWG and asked members to work on Recommendations 3 and 4.

In January 2003, at the request of the BTWG, the NAACCR established a Benign Brain Tumor Subcommittee of its Registry Operations Committee. Members of the subcommittee were asked to develop procedure guidelines for registry operations to follow when including data on nonmalignant CNS tumors in data collection efforts.

Concurrently, the North American Brain Tumor Coalition and brain tumor activists brought the issue of collecting data on benign brain tumors to Congress. Representative Barbara Lee (D-California) introduced HR 239, the Benign Brain Tumor Bill, "to amend the Public Health Service Act to provide for the collection of data on benign brain-related tumors through the National Program of Cancer Registries." The Centers for Disease Control and Prevention's (CDC) legislative staff worked with Representative Lee and the brain tumor community to ensure that the bill included the definition for primary intracranial and CNS tumors, as given in Recommendation 1 from the BTWG. A Senate bill was drafted by Senator Jack Reed (D-Rhode Island).

In October 2002, President Bush signed Public Law 107-260, the Benign Brain Tumor Cancer Registries Amendment Act (Appendix B).

Definitions of Reportable Cases

The Benign Brain Tumor Cancer Registries Amendment Act refers to CNS tumors as "brain-related tumors." The law defines these tumors 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 *International Classification of Diseases for Oncology (ICD-O-3)* (Appendix D).[2]

All cancer registry standard setters have adopted this definition for reporting brain tumors. The CoC requires that clinically and pathologically diagnosed analytic (Class of Case 0-2) nonmalignant 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 primary sites listed in Table 1. Both SEER and CoC agreed to require reporting of nonmalignant brain tumors, beginning with cases diagnosed on or after January 1, 2004.

Anticipated Impact on Registries

Registries that begin reporting data on nonmalignant CNS tumors can expect their overall number of CNS cases each year to double, according to estimates from the BTWG (Appendix E).[1] So if a registry has an average of 50 malignant CNS tumors annually, the estimated caseload increase from adding nonmalignant CNS tumors would be 50 cases annually, for a total of 100 CNS cases a year. The new

Table 1. ICD Site and Case-Finding Codes

ICD-O-3

Code	Site
	Meninges
C70.0	Cerebral meninges
C70.1	Spinal meninges
C70.9	Meninges, not otherwise specified
	Brain
C71.0	Cerebrum
C71.1	Frontal lobe
C71.2	Temporal lobe
C71.3	Parietal lobe
C71.4	Occipital lobe
C71.5	Ventricle
C71.6	Cerebellum
C71.7	Brain stem
C71.8	Overlapping lesions of the brain
C71.9	Brain, not otherwise specified
	Spinal cord, cranial nerves, and other parts of the CNS
C72.0	Spinal cord
C72.1	Cauda equina
C72.2	Olfactory nerve
C72.3	Optic nerve
C72.4	Acoustic nerve
C72.5	Cranial nerve, not otherwise specified
C72.8, C72.9	Other parts of the CNS
C75.1	Pituitary gland
C75.2	Craniopharyngeal duct
C75.3	Pineal gland

ICD-9-CM

Code	Site
	Tumor
225.0	Benign neoplasm of brain
225.1	Benign neoplasm of cranial nerves
225.2	Benign neoplasm of cerebral meninges; meninges, not otherwise specified; cerebral meningioma
225.3	Benign neoplasm of spinal cord, cauda equina
225.4	Benign neoplasm of spinal meninges; spinal meningioma
225.8	Benign neoplasm of other specified sites of nervous system
225.9	Benign neoplasm of nervous system, part unspecified
227.3	Benign neoplasm of pituitary, craniopharyngeal duct, craniobuccal pouch, hypophysis, Rathke's pouch, sella turcica
227.4	Benign neoplasm of pineal gland, pineal body
237.0	Neoplasm of uncertain behavior of pituitary gland and craniopharyngeal duct
237.1	Neoplasm of uncertain behavior of pineal gland
237.5	Neoplasm of uncertain behavior of brain and spinal cord
237.6	Neoplasm of uncertain behavior of meninges: not otherwise specified, cerebral, spinal
237.7	Neurofibromatosis, unspecified von Recklinghausen's disease
237.71	Neurofibromatosis, type one von Recklinghausen's disease
237.72	Neurofibromatosis, type two von Recklinghausen's disease
237.9	Neoplasm of uncertain behavior of other and unspecified parts of nervous system; cranial nerves

ICD-10

Code	Site
D32	Benign neoplasm of meninges
D32.0	Cerebral meninges
D32.1	Spinal meninges
D32.9	Meninges, unspecified
D33	Benign neoplasm of brain and other parts of the central nervous system
D33.0	Brain, supratentorial
D33.1	Brain, infratentorial
D33.2	Brain, unspecified
D33.3	Cranial nerves
D33.4	Spinal cord

continued on pg. 8

Table 1. ICD Site and Case-Finding Codes (continued)

***ICD-10* (continued)**

Code	Site
D33.7	Other specified parts of central nervous system
D33.9	Central nervous system, part unspecified
D35	Benign neoplasm of other and unspecified endocrine glands
D35.2	Pituitary gland
D35.3	Craniopharyngeal duct
D35.4	Pineal gland
D42	Neoplasm of uncertain or unknown behavior of meninges
D42.0	Cerebral meninges
D42.1	Spinal meninges
D42.9	Meninges, unspecified
D43	Neoplasm of uncertain or unknown behavior of brain and central nervous system
D43.0	Brain, supratentorial
D43.1	Brain, infratentorial
D43.2	Brain, unspecified
D43.3	Cranial nerves
D43.4	Spinal cord
D43.7	Other parts of central nervous system
D43.9	Central nervous system, unspecified
D44	Neoplasm of uncertain or unknown behavior of endocrine glands
D44.3	Pituitary gland
D44.4	Craniopharyngeal duct
D44.5	Pineal gland
Q85.1	Neurofibromatosis (non-malignant); Von Recklinghausen disease

requirement usually equates to a 1% total increased caseload for cancer registries. For hospital programs with few malignant CNS cases, the estimated caseload increase will be minimal. For hospital programs with a large neurology service, the caseload increase could be greater.

Hospital registries that need additional financial support to collect the new data might be able to acquire such support from clinicians who are interested in using information on benign and malignant CNS tumors.

For central cancer registries, adding nonmalignant CNS tumors will probably be the same as for hospitals—about 1% of the total annual caseload. For the 21 state central cancer registries that already collected data on all CNS tumors as of 2002, the new requirements should have a minimal effect, because nonmalignant cases are already part of the caseload.

If the central cancer registry's definition for CNS sites is not the same as the definition in the public law, the definition must be changed. The central registries that do not currently collect data on nonmalignant CNS tumors must make sure that their state reporting laws allow them to include these cases. If not, the state reporting laws will need to be changed.

Case-Finding Sources

Cancer registries should first examine the sources used to identify malignant CNS tumors and expand their procedures to include those sources needed to identify nonmalignant CNS tumors. Here are some of the many sources where reportable cases of CNS tumors can be found:

- **Pathology reports.** Because surgery is often the treatment of choice for CNS tumors of all behaviors, pathology reports are an excellent case-finding source. Inpatient and outpatient surgery logs should also be reviewed.

- **Radiation oncology appointment logs.** Many patients with CNS tumors of all behaviors are treated with adjuvant or primary radiation therapy. A review of radiation oncology appointment logs can identify these cases of primary CNS tumors.

- **Hospital and clinic appointment logs.** Logs or schedules from hospital departments or clinics for radiation oncology, neurology, and medical oncology should be reviewed. In facilities with large neurology services, many cases can be identified through the neurology clinic schedules.

- **Gamma or cyber knife center appointment logs.** Gamma or cyber knife is becoming a common treatment for nonmalignant CNS tumors. If the hospital has a gamma or cyber knife center, logs and schedules should be reviewed as part of case-finding.

Percentage of Tumors That Are Nonmalignant

- An estimated 46% of the primary CNS tumors reported to the Central Brain Tumor Registry of the United States were nonmalignant (1990–1993 data).

- About 51% of the primary CNS tumors reported to the Minnesota Cancer Surveillance System were nonmalignant (1989–1994 data).

- More than 33% of primary CNS tumors reported to the National Cancer Database were nonmalignant (1989–1994 data; see the BTWG report in Appendix E).[1]

- **Disease indexes.** The hospital disease index is a good data source for both hospital cancer registrars and central registry staff. Data are stored in the index by codes specified in the *International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM)*.[3] U.S. hospitals continue to assign *ICD-9-CM* codes to final diagnoses. Disease indexes include diagnosis codes for both inpatients and outpatients. Cases coded with a diagnosis of nonmalignant or malignant neoplasm of a CNS site should be reviewed. The *ICD-9-CM* codes in Table 1 should be added to case-finding lists to identify nonmalignant CNS tumors through the hospital disease index.

- **Imaging reports.** Diagnostic imaging is often the first source of diagnosis for CNS tumors. Therefore, 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.

- **Autopsy reports.** Autopsy reports should be reviewed because occasionally a nonmalignant intracranial tumor is identified only at autopsy.

- **Logs from free-standing centers.** Central cancer registries have additional case-finding sources, including freestanding radiation therapy, magnetic resonance imaging (MRI), oncology, and gamma or cyber knife centers that are not associated with a hospital. Central cancer registries should consult with licensing boards in their state or region or identify other sources to ensure that they have identified all facilities using nuclear sources for treatment. In addition, central cancer registries can identify residents with tumors diagnosed in other states by establishing case-sharing agreements with other central cancer registries. Central registries can also identify CNS tumor cases by exchanging data with other central registries and through the death clearance process. Death certificate diagnoses are coded using the *International Statistical Classification of Diseases and Related Health Problems, Tenth Revision (ICD-10)*.[4] A list of *ICD-10* codes used for case identification is included in Table 1.

Unusual Terminology

Unusual and ambiguous terminology used in the diagnosis of CNS tumors can make it difficult to determine if a case is reportable, and if it is reportable, to determine the tumor's site and histology. Here are several guidelines to help you:

- If the final pathologic diagnosis is a CNS neoplasm or mass, an *ICD-O-3* code must be assigned for the mass or neoplasm to be reportable. If no *ICD-O-3* code is assigned 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 should be 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.

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Part II
Anatomy and Function

National Program of Cancer Registries Training Materials

2004

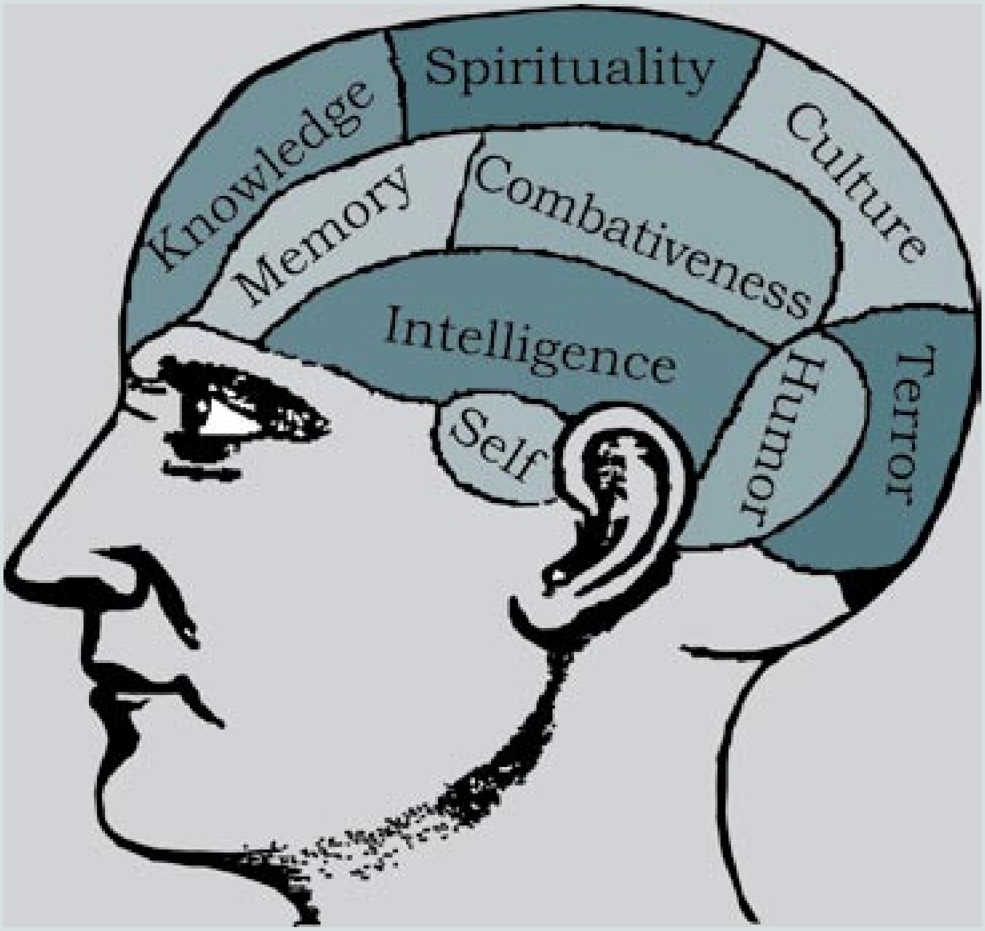


Figure 1. Functional anatomy of the central nervous system

Part II
Anatomy and Function

Because the brain and central nervous system (CNS) control the functions of the human body, CNS tumors are a grave concern. The brain controls thought, feeling, and function, including knowledge and memory, as well as the senses of smell, sight, hearing, taste, and touch (Figure 1). Thus, any abnormal growth in the CNS can affect a person's ability to function.

The skull or cranium is bone that covers the brain. The CNS includes both intracranial sites (inside the cranium) and extracranial sites (outside the cranium) (Figure 2). Nonmalignant tumors of tissues inside the cranium are reportable (Figure 3).

The brain is the largest intracranial organ. The pituitary and pineal glands and the cranial nerves are found inside the brain tissue. The brain is attached to the spinal cord.

The spinal cord is part of the CNS even though it is not intracranial. To be reportable, nonmalignant tumors must originate in the brain and spinal cord, not in the skull or vertebrae, which are bone.

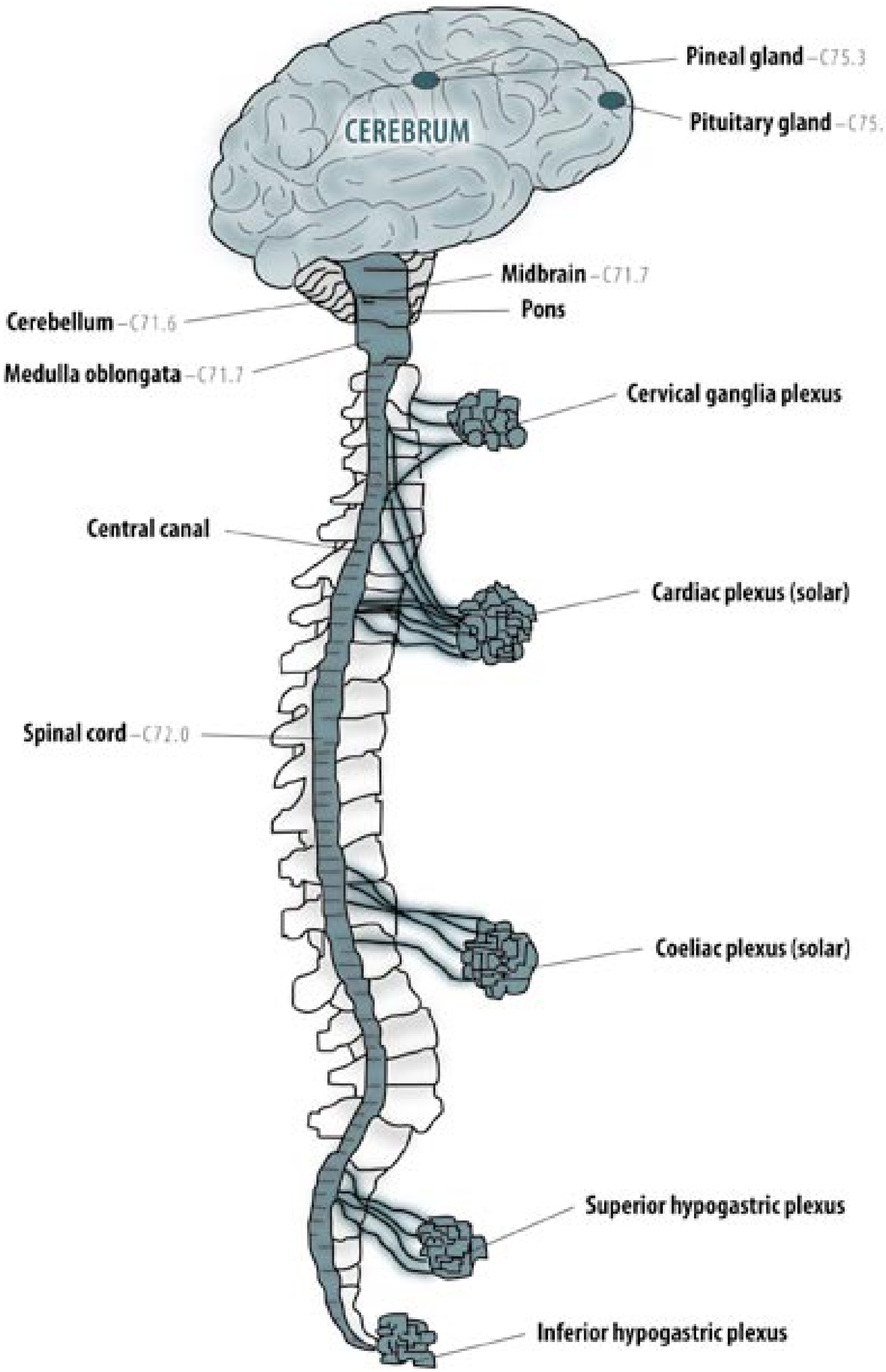


Figure 2. Anatomy of the central nervous system

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

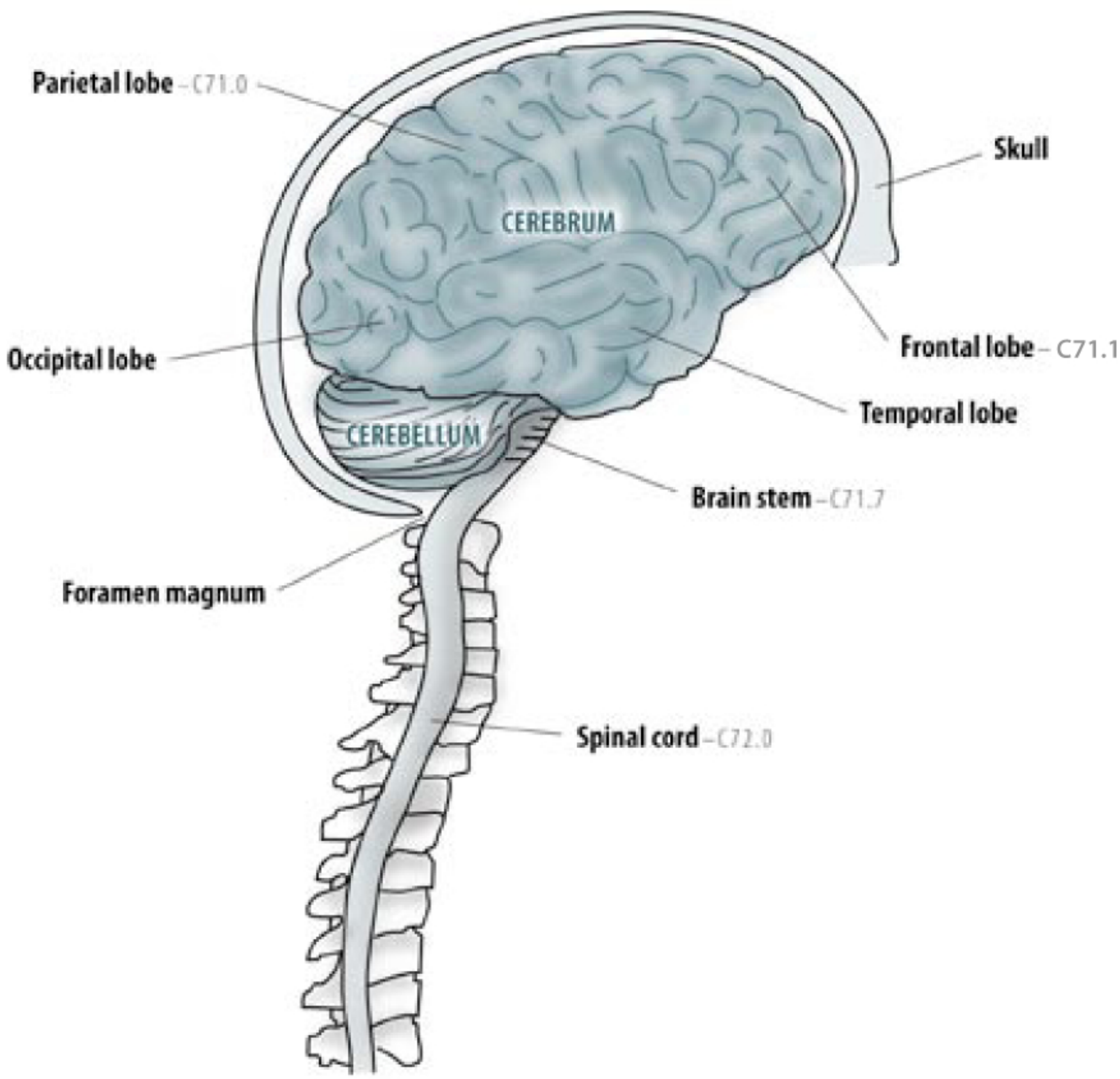


Figure 3. Intracranial sites

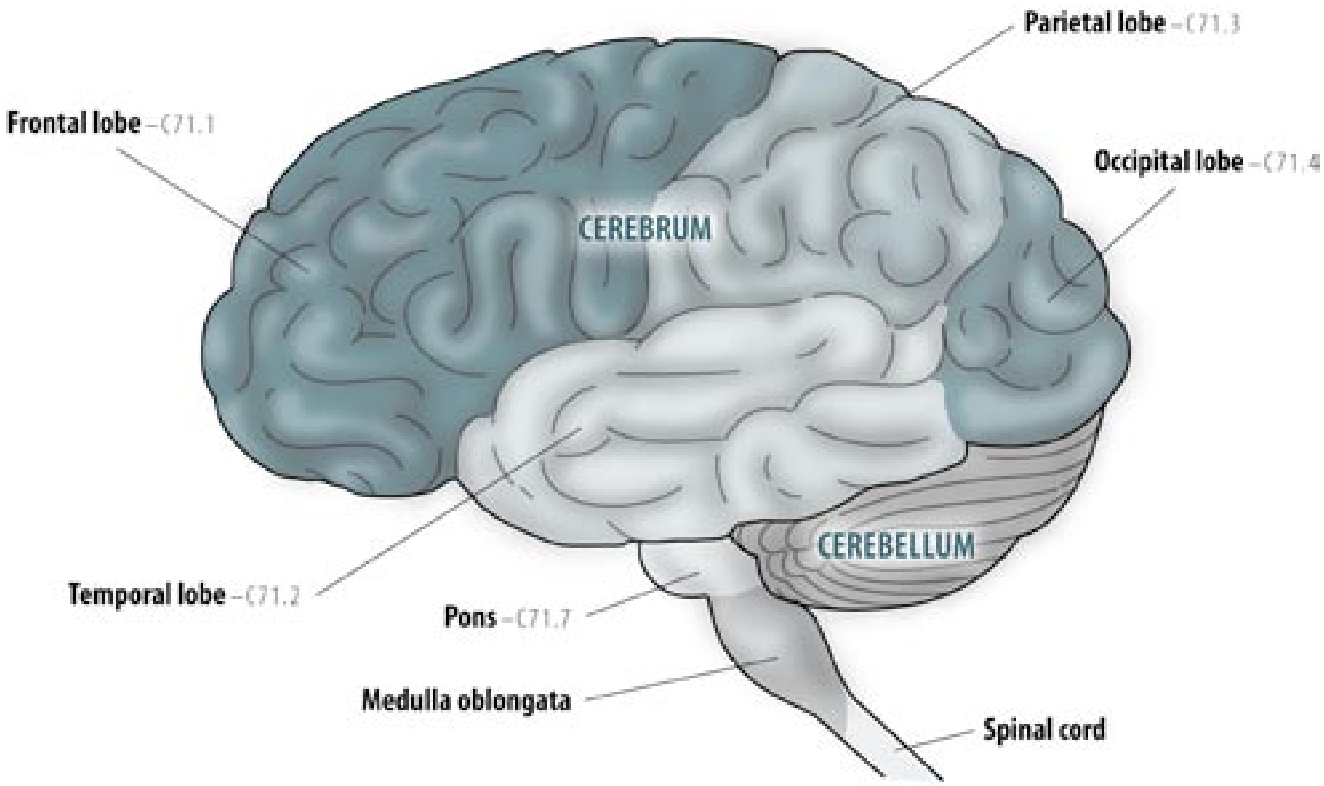


Figure 4. Cerebrum

Cerebrum

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 the following four lobes (Figure 4):

- The **frontal lobe** controls cognitive ability, memory, behavior, and the ability to speak and write. Symptoms of 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 a person's understanding of visual images. Symptoms of a tumor in the occipital lobe include blindness in one direction and seizures.

- The **temporal lobe** controls a person's hearing and ability to understand the spoken word. Seizure is the most common symptom of a tumor in the temporal lobe.

Cerebellum and Brain Stem

The cerebellum is the second largest area of the brain (Figure 5). It plays a role in muscle coordination, walking, and speech. Symptoms of a tumor in the cerebellum can include 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:

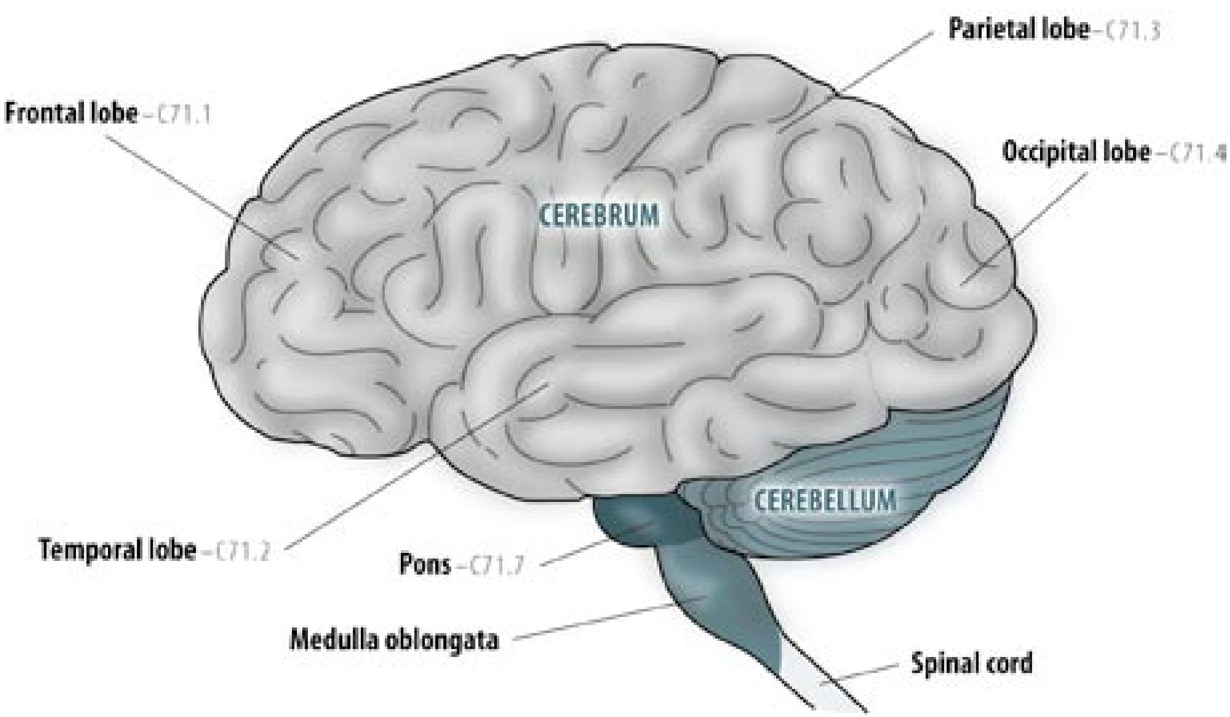
- The **medulla oblongata** functions primarily as a relay station for the crossing of motor tracts between the spinal cord and the brain. It also contains the respiratory, vasomotor, and cardiac centers as well as many mechanisms for controlling reflex activities such as coughing, gagging, swallowing, and vomiting.

- The **midbrain** serves as the nerve pathway of the cerebral hemispheres and contains auditory and visual reflex centers.

- The **pons** is a bridge-like structure that links different parts of the brain and serves as a relay station from the medulla to the higher cortical structures of the brain. It contains the respiratory center.

The brain stem controls blood pressure, heartbeat, breathing, consciousness, and eating and sleeping patterns. Symptoms of a brain stem tumor can include vomiting, muscle weakness on one side of the face, difficulty swallowing, double vision, and headache just after waking.

Figure 5. Cerebellum and brain stem



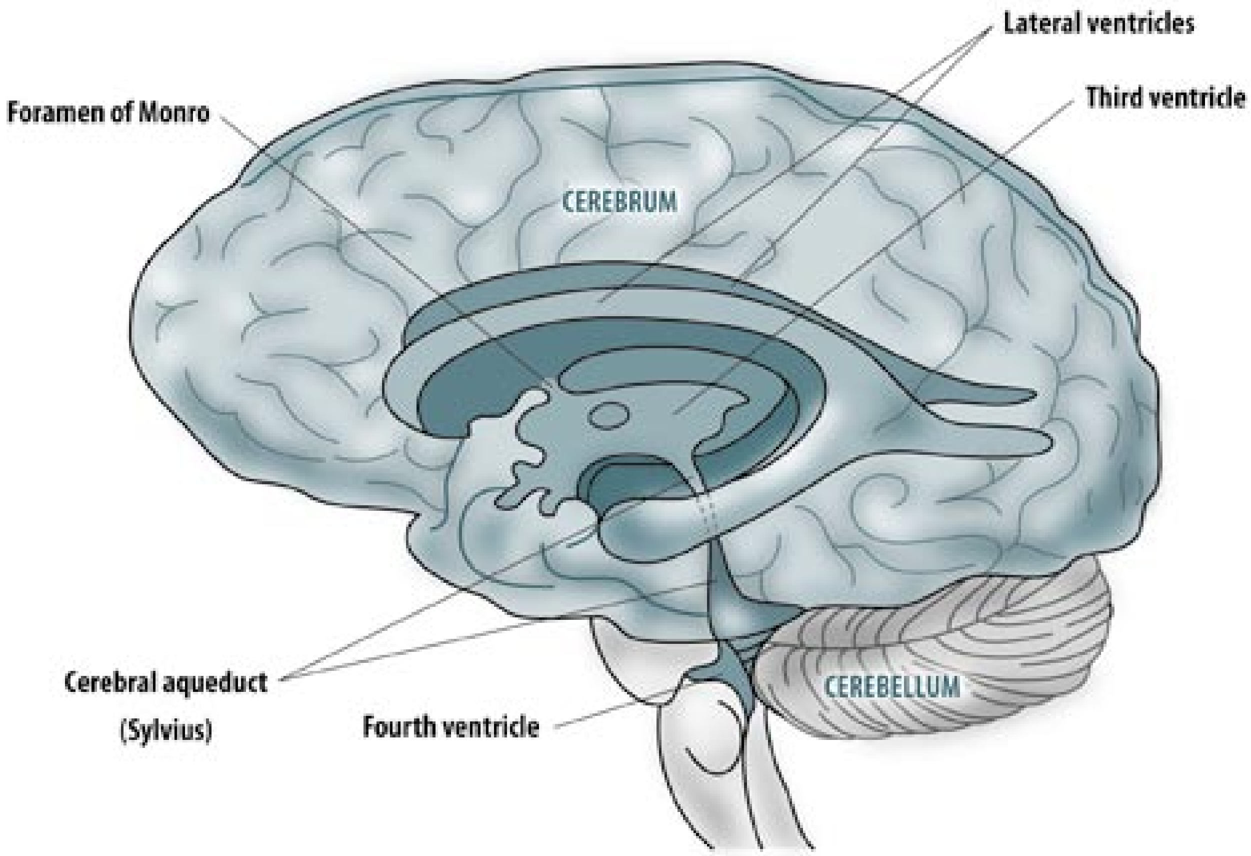


Figure 6. Ventricular system

Ventricular System

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) (Figure 6). The choroid plexus produces cerebrospinal fluid (CSF) that flows through these ventricles and the subarachnoid space of the meninges.

Here are more details about the four ventricles that make up this system:

- **Lateral ventricles.** The two ventricles enclosed in the cerebral hemispheres are called the lateral ventricles. They are called lateral rather than first and second ventricles because they are paired, but they count as two of the four ventricles. 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 lateral ventricles are assigned the *ICD-O-3* code of C71.5 (ventricle, not otherwise specified).

- **Third and fourth ventricles.** Unlike the lateral ventricles, the third and fourth ventricles are unique, so they each have their own numbered names. The third ventricle is in the bottom center of the brain, and its walls are made up of the thalamus and hypothalamus. The third ventricle connects with the fourth ventricle through a long tube called the aqueduct of Sylvius. Tumors in the third ventricle are rare and include choroid gliomas. Tumors in this location are assigned the *ICD-O-3* code of C71.5 (ventricle, not otherwise specified).

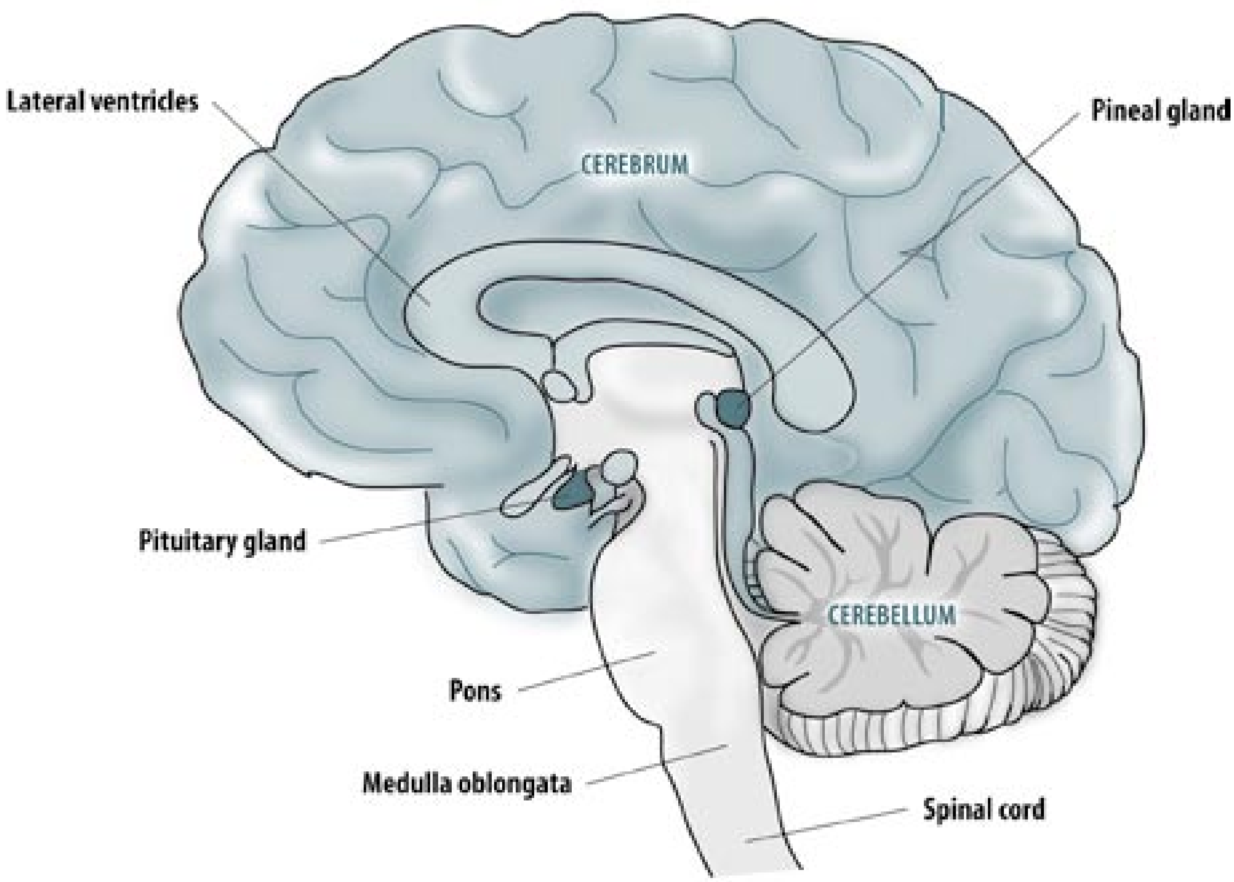
Most of the fourth ventricle is between the pons and cerebellum. Medulloblastomas and epedymomas commonly occur in the fourth ventricle. Tumors of the fourth ventricle are assigned the *ICD-O-3* code of C71.7 (brain stem).

When there is blockage of the normal flow of CSF, the ventricles can expand markedly. This condition (called hydrocephalus) can also arise when there is an overproduction of fluid or 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, including headaches, vomiting, drowsiness, and in some cases confusion. CNS tumor growth can cause blockage of CSF, and rare tumors involving the choroid plexus within the ventricles can affect the production and absorption of CSF. A similar condition (called syringomyelia) can arise in the spinal cord when a tumor blocks the flow of fluid down the central canal of the spinal cord.

Pineal and Pituitary Glands

The pineal and pituitary glands are located deep inside the brain (Figure 7). The pineal gland produces melatonin, the hormone that controls biological body

Figure 7. Pineal and pituitary glands



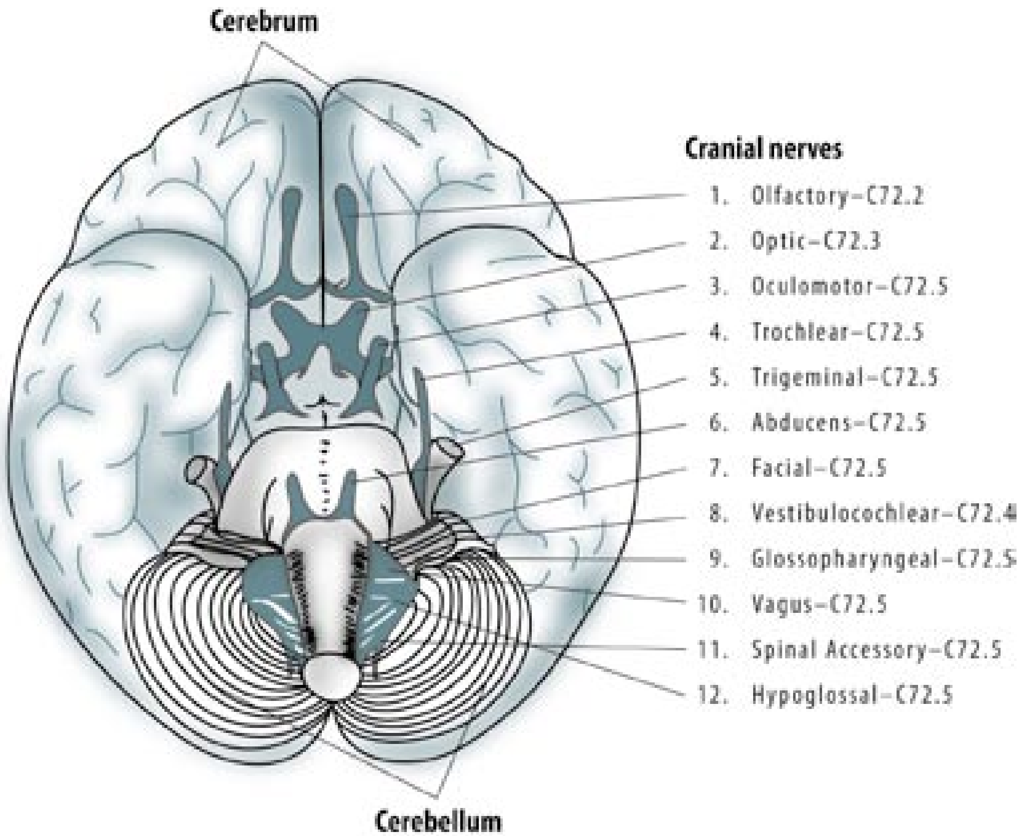


Figure 8.
Cranial nerves

rhythms. Because of the pineal gland's proximity to the ventricular system, hydrocephalus is a symptom of a pineal tumor. In children, one symptom of a pineal tumor is extremely early puberty due to excessive hormonal production caused by the tumor.

The pituitary gland produces several hormones, including growth hormone. Symptoms of pituitary tumors can include diabetes, headache, vision changes, and breast enlargement caused by inappropriate hormone secretion.

Cranial Nerves

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

1. The **olfactory nerve** controls sense of smell.
2. The **optic nerve** controls vision.
3. The **oculomotor nerve** controls eye movement and pupil size.
4. The **trochlear nerve** controls eye movement.
5. The **trigeminal nerve** controls sensation in the face, nose, mouth, teeth, and cornea, as well as chewing, and facial expression.
6. The **abducens nerve** controls eye muscles.

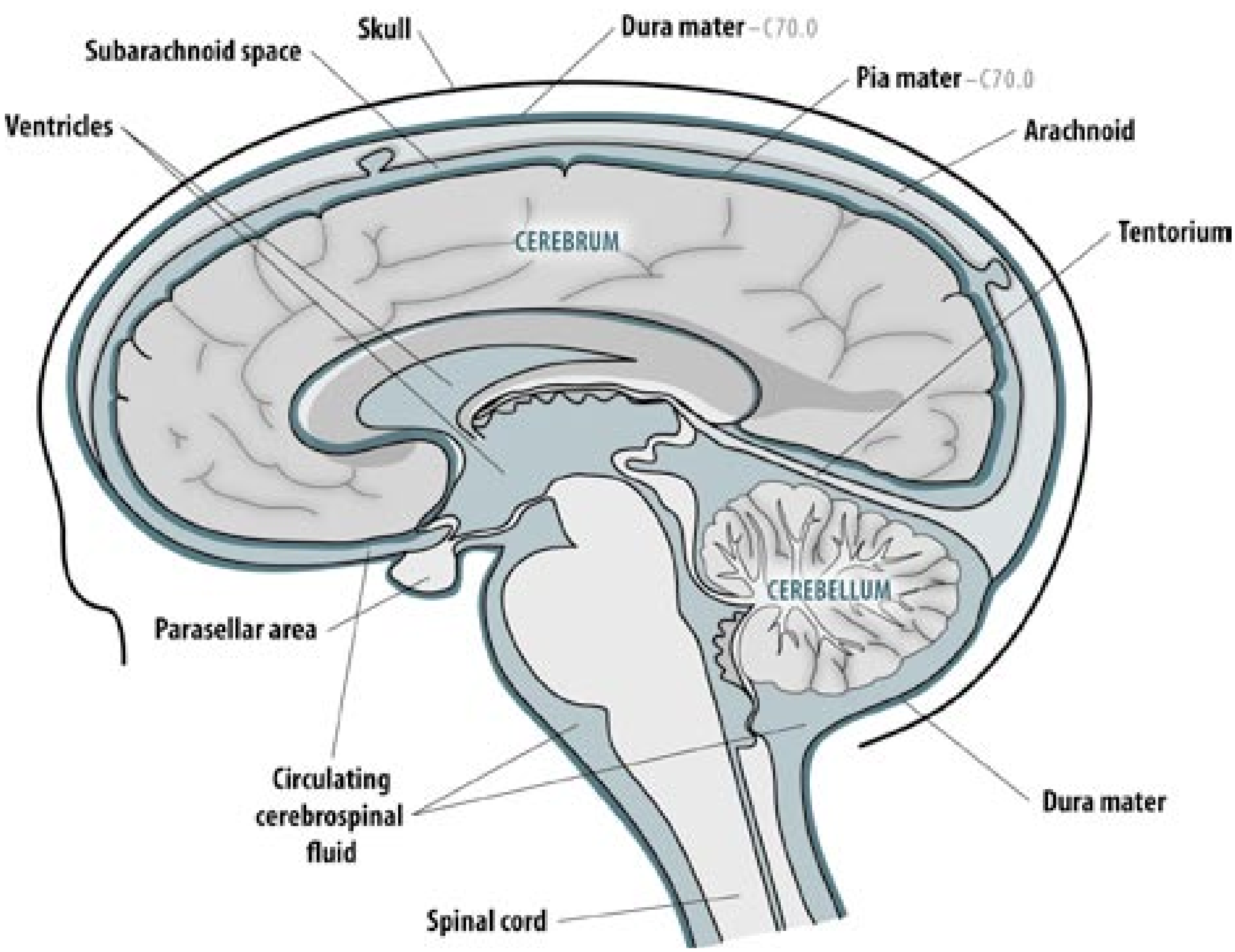
7. The **facial nerve** controls facial expression, tears, and saliva taste.
8. The **vestibulocochlear**, also known as the *acoustic nerve*, controls hearing and balance.
9. The **glossopharyngeal nerve** controls throat movement and sensation as well as taste.
10. The **vagus nerve** controls sensation and muscles in the throat and windpipe.
11. The **accessory nerve** controls movement of the neck.
12. The **hypoglossal nerve** controls tongue movement and swallowing.

Problems with any of the functions described above could be symptoms of a cranial nerve tumor.

Meninges

The meninges are three membranes that cover the brain and spinal cord and protect the CNS (Figure 9):

Figure 9. Meninges



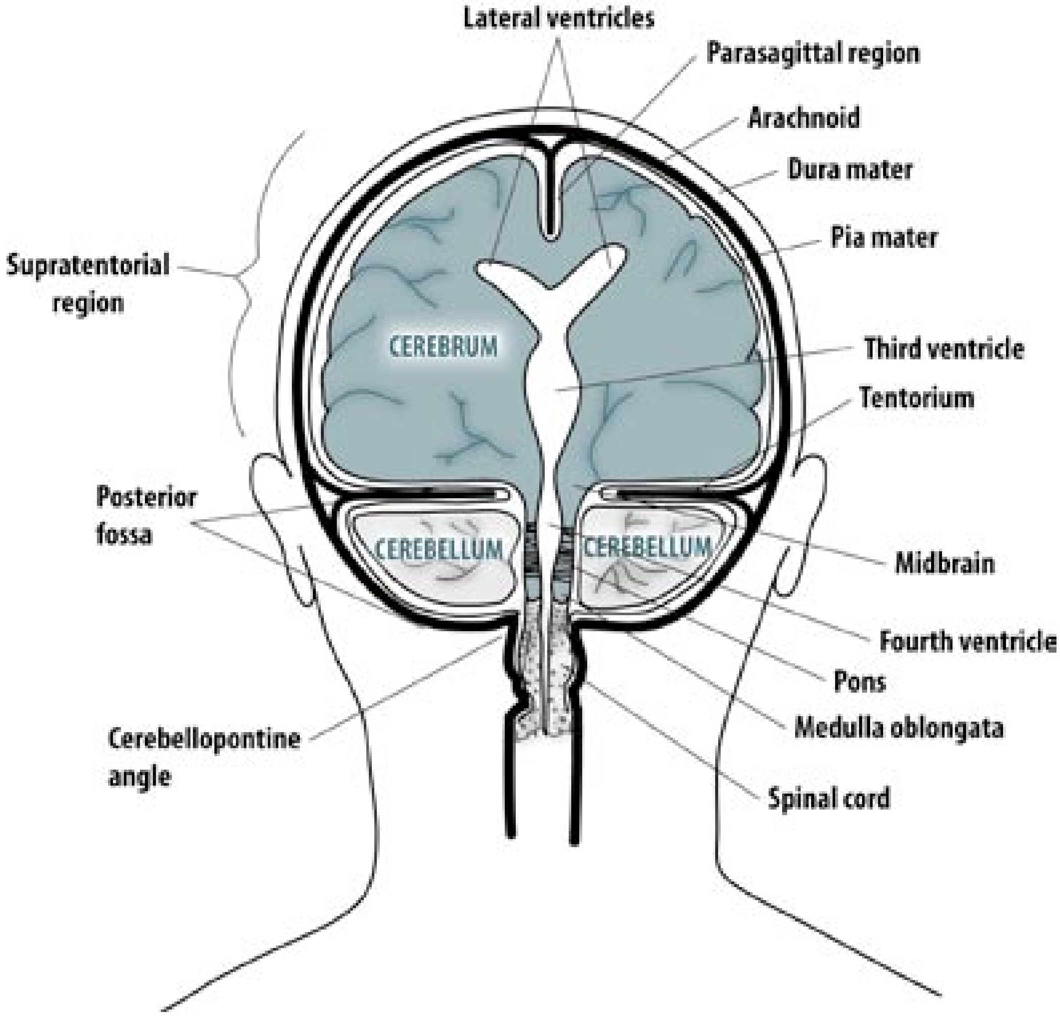


Figure 10. Tentorium

- The **dura mater** is the tough outer membrane.
- The **arachnoid** is the middle web-like membrane.
- The **pia mater** is the delicate, highly vascular, innermost membrane.

The subarachnoid space is between the arachnoid and pia mater and contains CSF. Seizures are the most common symptom of tumors of the meninges. Such symptoms are usually caused by compression and pressure, not by growth into brain tissue.

Tentorium

The tentorium is a flap of meninges that separates the cerebral hemispheres from the posterior fossa (Figure 10). 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.

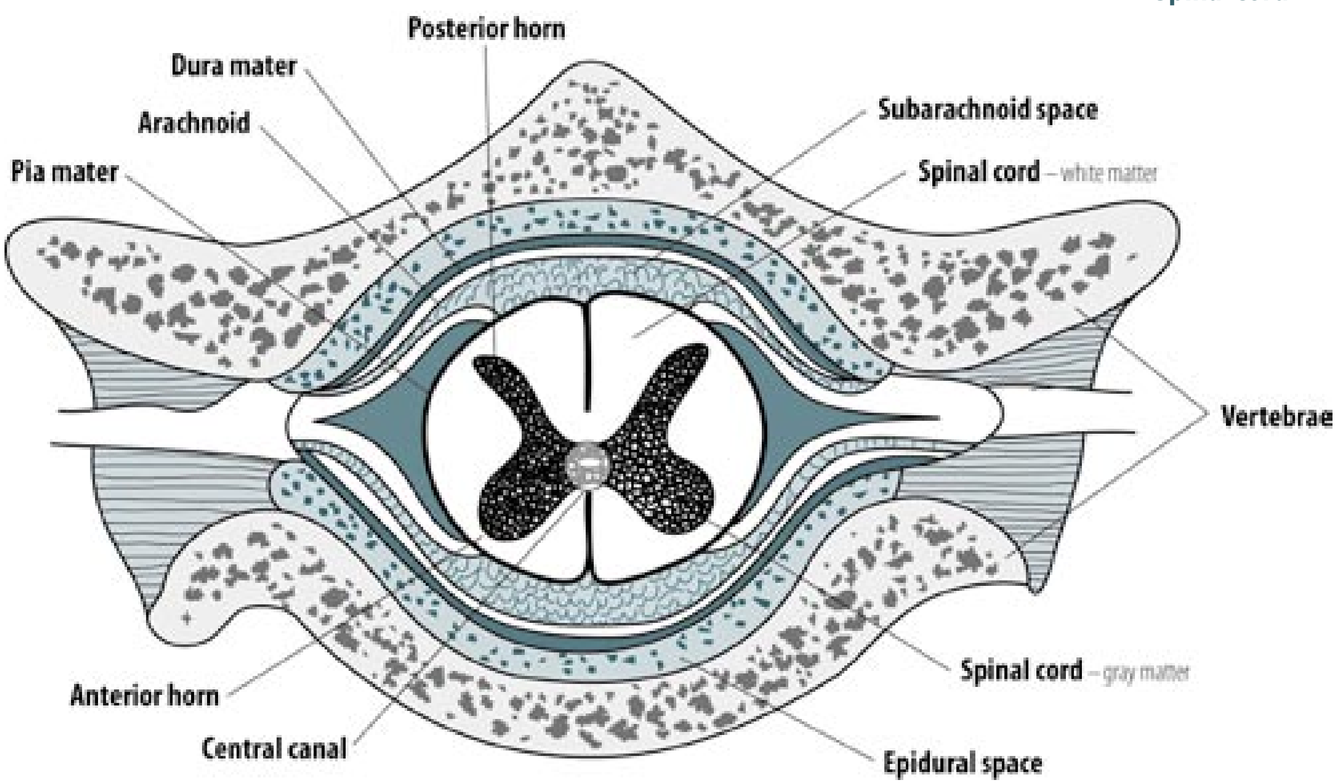
- **Infratentorial** tumors are located below the tentorium in the cerebellum or brain stem and include tumors in the medulla oblongata, which is part of the brain stem.

Spinal Cord

The spinal cord begins in the medulla oblongata and is made up of nerve fibers (Figure 11). Meninges cover and protect the spinal cord. Symptoms of spinal cord tumors vary, depending on the nerves that are involved. With tumors of the thoracic portion of spinal cord, symptoms can include pain in the chest. With tumors of the lumbar or cervical portion of the spinal cord, symptoms might include pain in the neck, arm, back, or leg.

The spinal cord ends in the lumbar area and continues through the vertebral canal as spinal nerves. Because of its resemblance to a horse's tail, the collection of these nerves at the end of the spinal cord is called the cauda equina. These nerves send and receive messages to and from the lower limbs and pelvic organs.

Figure 11.
Spinal cord



Cellular Classification of CNS Tumors

The World Health Organization (WHO) classifies tumors affecting the CNS into two groups:

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

- **Other CNS tumors** are derived from nonglial cells and include sellar tumors, hematopoetic tumors, germ cell tumors, meningiomas, and tumors of cranial nerves.

However, many researchers do not use the WHO classifications; instead, they group brain tumors into the categories of glial tumors and nonglial tumors.

Glial Tumors

Glial tumors develop from glial tissue (the supportive tissue of the brain, made up of astrocytes and oligodendrocytes) and can be benign, borderline, or malignant. Glial tumors are assigned *ICD-O-3* histology codes from the glioma series, codes 938–948. Some histologies of glial origin include glioma, subependymoma, astrocytoma, glioblastoma, and medulloblastoma. Here are more details about the specific types of glial tumors:

- **Astrocytic tumors** are divided into noninfiltrating and infiltrating categories. *Noninfiltrating* tumors include juvenile pilocytic and subependymal astrocytomas, which are often curable. *Infiltrating* tumors include well-differentiated mildly and moderately anaplastic astrocytomas, which are less often curable. For anaplastic astrocytomas of higher grade, the cure rate is low with standard local treatment. The cure rate for glioblastoma multiforme is also very 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. In studies, the overall median survival time of patients with brain stem gliomas has been 44–74 weeks. The best results have been attained with hyperfractionated radiation therapy.

- **Myxopapillary** and **well-differentiated ependymomas** are often curable. Malignant ependymomas have variable prognoses that depend on the location and extent of disease. Frequently, but not invariably, anaplastic ependymomas have a worse prognosis than well-differentiated ependymomas. The rare ependymoblastoma has a much poorer prognosis.

- **Oligodendrogliomas** are more slow-growing tumors that usually occur in young adults. Adult well-differentiated oligodendrogliomas behave very similarly to the well-differentiated mildly and moderately anaplastic astrocytomas. They are frequently located within the frontal, temporal, or parietal lobes and cause seizures in a relatively high percentage of patients. Many oligodendrogliomas contain little specks of calcium (bone) and can easily bleed. Treatment is usually surgery, and, when the tumor is benign, no additional chemotherapy or radiation therapy is recommended following surgery. Anaplastic or mixed oligodendrogliomas are more aggressive forms of this tumor. For anaplastic oligodendrogliomas of higher grade, the cure rate is low with standard local treatment. Both chemotherapy and radiation therapy are usually advised.

- The **mixed glial tumors** have a prognosis similar to that for anaplastic astrocytomas and can be treated as such.

- **Ganglioneuromas** are the rarest form of glioma. These tumors grow relatively slowly and can occur in the brain or spinal cord. They are usually treated with surgery.

- **Optic nerve gliomas** are found on the optic nerve and are particularly common in individuals who have neurofibromatosis. Treatment can include surgery, radiation, or chemotherapy. Some of these tumors are slow-growing and are best managed by observation alone.

Nonglial Tumors

Nonglial tumors are those CNS tumors that develop in areas other than glial tissues. These tumors also can be benign, borderline, or malignant. Some nonglial histologies include meningioma, germ cell tumor, and pituitary adenoma. Here are more details about the specific types of nonglial tumors:

- **Pineal region tumors** include pineocytomas, which 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 germinomas, embryonal cell carcinomas, choriocarcinomas, and teratomas. 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 can be a primary localization or metastatic disease from pineal lesions. Synchronous occurrence in the pineal gland and suprasellar cistern are frequent.

Embryonal cell carciniomas and choriocarcinomas are rare, highly malignant germ cell tumors of the pineal region. In imaging studies, these tumors are devoid of specific characteristics compared with other germ cell tumors. Before biopsy, the physician might suspect the diagnosis after analyzing cerebrospinal fluid for tumor markers. Embryonal cell carcinomas express both alpha fetoprotein (AFP) and the beta subunit of human chorionic gonadotrophin (beta-hCG). Choriocarcinomas express beta-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 or malignant (formerly called teratocarcinomas). Teratomas may also be classified as immature or mature. Mature teratomas are usually benign, while immature teratomas are usually malignant. Teratomas are the second most common pineal region tumor, accounting for 15% of pineal masses. Male predominance ranges from 2:1 to 8:1.

- **Meningiomas** can be benign or malignant. 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 capacity for these tumors to proliferate is greater.

- **Multiple meningiomas** (also called meningiomatosis) are almost always benign and are strongly associated with neurofibromatosis type 2 (NF2) and other genetic disorders. Occasionally multiple meningiomas develop that are not associated with a genetic disorder. These can be referred to as sporadic meningiomas. The *ICD-O* code for multiple meningiomas is M9530/1. Note that this has a behavior code of 1 and should not be used to code multiple or sequential *malignant* meningiomas, which have a behavior code of 3. Multiple malignant meningiomas are very rare and should be coded to 9530/3 (malignant meningioma) unless there is a description of a specific type of meningioma that has a different morphology code.

- **Tumors of the choroid plexus** can be primary or secondary as well as 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.

Magnetic resonance imaging shows a thin rim of cerebrospinal fluid around the tumor. Sometimes, however, the tumor becomes so large that it is difficult to determine its primary intraventricular origin.

Other CNS Tumors

Other CNS tumors include craniopharyngiomas, chordomas, schwannomas, retinoblastomas, primitive neuroectodermal tumors, hematopoetic tumors including cerebral lymphomas and vascular tumors, and other cysts and tumor-like lesions. Here are more details about the specific types of CNS tumors that fall into this category:

- **Craniopharyngiomas** are tumors usually located in the sellar and parasellar region, deriving from squamous epithelium 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, and there is a second smaller peak in the sixth decade. These tumors are often curable.

- **Chordomas** are most common among 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 can 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 can include loss of hearing, ringing in the ears, dizziness, and vertigo. When the condition is detected early, doctors might order an MRI scan and conduct hearing tests, which could 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 might be needed.

- **Retinoblastomas** cause the growth of malignant tumors in the retinal cell layer of the eye. Although the disease is very rare, it is the third most common cancer overall among children, representing about 2% of childhood malignancies. The disease can be inherited or result from a new germinal mutation. About 10% of patients have a family history of retinoblastoma and another 20% to 30% have bilateral disease. Most of the remaining 60% of patients, with unilateral disease and no family history of retinoblastoma, have nonheritable disease. However, about 5% of these patients can also carry the gene for retinoblastoma and risk passing the trait to their children. Tumors appear as single or multiple gray-white elevations in the retina; tumor seeds might be visible in the vitreous. In almost all tumors, calcification can be detected by computed tomography (CT). Untreated, retinoblastoma is almost always fatal; therefore, early diagnosis and treatment are critical in saving lives and preserving visual function. More than 90% of children with retinoblastoma can be cured by early detection and treatment of the affected eye.

- **Primitive neuroectodermal tumors (PNETs)** can occur anywhere in the brain, although the most common site is in the back of the brain near the cerebellum. When they occur here, they are called medulloblastomas. The classification is based on both histopathological characteristics and location in the brain. Undifferentiated neuroectodermal tumors of the cerebellum have historically been referred to as medulloblastomas, whereas tumors of identical histology in the pineal region are referred to as pineoblastomas. The symptoms depend on their location in the brain but typically include increased intracranial pressure. These tumors are fast-growing and often malignant, with occasional spreading throughout the brain or spinal cord. They can spread contiguously to the cerebellar peduncle, the floor of the fourth ventricle, and into the cervical spine, or above the tentorium. In addition, they can spread via CSF intracranially or to the spinal cord. The tumors often block drainage of CSF, causing symptoms associated with increased intracranial pressure. A combination of surgery, radiation, and chemotherapy is usually necessary to control these tumors.

  Neuroblastoma tumors are another type of PNET. These tumors occur in young children, with over 50% of tumors found within the first 5 years of life. Increased intracranial pressure is a symptom of neuroblastoma. These tumors are large solid masses that often hemorrhage and produce calcification and cysts. Neuroblastomas typically originate above the tentorium and are deep-seated within the frontoparietal lobes, adjacent to the lateral ventricles. However, they can be found anywhere in the nervous system.

- **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, these lymphomas affect immunosuppressed individuals, such as patients with AIDS, but they can also develop in people with intact immune systems. The high incidence of these lymphomas in patients with AIDS and frequency in people with intact immune systems have made primary cerebral lymphomas a relatively common brain tumor. The brain, especially the 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.

- **Vascular tumors** are rare, noncancerous 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. Vascular tumors of the CNS are coded to the site within the CNS where they occur, not to blood vessel.

- **Cysts and tumor-like lesions** are reportable only if they are listed in the *ICD-O*. WHO lists several cysts and tumor-like conditions, but only three of these conditions 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 can be found intracranially or in the spinal canal, mainly in or near the midline; some typical locations are in the frontobasal region, the parasellar region, or the posterior fossa. They are cystic masses, with a fibrous capsule. This capsule is lined with squamous epithelium and contains a thick fluid composed of cholesterol, keratin, and lipid metabolites, derived from decomposed epithelial cells. The cysts can rupture, and fatty components of their contents can 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 the tongue and subcutaneous tissue, but a variety of other sites can be involved, including the CNS. Most CNS GCTs arise in the pituitary gland, but rare cases involving the brain and leptomeninges have been described.[1]

- **Rathke pouch tumors** are synonymous with craniopharyngiomas (see earlier description).

The cysts and lesions that are not reportable because they do not have an *ICD-O-3* code include epidermoid cysts, colloid cysts, enterogenous cysts, neuroglial cysts, plasma cell granulomas, nasal glial heterotopias, and Rathke's cleft cyst. Some controversy exists among pathologists regarding the need to include some of these CNS cystic lesions in the *ICD-O-3*. A consensus conference to discuss these issues was held in November 2003. Some pathologists have suggested that the *ICD-O-3* be revised in a few years.

Childhood Versus Adult Tumors

The histologies and locations of CNS tumors differ for children and adults. Brain tumors are classified according to histology, but the tumors' locations and extent of spread are important factors that affect the treatment and prognosis for 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 occur more often in children aged 7 years and younger than in older children. The most common type of childhood CNS tumor is medulloblastoma, a malignant tumor.

Infratentorial Tumors

Approximately 50% of brain tumors in children are infratentorial—originating below the tentorium, which separates the cerebrum from the cerebellum and brain stem. Three-fourths of these tumors are located in the cerebellum or fourth ventricle. Here are some common infratentorial (posterior fossa) tumors that can occur during childhood:

- Cerebellar astrocytomas (usually pilocytic but also fibrillary and high-grade).
- Medulloblastomas (primitive neuroectodermal tumors).
- Ependymomas (low-grade or anaplastic).
- Brain stem gliomas (high-grade or low-grade).
- Atypical teratoid tumors.

Supratentorial Tumors

Brain tumors among children can also be supratentorial—originating above the tentorium, in the sellar or suprasellar region around the sella turcica (the bone that contains the pituitary gland), in the cerebrum, or in the diencephalon (part of the cerebrum). Approximately 20% of childhood CNS tumors are located in the sellar or suprasellar region. Here are some of the supratentorial tumors that can occur during childhood:

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

The histopathology of childhood spinal cord tumors is the same as for childhood brain tumors. Primary spinal cord tumors make up about 1%–2% of all childhood CNS tumors. As with primary brain tumors, such lesions are histologically diverse. Approximately 70% of all intramedullary spinal cord tumors are low-grade astrocytomas or gangliogliomas. Other tumor types include ependymomas, high-grade glial tumors, and rarely PNETs. Myxopapillary ependymomas have a proclivity to develop in the conus and cauda equina regions.

Symptoms and signs of spinal cord tumors differ, depending on the location of the tumors and their extent of spread. Some low-grade spinal cord tumors are associated with large cysts that extend rostrally and caudally and have been termed “holocord astrocytomas.” At times, it is impossible to distinguish a tumor arising in the medulla from a tumor arising in the upper cervical cord.

No uniformly accepted staging system exists for childhood primary spinal cord tumors. These tumors are classified on the basis of their location within the spinal cord and histology. Low-grade spinal cord tumors rarely disseminate elsewhere in the nervous system; however, higher grade tumors can disseminate. Despite this, and because of the location of the tumors and concerns about causing further neurological deterioration by CSF attainment, routine lumbar spinal punctures are not indicated in the evaluation of children with spinal cord tumors. For high-grade glial spinal cord tumors, and possibly lower grade tumors and ependymomas, neuroimaging of the entire neuroaxis (brain and entire spine) is indicated at the time of diagnosis to determine the extent of disease.

The cause of the vast majority of childhood brain tumors remains unknown. More than half of children diagnosed with brain tumors survive 5 years from the diagnosis. In some subgroups of patients, the survival and cure rate is higher. Guidelines for pediatric cancer centers and their role in the treatment of children with cancer have been outlined by the American Academy of Pediatrics.[2,3]

ICD-O-3 Coding Issues

Intracranial Versus Skull Origin

With some tumors, it is difficult to determine if the primary site is intracranial or in the skull (C41.0). Making this distinction is important because nonmalignant tumors that originate in intracranial sites are reportable, whereas those originating in the skull are not reportable. In comparison, all malignant tumors are reportable, regardless of their origin. Here are more details about the specific types of tumors that should be reported:

- **Chondromas** (9220/0) are benign tumors of cartilage cells. The *ICD-O-3* manual shows the code for bone in parentheses next to the morphology. Registrars should review the record carefully to determine if the tumor originated in bone or in an intracranial site. Because a chondroma is a benign tumor, an abstract should only be completed if the primary tumor is in an intracranial site. A chondroma of the skull is not reportable.

- **Chordomas** are malignant tumors arising from the embryonic notochord, and *chondrosarcomas* (9220/3) are malignant tumors of cartilage cells. These types of tumors are reportable, but registrars must determine if the primary site is bone or an intracranial site because intracranial tumors should be analyzed separately.

Pilocytic Astrocytoma

When the *ICD-O-3* was published, the behavior code for pilocytic astrocytoma changed from 3 (malignant behavior) to 1 (borderline behavior). Registrars were instructed to continue to assign the code for malignant behavior. To ensure data consistency, registrars should continue to assign the malignant behavior code (3) to pilocytic astrocytoma.

Intracranial Schwannoma

Cases of intracranial schwannoma (9560/0) diagnosed on or after January 1, 2004, should be reported. It is difficult to determine the intracranial site of a schwannoma. When the primary site of an intracranial schwannoma is not documented in the health record, registrars should assign the *ICD-O-3* site code for cranial nerves, not otherwise specified (C72.5).

CNS Grading Systems and Coding Grade

ICD-O-3 Grade

The sixth digit of the *ICD-O-3* morphology code describes the histologic grade or differentiation of the tumor. Pathologists do not always describe the *ICD-O-3* grade or differentiation for CNS tumors. When the tumor's grade or differentiation is not described, registrars should assign code 9 (not determined, not stated, or not applicable).

Some histologies include differentiation in the terms. When this is the case, the differentiation can be coded. Also, other terms for grade might be used, as is the case with low-grade astrocytomas (M9400/3). In such instances, registrars should assign codes according to the *Facility Oncology Registry Data Standards* (*FORDS*) manual[4] and *SEER Summary Staging Manual*.[5] In this particular case, low-grade should be assigned a grade I–II or code 2.

The *ICD-O-3* grade or differentiation code for nonmalignant CNS tumors is always code 9, as documented in the *ICD-O-3*.[6]

Other grading systems used to describe CNS tumors are the WHO grade, Kernohan grade, and St. Anne-Mayo grade. *These grades are not the same as the ICD-O-3 grade or differentiation* and should not be recorded in the sixth digit histology code data field for grade. Clinicians often use these other grading systems to plan treatment and predict prognosis.

WHO Grade

The WHO grade has four categories of tumors:

- **Grade I** tumors are slow-growing, nonmalignant, and associated with long-term survival.

- **Grade II** tumors are relatively slow-growing but sometimes recur as higher grade tumors. They can be nonmalignant or malignant.

- **Grade III** tumors are malignant and often recur as higher grade tumors.

- **Grade IV** tumors reproduce rapidly and are very aggressive malignant tumors.

Registrars should record the WHO grade in the collaborative stage data field, site-specific factor 1 for brain. If different WHO grades are reported, the rule to use is "grade up, stage down" and code according to the highest grade (worst prognosis). If the pathology report does not give a WHO grade but another diagnostic test such as an MRI does, registrars may use the WHO grade from the diagnostic test.

Kernohan Grade

The Kernohan grade defines progressive malignancy of astrocytomas as follows:

- **Grade 1** tumors are benign astrocytomas.
- **Grade 2** tumors are low-grade astrocytomas.
- **Grade 3** tumors are anaplastic astrocytomas.
- **Grade 4** tumors are glioblastomas multiforme.

There is no North American Association of Central Cancer Registries (NAACCR) data field available to record the Kernohan grade.

St. Anne-Mayo Grade

The St. Anne-Mayo grade also is used to grade astrocytomas; however, this system uses four morphologic criteria to assign a grade: nuclear atypia, mitosis, endothelial proliferation, and necrosis. The St. Anne-Mayo grade has four categories of tumors:

- **Grade 1** tumors do not meet any of the criteria.
- **Grade 2** tumors meet one criterion, usually nuclear atypia.
- **Grade 3** tumors meet two criteria, usually nuclear atypia and mitosis.
- **Grade 4** tumors meet three or four of the criteria.

As with the Kernohan grade, there is no NAACCR data field available to record the St. Anne-Mayo grade.

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

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5. Young JL Jr, Roffers SD, Ries LAG, Fritz AG, Hurlbut AA, eds. *SEER summary staging manual—2000: codes and coding instructions.* Bethesda, Maryland: National Cancer Institute, 2001:102. (NIH publication no. 01-4969.)

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Part II Exercises
Reportability, Coding Site, and Histology

For these exercises, assume that the patient had no previous nonmalignant or malignant tumors of other sites.

Assign ICD-O-3 codes for site and histology even if the case is not reportable.

1. **The patient was seen in the hospital neurology clinic on March 4, 2004 and was prescribed tamoxifen for cerebral meningioma. The patient was first diagnosed with cerebral meningioma in December 2001.**

   Reportable:
   Primary site:
   Histology:

2. **The patient was diagnosed on April 15, 2004 with a chondroma originating in the skull.**

   Reportable:
   Primary site:
   Histology:

3. **The patient was diagnosed on December 1, 2004 with a chordoma of the right frontal lobe extending into the skull.**

   Reportable:
   Primary site:
   Histology:

4. **On February 2, 2002 the patient was diagnosed with low-grade astrocytoma of the cerebellum, Kernohan grade 2.**

   Reportable:
   Primary site:
   Histology:

5. **The patient had an intracranial biopsy on July 1, 2004, and the tumor pathology was WHO grade I schwannoma.**

   Reportable:
   Primary site:
   Histology:

6. **The final pathologic diagnosis for a procedure performed on January 2, 2004 was well-differentiated pituitary adenoma.**

   Reportable:
   Primary site:
   Histology:

7. **The patient had hearing loss on the right side first documented in 2002. In August 2002, a computerized tomography (CT) scan showed acoustic neuroma but no treatment was given. On July 25, 2004 the patient had surgical resection of an intracranial tumor. The final pathologic diagnosis was right acoustic neuroma.**

   Reportable:
   Primary site:
   Histology:

8. **A CT scan in May 2004 identified a lesion in the cerebral meninges. A biopsy of the lesion was used to diagnose cholesteatoma.**

   Reportable:
   Primary site:
   Histology:

9. **Magnetic Resonance Imaging (MRI) was used to identify a pinealoma on February 20, 2004. The patient had gamma knife radiosurgery on March 15, 2004.**

   Reportable:
   Primary site:
   Histology:

10. **A CT scan identified a non-glial tumor in the temporal lobe on October 1, 2004. The tumor was removed and final pathologic diagnosis was meningioma of the left temporal dura.**

    Reportable:
    Primary site:
    Histology:

Part III

Data Reporting Rules

National Program of Cancer Registries Training Materials

2004

Part III
Data Reporting Rules

Multiple Primary Tumors

Coding Laterality

The brain is not a paired organ, but data on which side of the brain is affected (coded in the laterality data item) should be collected for both nonmalignant and malignant central nervous system (CNS) tumors.* Researchers, including epidemiologists, have asked that data on laterality be collected because the location of certain tumors can help in determining the cause. Certain investigations, such as those involving cell phone use, would benefit from having this information routinely available. Also, factors not related to treatment, such as the location of the tumor by hemisphere, can help doctors predict the patient's cognitive outcome.

Registrars should use laterality to determine if multiple *nonmalignant*† CNS tumors are counted as multiple primary tumors. In other words, multiple nonmalignant tumors of the same histology in different locations are counted as multiple primary tumors. Registrars should *not*, however, use laterality to determine if multiple *malignant* tumors of the same CNS site are multiple primary tumors. These malignant tumors should be counted only once (see rationale in box, page 44).

Registrars should code laterality for the following CNS sites:

- Cerebral meninges, not otherwise specified (C70.0).
- Cerebrum (C71.0).
- Frontal lobe (C71.1).
- Temporal lobe (C71.2).
- Parietal lobe (C71.3).
- Occipital lobe (C71.4).
- Olfactory nerve (C72.2).
- Optic nerve (C72.3).
- Acoustic nerve (C72.4).
- Cranial nerve, not otherwise specified (C72.5).

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* CNS is used to describe intracranial and central nervous system tumors.
† A nonmalignant tumor is any tumor with an *ICD-O-3* behavior code of 0 (benign) or 1 (borderline).

When coding these tumors for laterality, registrars should use codes 1–4 or 9 (paired site but lateral origin unknown; midline tumor). The laterality for all other CNS sites should be coded 0 (not a paired organ).

Rationale for Multiple Primary Rule Changes

The North American Association of Central Cancer Registries (NAACCR) Registry Operations Brain Tumor Subcommittee reviewed the rules for determining multiple primary tumors of all CNS sites and histologies and recommended changes to these rules. The NAACCR Uniform Data Standards Committee approved the changes recommended for nonmalignant tumors but not those for malignant tumors. The changes suggested for malignant CNS tumors have been submitted to the Surveillance Epidemiology and End Results (SEER) Histology Coding Committee which is reviewing multiple primary rules for all sites.

The changes to the multiple primary rules for nonmalignant tumors are based on the rationale that the natural biology of nonmalignant tumors is that of expansive localized growth. Local recurrences are common, and metastases are uncommon. Nonmalignant brain tumors confine themselves to one location, their site of origin. Thus, any new nonmalignant brain tumor in another location represents a new tumor, a clinically significant event in the life of the patient worthy of capture. Therefore, if multiple nonmalignant tumors of the same histology are identified in different locations, they should be counted as separate primary sites. However, if nonmalignant tumors of the same histology, same site, and same side recur in the same location—even after 20 or so years—they should still be counted as the same tumor.

Multiple Primary Rules for Malignant CNS Tumors

Although the NAACCR Registry Operations Brain Tumor Subcommittee has suggested some rule changes for *malignant* CNS tumors (see box), at this time the rules for determining multiple primary tumors for malignant CNS tumors will remain the same as they are now. These rules are as follows:

- Each category (first three characters), as delineated in the *ICD-O-3*, is considered a separate site. Therefore, multiple tumors occurring in the cerebrum (**C71**.0) and the temporal lobe of the brain (**C71**.2) are considered the same tumor, and only one abstract is completed. Multiple tumors occurring in the meninges (**C70**.0–**C70**.9) and in the cerebrum (**C71**.0) are considered different tumors, and separate abstracts are completed.

- Differences in histologic type refer to differences in the first three digits of the morphology code. Therefore, if multiple tumors occur in the *same site*, and the first three digits of the histology code are the same, they are considered the *same tumor*, and one abstract should be completed. An example of this is a choroid plexus carcinoma (M**939**0) and an ependymoma (M**939**1).

  If the first three digits of the histology codes are different, the tumors are considered different, and separate abstracts should be completed. An example of this is an astrocytoma (M**940**0) and a gemistocytic astrocytoma (M**941**1).

  If the first three digits are the same, and the fourth digit is a 9 or not otherwise specified (NOS) site, this is considered one site and should be coded to the *more specific site*. For example, if a tumor is identified as meninges, NOS (C70.**9**), and a separate tumor is identified as occurring in either the spinal (C70.**1**) or cerebral (C70.**0**) meninges, this is considered one tumor, and only one abstract should be completed using the more specific site code.

New Multiple Primary Rules for Nonmalignant Tumors

Determining If Site Is Same or Different

Each *subsite* (fourth-digit level) as delineated in *ICD-O-3* is considered a separate site.

- If separate tumors with the same histology occur *in the same subsite* they are considered the same tumor and one abstract is completed. Therefore, if multiple tumors of the same histology occur in the cerebrum (C71.**0**), they are considered the same tumor regardless of when they occur and only one abstract is completed.

- If separate tumors with the same histology occur in *different subsites*, they are different tumors and separate abstracts are completed. Therefore, if a tumor occurs in the cerebral meninges (C70.**0**), and a separate tumor occurs in the spinal meninges (C70.**1**), they are considered separate tumors. Likewise, if separate brain tumors of the same histology occur in the frontal lobe (C71.**1**), and in the occipital lobe (C71.**4**), they are also considered separate tumors.

- As with malignant tumors, if the first three digits are the same, and the fourth digit is a **9** or not otherwise specified (NOS) site, this is considered one site and should be coded to the *more specific site*. For example, if a tumor is identified as meninges, NOS (C70.**9**), and a separate tumor is identified as occurring in either the spinal (C70.**1**) or cerebral (C70.**0**) meninges, this is considered one tumor, and only one abstract should be completed using the more specific site code.

- Laterality is used to determine multiple primaries for nonmalignant CNS tumors for sites listed as being lateral. If multiple tumors of the same site and same histologic type are identified and *both sides* of a site (listed as lateral) are involved, the tumors should be considered to be separate tumors, and separate abstracts should be completed. For example, the right and left temporal lobes of the brain or the right and left acoustic nerves.

Determining If Histology Is Same or Different

The codes in Table 2 apply only to nonmalignant histologies. A separate table for malignant histologies is being reviewed by the SEER Histology Coding Committee. Originally, *ICD-O* histology codes were grouped into categories that were considered biologically related. Hence, the epithelial neoplasms were coded M801–804, and the squamous cell neoplasms were coded M805–808. This is the origin of the multiple primary rule—that differences in histologic type refer to differences in the first three digits of the histology code.

Over time the World Health Organization (WHO) classification of brain tumors has undergone several revisions, and some new codes have been inserted in the *ICD-O*. However, these codes were inserted where numbers were available rather than where the code would logically fit. Thus, all codes in a three-digit rubric might not be part of the same histologic group.

Brain tumor histologies grouped in Table 2 do not follow the standard three-digit histology difference rule because they represent a progression, differentiation, or subtype of a single histologic category. In reviewing the histology codes, it was found that applying the current three-digit histology rule to nonmalignant CNS tumors would combine tumors that are no longer considered to be biologically related.

To determine if the histology in multiple nonmalignant CNS tumors is the same, registrars should consult Table 2 and follow these rules, in priority order:

A-1. When multiple tumors are in the *same site*; the first three digits of the histology code are *the same*; and the codes are not found in Table 2, then the histology is considered to be *the same*. Only one abstract should be completed.

A-2. When multiple tumors are in the *same site*; the first three digits of the histology code are *different*, and the codes are not found in Table 2,

Table 2. *ICD-O-3* Code Groupings to Determine If Histology Is the Same for Multiple Nonmalignant Brain Tumors

Tumor	*ICD-O-3* Code Groupings
Choroid plexus neoplasms	9390/0, 9390/1
Ependymomas	9383, 9394, 9444
Neuronal and neuronal-glial neoplasms	9384, 9412, 9413, 9442, 9505/1, 9506
Neurofibromas	9540/0,9540/1, 9541, 9550, 9560/0
Neurinomatosis	9560/1
Neurothekeoma	9562
Neuroma	9570
Perineurioma, not otherwise specified	9571/0

then the histology is considered to be *different*. Separate abstracts should be completed.

This follows the existing rule for histology—that differences in histologic type refer to differences in the *first three* digits of the morphology code. This is the same rule as for malignant CNS tumors. In addition, when multiple tumors are determined to be the same primary tumors but with different histology codes, registrars should follow existing rules for selecting which histology to code.[1] When no single code includes all diagnostic terms, use the numerically higher code number.

B. If all histologies are in the *same histologic* group in Table 2, then the histology is considered to be the *same*, even though the first three digits are different. These two histologies represent a progression, differentiation, or subtype of a single histologic category. One abstract should be prepared.

If one of the histologies is nonspecific and one is specific, the more specific histology should be coded. If separate tumors occur with *different histologies* in the same group in Table 2, the *first histology* (the one with the earliest diagnosis date) should be coded.

Example. A patient has a diagnosis of a choroid plexus papilloma, *not otherwise specified* (9390/0), a *nonspecific term*, and subsequently has a diagnosis of atypical choroid plexus papilloma (9390/1), a *more specific* histology, in the same site. These terms are both in the grouping of choroid plexus neoplasms in Table 2. The 9390/0 is the first diagnosis (earliest diagnosis date), but 9390/1 is more specific. Therefore, 9390/1 should be used.

Example. A patient has subependymoma (9383/1), a specific histology, and is subsequently diagnosed with a choroid glioma (9444/1), also a specific histology. Both histologies are listed in Table 2 under the ependymomas grouping. In this instance, because both histologies are specific and in the same grouping, the *first* histology of subependymoma (9383/1) should be coded even though the second histology has a higher code.

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

Example. A patient has a ganglioglioma (M9505/1) listed in Table 2 in the grouping neuronal and neuronal-glial neoplasm, as well as a separate Pacinian tumor (M9507/0) which is not listed in Table 2. These two tumors have the same first three digits. In this instance, the Pacinian tumor is considered the same tumor as ganglioglioma, and only one abstract should be completed. The first histology of ganglioglioma (M9505/1) should be coded even though the second histology has a higher code.

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

Example. A patient has a choroid plexus papilloma (M9390/0), listed in Table 2 in the choroid plexus neoplasm grouping, as well as a myxopapillary ependymoma (M9394), which is listed in the ependymoma grouping. In this case, even though the first three digits are the same, the histologies are considered to be different for these tumors because they are listed in different groupings in Table 2. Thus, two abstracts should be completed.

General Rules for Determining Multiple Primaries of CNS Sites

If a patient has multiple CNS tumors that are *all nonmalignant* and—

- they are in different sites, then they are *separate primaries*.
- they have different histologies, then they are *separate primaries*.
- the site and histology are the same and the laterality of one tumor is unknown or not applicable, they are counted as a *single primary*.
- the site and histology are the same and the laterality is both sides, these tumors should be considered *separate primary tumors*.

Current Timing Rule

The current timing rule for determining multiple primary tumors applies to malignant CNS tumors. If two or more primary malignant CNS tumors are diagnosed in the same site with the same histology within 2 months of the diagnosis of the first primary, the tumors are counted as one primary. If multiple tumors with the same histology in the same site are diagnosed more than 2 months apart, the tumors are counted as separate primary sites.

The current 2-month rule *does not apply* to *nonmalignant* CNS tumors. Nonmalignant tumors often recur in the same location and are considered the same tumor, regardless of the time frame. If a nonmalignant tumor of the same histology, same site, and same side recurs in the same location—even after 20 or so years—the two tumors are still considered the same tumor.

If a patient has multiple tumors and one is *malignant* and one is *nonmalignant*, they are *always* considered to be separate primary tumors regardless of the time of diagnosis.

- If a nonmalignant tumor is followed by malignant tumor, they are considered *different* primaries, regardless of timing.
- If a malignant tumor is followed by a nonmalignant tumor, they are considered *different* primaries, regardless of timing.

Histologic Transformation to Higher Grades

CNS tumors can transform or progress to a higher grade. This process will be documented in physician statements in the patient medical records or pathological reviews. Pathologists develop a final diagnosis of transformation or progression by comparing slides from previous biopsies or excisions of the original tumor with slides of newly biopsied or resected CNS tumors. Transformation may be reported as a recurrence with a different behavior. This transformation or grade change can result in a difference in the morphology code of the tumor. Histologic transformation can change a nonmalignant tumor into a malignant tumor or change a malignant tumor to a higher grade with a different histologic code.

If a *malignant* CNS tumor recurs or progresses to a higher grade tumor, it is still considered the same tumor. The original diagnosis should not be changed, and a separate abstract should not be created. For example, if an astrocytoma (M9400) *recurs* (transforms) as a glioblastoma multiforme (M9440), this should be coded as one primary tumor because a glioblastoma multiforme is defined as a grade 4 astrocytoma. The histology code (M9400) should not be changed.

Malignant Transformation‡

In rare cases, a diagnosed nonmalignant tumor transforms into a malignant tumor. In these cases, the morphology changes from WHO grade I to WHO grade II, III, or IV, or the behavior changes from code 0 or 1 to code 2 or 3.

When malignant transformation occurs in a *previously diagnosed nonmalignant* tumor, the tumors are considered *separate primaries*, and two abstracts should be completed because of the change from nonmalignant to malignant. Recording these tumors as separate primaries will allow researchers investigating these conditions to identify cases. This will also allow these cases to be included when registries report only malignant cases. This rule is similar to the rule that if an *in situ* tumor recurs as an invasive tumor, a second abstract should be completed for the invasive tumor.

Sequence Numbers Associated with Malignant Transformation

When malignant histologic transformation has occurred, registrars should assign the sequence number for the nonmalignant tumor using the reportable-by-agreement series of sequence numbers (60–87); this series includes other non-CNS reportable-by-agreement cases. The malignant tumor should be assigned a sequence number from the malignant series of sequence numbers (00–35). For example, if a patient had one nonmalignant CNS tumor that progressed into a malignant CNS tumor, the sequence number for the nonmalignant tumor would be 60, and the sequence number for the malignant tumor would be 00.

Code 60 is used in the same way as 00 in that it represents the occurrence of only one reportable-by-agreement tumor. If a second nonmalignant tumor is diagnosed or if other reportable-by-agreement cases occur, the first tumor is coded as 61 and the second as 62.

Date of Diagnosis for Transformation Cases

When recording the date of diagnosis for transformation cases, the date for a nonmalignant tumor is the date that a medical practitioner first diagnosed the nonmalignant tumor either clinically or histologically. The date of diagnosis for the malignant tumor is also the date that the malignant transformation was first diagnosed by a medical practitioner either clinically or histologically.

‡ Malignant transformation and progression to malignancy are the same thing.

Coding Sequence Numbers: General

The sequence of the occurrence of neoplasms throughout the lifetime of a patient is recorded in the sequence number data field. Instructions for assigning sequence numbers are found in the *Facility Oncology Registry Data Standards (FORDS)* manual.[2]

Malignant and *in situ* neoplasms, including malignant CNS neoplasms, are assigned codes 00–35. If a patient has only one primary malignant or *in situ* neoplasm, the sequence number assigned is 00. If a patient has multiple malignant primary neoplasms during his or her lifetime, the sequence number for the first tumor is 01, the sequence number for the second primary tumor is 02, and so forth.

Nonmalignant tumors are coded in the reportable-by-agreement range of codes 60–87. If only one reportable-by-agreement tumor occurs, it is coded as 60. If subsequent nonmalignant tumors or other reportable-by-agreement tumors are diagnosed, the first tumor should be coded as 61 and the second tumor as 62.

Reportable-by-agreement neoplasms are defined by each facility and central cancer registry. They are not considered reportable by the Commission on Cancer (CoC) of the American College of Surgeons. Nonmalignant CNS tumors are assigned reportable-by-agreement sequence numbers in the 60–87 range, even though they are defined as reportable. This allows for consistency in sequence numbers for registries that abstracted these cases before the Benign Brain Tumor Cancer Registries Amendment Act, Public Law 107-260 was enacted.

Sequence numbers for both malignant cases and reportable-by-agreement cases are assigned over a lifetime. Therefore, if a patient was diagnosed with a nonmalignant CNS neoplasm before reporting was required (January 1, 2004) and had a second nonmalignant CNS neoplasm diagnosed in 2004, the second neoplasm should be assigned sequence number 62, even though an abstract is required only for the second tumor.

Date of Diagnosis

The same rules are used to assign the date of diagnosis for nonmalignant and malignant CNS tumors. These rules are found in the *FORDS* manual.[3]

However, it should be noted that it is not unusual for a nonmalignant CNS tumor to be diagnosed in a physician's office and for the patient to be treated with watchful waiting. Several years might go by before the patient undergoes surgery, radiation therapy, or some type of systemic therapy at a health care facility. Also, nonmalignant CNS tumors, especially meningiomas, often recur.

The date of *initial* diagnosis, not the date of subsequent treatment or date of recurrence, should be recorded in the abstract. Health records must be reviewed carefully to determine the initial date of diagnosis by a medical practitioner, regardless of whether the initial diagnosis was clinical or histologic.

References

1. Fritz A, Percy C, Jack A, et al., eds. *International classification of diseases for oncology, 3rd ed.* Geneva: World Health Organization, 2000:34, rule K.

2. American College of Surgeons. *Facility oncology registry data standards.* Chicago, Illinois: American College of Surgeons, 2002:34-5, section 2, Coding Instructions.

3. American College of Surgeons. *Facility oncology registry data standards.* Chicago, Illinois: American College of Surgeons, 2002:89-90, section 2, Coding Instructions.

Part III Exercises

Multiple Primaries, Diagnosis Date, Sequence Number, Laterality, Collaborative Stage

For these exercises, assume that the patient had no previous benign or malignant tumors of other sites.

1. **The patient had a computerized tomography (CT) scan on January 2, 2004, showing an acoustic neuroma. On December 3, 2004, the patient had a craniotomy and removal of the tumor. In the pathology report the final diagnosis was acoustic neuroma.**

   What is the date of diagnosis?
   What is the sequence number?
   Primary site:
   Histology:

2. **The patient had an excisional biopsy on March 1, 2004, and the pathology was WHO grade I gangliocytoma of the basal ganglia. On October 15, 2004, the patient had re-resection of a tumor of the basal ganglia. The final pathologic diagnosis was anaplastic ganglioglioma, WHO grade III.**

   What is the date of diagnosis?
   What is the sequence number?
   Primary site:
   Histology:

3. **The patient is deaf. In 1998, the patient had surgery to remove an acoustic neuroma. A CT scan showed a spinal cord tumor on March 3, 2004. On March 21, 2004, the patient had a laminectomy and partial removal of tumor at T7. The pathology report documented psammomatous meningioma of the dura.**

   What is the date of diagnosis?
   What is the sequence number?
   Primary site:
   Histology:

4. **On April 1, 2004 the patient had a CT scan of the head that showed cholesteatoma. On April 15, 2004, an MRI of the head showed left temporal meningioma. On April 30, 2004, the patient had surgery to remove the meningioma. The final pathologic diagnosis was meningioma of the left inferior temporal dura.**

   What is the date of diagnosis?
   What is the sequence number?
   Primary site:
   Histology:

5. **An MRI on January 3, 2004, was used to diagnose subependymoma. On January 31, 2004, the patient had a stereotactic craniotomy and removal of the subependymoma from the medulla oblongata. The patient later had a bulge in the lumbar spinal cord, and on December 15, 2004, an MRI was used to diagnose meningioma. On December 30, 2004, the meningioma was removed. The pathology report documented intradural meningioma.**

   What is the date of diagnosis?
   What is the sequence number?
   Primary site:
   Histology:

Part IV
Staging, Diagnosis, Treatment, and Related Data Issues

National Program of Cancer Registries Training Materials

2004

Part IV
Staging, Diagnosis, Treatment, and Related Data Issues

Collaborative Staging

Collaborative staging (CS) is a set of data fields that describe the extent of disease. These codes are organized by site in schemas (sets of codes).[1] CS data are collected for all tumors diagnosed on or after January 1, 2004. The CS data fields for *extension*, *lymph node involvement*, and *metastasis at diagnosis* are site-specific. In addition, there is a CS data field for all central nervous system (CNS) sites (*site-specific factor 1*) where registrars should record the World Health Organization (WHO) grade of the tumor. A computer algorithm is applied to the CS data fields to calculate T-N-M (tumor; regional lymph node; metastasis) stage; Surveillance, Epidemiology, and End Results (SEER) Summary Stage 1977; and SEER Summary Stage 2000 for specific sites.[2,3] No T-N-M stage is available for brain tumors at this time.

CS Extension

Three schemas are used to describe the area affected and the extent of the tumor's spread for CNS sites (Table 3):

Brain and Cerebral Meninges. These extension codes describe what areas of the cerebral meninges (C70.0) or brain (C71.0–C71.9) are involved by tumor and how far the tumor has spread. Registrars should assign code 05 to nonmalignant tumors. All of the other codes are used for malignant tumors only:

- **Codes 10–12** should be assigned when the tumor is confined to a single intracranial location and is localized.
- **Codes 15–30** are used to describe localized tumors.
- **Codes 40–51** are used to indicate that the tumors have crossed the midline of the brain or extended across the tentorium and are categorized as regional disease.
- **Code 60** is for regional disease and represents tumor invasion into the skull, blood vessels, dural meninges, nerves, or spinal cord.

- **Code 70** is used to describe distant disease resulting from cells in the cerebrospinal fluid circulating to the spinal cord and brain. This code also represents direct tumor extension through the skull into the nasal cavity, posterior pharynx, or further.

- **Code 80** is assigned for distant extension not described in code 70.

- **Code 95** is used when intracranial malignancy is diagnosed, but the tumor itself cannot be found.

- **Code 99** indicates that the tumor extension is unknown.

Other Parts of the CNS. These extension codes describe the area affected and extent of spread for tumors of the spinal meninges (C70.1); meninges, not otherwise specified (C70.9); and spinal cord, cranial nerves, and other parts of the CNS (C72.0–C72.9).* The descriptions in this schema are less detailed than those for the cerebral meninges and brain. Registrars should use code 05 for nonmalignant tumors. The other codes are to be used for malignant tumors only:

- **Codes 10 and 30** are used to describe localized disease.

- **Code 40** is used to define regional extension for spinal and not otherwise specified tumors of the meninges.

- **Code 50** is used to describe regional extension for any of the included sites.

- **Code 60** is used to describe regional disease for cranial nerve tumors.

- **Codes 70 and 80** are used to describe distant disease.

- **Code 95** is used when intracranial malignancy is diagnosed but the tumor itself cannot be found.

- **Code 99** indicates that the tumor extension is unknown.

Thymus, Adrenal Gland, and Other Endocrine Glands. The third set of extension codes is used to describe tumor extension for the pituitary gland (C75.1), craniopharyngeal duct (C75.2), and pineal gland (C75.3), in addition to several other sites. Code 00 is used to describe *in situ* neoplasms, and code 05 is used to describe benign tumors. The other codes are to be used for malignant tumors only:

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* These codes are compatible with the American Joint Committee on Cancer's *Cancer Staging Manual, 4th Edition*[4] schema for spinal cord; however, the 6th edition of this manual[5] does not recommend a schema for these sites.

- **Codes 10–30** indicate local disease for all sites listed.
- **Codes 40–60** indicate regional tumor spread. Adjacent connective tissue in code 40 indicates involvement with tissues that immediately surround the structure with primary cancer, but these are not adjacent organs. Code 60 lists adjacent organs specific to the pituitary gland and the craniopharyngeal duct as well as the pineal gland.
- **Code 80** is used for distant disease.
- **Code 95** is used when malignancy is diagnosed in the gland but the tumor itself cannot be found.
- **Code 99** indicates that the tumor extension is unknown.

CS Lymph Node Involvement

Only two codes are used to describe a CNS tumor's spread into the regional lymph nodes (Table 3):

Brain and Cerebral Meninges and Other Parts of the CNS. The code used for lymph node involvement for these two schemas is the same. Because no lymph node drainage occurs for these sites, these schemas contain only one code for CS Lymph nodes which is code 88 (not applicable).

Thymus, Adrenal Gland, and Other Endocrine Glands. The CS lymph node code 99 (unknown) for this schema lists pituitary gland (C75.1), craniopharyngeal duct (C75.2), or pineal gland (C75.3), indicating that for these sites, lymph node involvement is always unknown.

CS Metastasis at Diagnosis

Two unique sets of codes are used to describe distant metastasis identified at the time the tumor is diagnosed for the CNS sites (Table 3).

Brain and Cerebral Meninges. The codes for this schema describing the areas that are affected by metastasis at diagnosis are unique.

Other Parts of the CNS and **Thymus, Adrenal Gland, and Other Endocrine Glands**. The codes for these two schemas, describing the areas that are affected by metastasis at diagnosis are the same, but they are different than those for the brain and cerebral meninges.

Table 3. Collaborative Staging Codes for All Intracranial and CNS Tumors

CS Extension

Brain and Cerebral Meninges

05	Benign or borderline brain tumors
10	Supratentorial tumor confined to cerebral hemisphere (cerebrum) or meninges of cerebral hemisphere on one side: frontal lobe, occipital lobe, parietal lobe, or temporal lobe
11	Infratentorial tumor confined to cerebellum or meninges of cerebellum on one side: vermis, which includes the lateral lobes and median lobe of cerebellum
12	Infratentorial tumor confined to brain stem or meninges of brain stem on one side: medulla oblongata, midbrain (mesencephalon), pons, hypothalamus, or thalamus
15	Confined to brain, not otherwise specified, confined to meninges, not otherwise specified
20	Infratentorial tumor: both cerebellum and brain stem involved with tumor on one side
30	Confined to ventricles; tumor invades or encroaches upon ventricular system
40	Tumor crosses the midline: involves the contralateral hemisphere, involves corpus callosum (including splenium)
50	Supratentorial tumor extends infratentorially to involve cerebellum or brain stem
51	Infratentorial tumor extends supratentorially to involve cerebrum (cerebral hemisphere)
60	Tumor invades bone (skull), major blood vessel(s), meninges (dura), nerves, not otherwise specified (cranial nerves), or spinal cord/canal
70	Circulating cells in cerebrospinal fluid; nasal cavity; nasopharynx; posterior pharynx; or outside CNS
80	Further contiguous extension
95	No evidence of primary tumor
99	Unknown extension, primary tumor cannot be assessed, not documented in patient record

Other Parts of the CNS

05	Benign or borderline tumors
10	Tumor confined to tissue or site of origin
30	Localized, not otherwise specified
40	Meningeal tumor infiltrates nerve, nerve tumor infiltrates meninges (dura)
50	Adjacent connective/soft tissue, adjacent muscle
60	Brain, for cranial nerve tumors, major blood vessel(s), sphenoid and frontal sinuses (skull)
70	Brain except for cranial nerve tumors; bone, other than skull; eye
80	Further contiguous extension
95	No evidence of primary tumor
99	Unknown extension, primary tumor cannot be assessed, not documented in patient record

Thymus, Adrenal Gland, and Other Endocrine Glands

00	*In situ*, noninvasive, intraepithelial
05	Benign or borderline tumors
10	Invasive carcinoma confined to gland of origin
30	Localized, not otherwise specified
40	Adjacent connective tissue
60	Pituitary and craniopharyngeal duct: cavernous sinus, infundibulum, pons, sphenoid body, sinuses. Pineal gland: infratentorial, central brain
80	Further contiguous extension
95	No evidence of primary tumor
99	Unknown extension, primary tumor cannot be assessed, not documented in patient record

CS Lymph Node Involvement

Brain and Cerebral Meninges and Other Parts of the CNS

88	Not applicable

Thymus, Adrenal Gland, and Other Endocrine Glands

99	For pituitary gland (C75.1), craniopharyngeal duct (C75.2), and pineal gland (C75.3): not applicable

CS Metastasis at Diagnosis

Brain and Cerebral Meninges

00	No; None
10	Distant metastases
85	Drop metastasis (cells in the cerebrospinal fluid have circulated to the spinal column and begun to grow)
99	Unknown, distant metastasis cannot be assessed, not documented in patient record

Other Parts of the CNS and Thymus, Adrenal Gland, and Other Endocrine Glands

00	No; None
10	Distant lymph node(s) metastases
40	Distant metastases other than distant lymph nodes (code 10); distant metastasis, not otherwise specified, carcinomatosis (10 and 40)
50	Combination of distant lymph nodes and other distant metastases
99	Unknown, metastasis cannot be assessed, not documented in patient record

Site-Specific Factor 1 (WHO Grade)

010	Grade I (slow-growing, nonmalignant, associated with long-term survival)
020	Grade II (relatively slow-growing but sometimes recur as higher grade tumors; can be nonmalignant or malignant)
030	Grade III (malignant and often recur as higher grade tumors)
040	Grade IV (reproduce rapidly; very aggressive malignant tumors)
999	Clinically diagnosed, WHO grade unknown, not documented, or not otherwise specified

Site-Specific Factor 1

Data fields for site-specific factors allow registrars to validate the extent of disease at diagnosis and record other prognostic information. Registrars should record the tumor's WHO grade in the *site-specific factor 1* data field—*not* in the sixth digit histology data field (Table 3) for meninges, brain, spinal cord, cranial nerves, and other parts of the CNS. It is not recorded for the pituitary gland, craniopharyngeal duct, and pineal gland.

The WHO grade describes a CNS tumor's aggressiveness and helps clinicians estimate prognosis. Registrars can usually find the WHO grade in the pathology report. If the pathology report does not give a WHO grade but another diagnostic test does, registrars may use the WHO grade from the diagnostic test. If different WHO grades are reported, the rule is to "grade up, stage down" and code according to the highest grade (worst prognosis). If the WHO grade is not recorded, code 999 should be used.

Risk Factors

Malignant and nonmalignant CNS tumors have been the subject of considerable study. Why they occur is still unclear, but several risk factors have been identified:

- **Genetic predispositions** to the development of brain tumors have been identified; however, population-based studies suggest that no more than 4% of these tumors can be attributed to heredity.

- Several **environmental factors** can be associated with CNS tumors, including exposure to ionizing radiation, electromagnetic fields, pesticides, vinyl chlorides, and polycyclic hydrocarbons.

- The presence of the Epstein-Barr virus in the DNA of primary lymphoma suggests that a **viral etiology** for CNS tumors cannot be entirely ruled out.

Accurate and complete data are necessary to develop hypotheses for identifying the causes of CNS tumors. The heterogeneity of CNS tumors can mask their causes when histology-specific studies are limited by the number of available cases (for details, see *Surveillance of Primary Intracranial and Central Nervous System Tumors: Recommendations from the Brain Tumor Working Group* in Appendix E).

Genetic Syndromes

Several genetic syndromes are associated with the occurrence of multiple CNS tumors:

- Neurofibromatosis I (von Recklinghausen's disease)
- Neurofibromatosis II (bilateral acoustic neurofibromatosis)
- Von Hippel-Lindau disease
- Tuberous sclerosis (Bourneville-Pringle syndrome)
- Gorlin syndrome (nevoid basal cell carcinoma syndrome)
- Li-Fraumeni syndrome
- Familial retinoblastoma
- Turcot syndrome (adenomatous polyposis syndrome)
- Lynch cancer family syndrome (nonpolyposis colorectal syndrome)
- Cowden disease
- Wermer syndrome
- Carney's complex

Information on the incidence of these syndromes is incomplete, because no population-based data are available. Moreover, data on genetic syndromes are not currently collected in any North American Association of Central Cancer Registries (NAACCR) data field. If a genetic syndrome is documented in the patient's health record, registrars should include this information in the narrative and document the presence of the syndrome in text fields.

Diagnostic Tools

Physical Examination

The first step in diagnosing a tumor in the brain or CNS is a neurological examination in which the physician evaluates eye movements, vision, hearing, reflexes, balance and coordination, the senses of smell and touch, abstract thinking, and memory. The results help to determine if a tumor is present and what clinical work-up is needed. Specific symptoms can also help identify the tumor location. One-sided hearing loss, for example, is a symptom associated with acoustic neuroma, a tumor of the acoustic nerve. Headache, muscle weakness, and seizures are symptoms of meningioma, a tumor of the cerebral or spinal meninges.

Radiologic Tests

- The first confirmation of CNS tumors is often a diagnosis by **computerized tomography** (CT) or **magnetic resonance imaging** (MRI) scan. CT scans use x-ray technology and a computer to view the intracranial and intervertebral structures and identify tumors. A CT scan can be used to identify the tumor location and type, as well as intracranial swelling or bleeding. MRI is used to identify CNS tumors utilizing magnetic fields and a computer. MRI provides a better picture of tumors that lie near bone.

- **Positron emission tomography** (PET), **single-photon emission computed tomography** (SPECT), and **magnetoencephalography** (MEG) scans are not usually used as diagnostic tools. PET is used to establish tumor grade, SPECT to determine if a tumor is low or high grade, and MEG to determine the function of the area of the brain that contains the tumor. All of these scans are used to help determine the best treatment course.

- **Angiography** may not be used in the diagnosis of CNS tumors, but it can help clinicians identify certain types of CNS tumors and make decisions before surgery. Dye is injected into blood vessels, and then x-rays identify the dye. Visualized blood vessels can help identify the tumor type or show if the tumor is near blood vessels that need to be avoided during surgery.

Laboratory Tests

- **Audiometry** is a hearing test that aids in the diagnosis of acoustic neuroma.

- For patients having seizures, an **electroencephalogram** (EEG) is performed to evaluate the electrical currents inside the brain. Abnormal currents can identify a brain tumor as the cause of the seizures.

- When an **endocrine evaluation** shows increased hormonal levels in blood and urine, a diagnosis of a tumor in the pituitary gland or hypothalamus is possible.

- The electrical activity of a nerve is evaluated through **evoked potentials testing,** which helps in the diagnosis of acoustic neuroma. Evoked potential testing can also be used to determine the role of specific nerves and to avoid damaging these nerves during surgery.

- **Lumbar puncture** is used to withdraw cerebrospinal fluid (CSF), which is examined for tumor cells and infection. Meningioma, lymphoma, and pineal gland tumors can be identified through the evaluation of CSF.

- A **myelogram** is the radiographic study of the spinal cord where a dye is injected into the spinal fluid.

- **Perimetry** is the quantification of the extent of the visual field for various types and intensities of stimuli using a perimeter apparatus. Manual and computerized machines are used, but both are based on the same principles.

Biopsies

A biopsy is not usually the first method used to diagnose CNS tumors. However, biopsy is used to identify the cell type and aid in determining of the best course of treatment. Two types of biopsies used are:

- **Needle biopsy:** a small burr hole is drilled into the skull, and the biopsy needle is inserted into the brain through the hole. Tissue is removed by the needle.

- **Stereotactic biopsy:** a computer is used to guide the needle to the tumor to extract tissue.

College of American Pathologists (CAP) Protocols

The College of American Pathologists has created site-specific protocols for pathologists to use when documenting pathologic information in patient health records. These protocols include a checklist as well as background documentation for the cytology or pathology report. The Commission on Cancer (CoC) of the American College of Surgeons requires that approved hospital cancer programs include a site-specific checklist for each pathology specimen in patient health records for cases diagnosed on or after January 1, 2004. The checklist for the brain/spinal cord includes macroscopic and microscopic categories. For more information, see the CAP Web site at http://www.cap.org/apps/docs/cancer_protocols/protocols_index.html.

Macroscopic Categories

- **Specimen type** indicates whether the specimen is from a biopsy or partial or total resection. Registrars should use this information when coding surgical procedure of the primary site.

- **Specimen size** indicates the greatest dimension of the specimen.

- **Tumor site** indicates different sites in the brain and CNS. The pathologist checks off the appropriate site. Registrars should use this information when determining the primary site.

- **Tumor size** indicates the tumor's largest dimension. Registrars should use this information when determining tumor size in the collaborative stage.

Microscopic Categories

- **Histologic type** indicates tumor morphologies commonly found in the brain and spinal cord, including nonmalignant and malignant morphologies. The appropriate morphology is checked. Registrars should use this information when assigning morphology codes.

- **Histologic grade** is used to indicate the WHO grade of the tumor. If another grading system is used, it can be recorded in narrative form by the pathologist. Registrars should use these data when recording WHO grade in a site-specific factor collaborative stage data field. This information should *not* be used to assign the *ICD-O-3* sixth digit.

- **Margins** indicate the postsurgical resection margins. Registrars should record this information in the surgical margins of primary site data field.

- **Additional studies** indicate additional pathologic findings. This information is not required by the CoC for accreditation.

- **Comments** are not required by the CoC for accreditation.

Treatment

Treatment options for brain tumors are driven by cell type, size, and location of the tumor as well as comorbid conditions and the overall health of the patient. For some patients with nonmalignant CNS tumors, the *first course* of treatment is *watchful waiting*. Over time, the patient might receive surgery, radiation therapy, or some type of systemic therapy. These treatments are then considered *subsequent treatment* and should not be coded in the abstract as first course treatment. Some hospital cancer registries record subsequent treatment on their abstracts, but most central registries do not collect these data. Health records should be reviewed carefully to delineate between first course and subsequent treatment. When the original treatment plan documents that treatment will be delayed for a specific period of time, once the treatment is given, it can be recorded as the first course treatment.

Patients with inoperable CNS tumors can be treated primarily with *radiation* or systemic therapies, such as *chemotherapy*, *immunotherapy*, or *hormone therapy*. Surgery performed to treat symptoms, such as the insertion of a shunt to reduce swelling, should not be coded as surgical treatment. However, such surgery can be coded as a *palliative procedure* by CoC-approved cancer programs.

Surgical Procedure of Primary Site

Two sets of site-specific surgery codes are used to identify the procedure used to remove, biopsy, or aspirate a CNS tumor at the primary site (Table 4).

Meninges, Brain, Spinal Cord, Cranial Nerves, or Other Parts of the CNS. These surgical codes are used when the primary site of surgery is the meninges (C70.0–C70.9), brain (C71.0–C71.9), spinal cord, cranial nerves, or other parts of the CNS (C72.0–C72.9):

- **Code 10** is assigned when a patient receives one of the listed treatments as the primary means of tumor destruction, and *no specimen is sent to pathology* from a surgical procedure. This code includes *laser surgery* (using light energy from a laser to destroy a tumor). Laser surgery can be assisted by *stereotactic surgery*, which uses a computer image to guide the surgeon to the tumor to be destroyed. Laser surgery can also be combined with photodynamic therapy. With photodynamic therapy, the patient ingests a photosensitive drug, and treated cancer cells are exposed to a laser light when most of the photosensitizing agent has left healthy cells but is still present in the cancer cells. The photosensitizing agent absorbs the light and produces an active form of oxygen that destroys the treated cancer cells. *Ultrasonic aspiration* uses ultrasonic waves to cause vibration, which breaks the tumor into small pieces. The pieces are then aspirated.

- **Code 20** is assigned when the most extensive surgery is a local excision or biopsy of the primary CNS tumor. A specimen is obtained and pathologically examined. For meninges, brain, and other CNS, an incisional biopsy can be coded as the surgical procedure of primary site. *Facility Oncology Registry Data Standards (FORDS)* changes, corrections, or clarifications in the wording for code 20 are found at the CoC Web site at http://www.facs.org/cancer/index.html.

- **Code 40** is assigned for a partial resection (part of the tumor is removed, and there is visible residual tumor remaining after resection or debulking).

- **Code 55** is assigned for a gross total resection (all of the tumor is removed with no macroscopic tumor remaining, but microscopic tumor can be present).

- **Code 90** is used for surgery, not otherwise specified.

All other sites. These surgical codes are used when the primary site of surgery is the pituitary gland (C75.1), craniopharyngeal duct (C75.2), or pineal gland (C75.3):

- **Codes 10–14** are used when tumor destruction is performed, but *no specimen is sent to pathology.* These codes apply to local tumor destruction, not otherwise specified, photodynamic therapy, electrocautery, cryosurgery, and laser surgery.

- **Codes 20–27** are used to code local tumor excision, and *a histologic specimen is sent to pathology.*

  - **Code 20** is used when the surgery is local tumor excision, not otherwise specified, with a pathologic specimen.

  - **Codes 21–24** are used for photodynamic therapy (PDT), electrocautery, cryosurgery, or laser ablation when they are *used alone* (without an excisional biopsy). This includes cases in which the tissue removed is *less than* an excisional biopsy (code 27), which is defined as removing all visible tumor. (*FORDS* changes, corrections, or clarifications are found at the CoC Web site at http://www.facs.org/cancer/index.html.)

  - **Code 25** is used for laser excision.

  - **Code 26** is used to code polypectomy; this procedure is not used for the intracranial glands.

  - **Code 27** is used for an excisional biopsy indicating that all visible tumor was removed; however, microscopic tumor might remain. Code 27 is used when any of codes 21–24 are used with *local tumor excision, not otherwise specified or excisional biopsy.* Codes 25–27 are also local tumor excision codes and are only used when a pathologic specimen is collected.

- **Code 30**, simple or partial surgical removal of the primary site, indicates that part of the gland was removed.

- **Code 40**, total surgical removal of the primary site, is used if the complete gland is resected.

- **Code 50** should be used when the surgery is described as debulking; however, debulking is not commonly used with intracranial glands.

- **Code 60** is for radical surgery, in which all or part of the primary site was removed with a resection of other organs. This procedure also is unusual surgery for intracranial glands.

- **Code 90**, surgery, not otherwise specified, is used when the surgery was conducted at another facility and no additional information is available.

Other surgical data fields should be completed for CNS tumors just as they are for other malignant primary sites.

Table 4. Codes for Surgical Procedures of Primary Site for CNS Tumors

Meninges, Brain, Spinal Cord, Cranial Nerves, or Other Parts of the CNS

10	Tumor destruction, not otherwise specified; no specimen is sent to pathology from surgical procedure. Includes: • Laser surgery • Laser surgery with photodynamic therapy • Ultrasonic aspirator.
20	Local excision (biopsy) of tumor, lesion, or mass; specimen sent to pathology from surgical event
40	Partial resection
55	Gross total resection
90	Surgery, not otherwise specified

All Other Sites: Includes Pituitary Gland, Craniopharyngeal Duct, or Pineal Gland

10	Local tumor destruction, not otherwise specified
11	Photodynamic therapy
12	Electrocautery; fulguration
13	Cryosurgery
14	Laser
20	Local tumor excision, not otherwise specified
21	Photodynamic therapy
22	Electrocautery
23	Cryosurgery
24	Laser ablation
25	Laser excision; specimen sent to pathology from surgical event 20–27
26	Polypectomy
27	Excisional biopsy (used when any of codes 21–24 are used with code 20, 26, or 27)
30	Simple or partial surgical removal of primary site
40	Total surgical removal of primary site; enucleation
50	Surgery stated to be “debulking”
60	Radical surgery; partial or total removal of the primary site with resection in continuity (partial or total removal) with other organs
90	Surgery, not otherwise specified

Surgical Margins of the Primary Site

This data item codes the final status of surgical margins. It is a CoC-required data item and serves as a quality control measure for pathology reports. The status of this field can be a prognostic factor in recurrence. (Directions for coding the surgical margins of the primary site data field are found in the *FORDS* manual, section 2.)[6] The final status of the margins should be coded after the tumor is removed.

Scope of Regional Lymph Node Surgery

These codes are used to identify the removal, biopsy, or aspiration of regional lymph nodes. This is a required data item for the National Program of Cancer Registries (NPCR), CoC, and SEER. (Directions for coding this data field can be found in the *FORDS* manual, section 2.)[6] For tumors of the meninges (C70.0–C70.9); brain (C71.0–C71.9); and spinal cord, cranial nerves, and other parts of the CNS (C72.0–C72.9), registrars should use always use code 9 (unknown or not applicable), because there are no regional lymph nodes for these sites.

Radiation Therapy

Radiation therapy is used to treat nonmalignant and malignant CNS tumors. There are several fields that record information about radiation therapy, including several data fields. They include the following:

- Location of radiation treatment
- Radiation treatment volume
- Regional treatment modality
- Regional dose
- Boost treatment modality
- Boost dose
- Number of treatments
- Radiation/surgery sequence
- Reason no radiation given.

CoC-approved cancer programs collect more specific radiation therapy data, and most of these fields are required by the CoC. However, only the regional treatment modality field is collected by SEER and NPCR. Codes in the radiation treatment volume field describe the anatomical structures targeted by regional radiation therapy. The reason no radiation was given is recorded in a separate field, unlike other modalities where the codes for no treatment are included with the other treatment codes.

The codes for the regional treatment modality field are used to indicate the type of radiation therapy performed as part of the first course of treatment (Table 5). They record the modality of radiation therapy used to deliver a significant regional dose to the primary volume of interest.

Some central cancer registries collect data on radiation therapy. SEER collects data on regional treatment modality from CoC-approved facilities only. The NPCR has recommended that such data be collected for cases diagnosed through 2005 and will require the collection of such data for cases diagnosed in 2006 forward.

Here are the types of radiation therapy used to treat CNS tumors:

Beam Radiation

The general category of beam radiation includes several types of radiation therapy. Beam radiation is most often fractionated, meaning that doses of radiation are delivered over a specified time period.

- **Codes 20–29** are assigned when conventional radiation therapy is delivered by *external beam* to the tumor. The radiation is delivered by *orthovoltage* (code 21), *cobalt* (code 22), or *linear accelerator* (codes 23–29). Orthovoltage and cobalt are old technologies that are rarely used to administer conventional radiation therapy to CNS sites. Linear accelerators deliver radiation using photon or electron energy. The amount of photon energy delivered determines the code. *Photon energy* is assigned codes 23–27. Electron energy is assigned code 28, and *mixed photon and electron energy* is assigned code 29.

- **Code 30** is used for another type of particle radiation that uses a beam of high-energy neutrons. *Neutrons* are electrically neutral particles that are part of all atoms. Unlike x-rays, gamma rays, and proton rays, neutrons disrupt atomic nuclei rather than electrons; the likelihood of cells repairing this kind of damage is very small. Neutron therapy can also more effectively treat larger tumors than conventional radiation therapy. Modern neutron machines and 3-dimensional treatment planning systems are now available in a few institutions and could further reduce the side effects of neutron radiation therapy. Another type of neutron therapy is *boron neutron capture therapy (BNCT)*. With BNCT, the patient ingests a boron compound that concentrates in the tumor cells. External beam radiation is delivered by using neutron energy. The boron concentrated in the tumor cells captures the neutron energy and destroys the tumor cells without damaging normal tissues. BNCT is not yet widely available.

- **Code 31** is used for *intensity-modulated radiation therapy (IMRT)*, a type of conformal radiation. A linear accelerator delivers energy in three dimensional beams that conform to the shape of the tumor. With IMRT, the intensity of the beams change as radiation is delivered allowing for short bursts of high intensity energy without damaging normal tissues. Registrars should use this code only when IMRT is documented in the health record.

- **Code 32** is used for *conformal radiation*, which also uses 3-dimensional beams of energy from a linear accelerator that conform to the shape of the tumor. But with conformal radiation, the intensity of the radiation delivered does not change. Conformal radiation allows for the delivery of more intense radiation without damaging normal tissues. Registrars should use this code only when conformal radiation is documented in the health record.

- **Code 40** is used for *particle or proton beam radiosurgery.* A cyclotron, an adapted nuclear reactor, produces proton particle beams that conform to the shape of the tumor. Particle beam radiosurgery is used for deep-seated tumors such as tumors in the pituitary gland. The treatment is usually fractionated. The use of this technology is growing but not yet widespread.

- **Code 41** is used when the health record documents that the patient received *stereotactic radiosurgery*, but the specific type of radiosurgery is not documented. Stereotactic radiosurgery is used for patients with localized intracranial tumors that are difficult to access through conventional surgery. Stereotactic radiosurgery is radiation therapy (not surgery) that is given focally in high doses with the use of a computer. Blocks are used to ensure that the radiation is delivered only to the tumor and not to surrounding normal tissues. There are several types of stereotactic radiosurgery. Treatment can be delivered in a single session or be fractionated.

- **Code 42** is used for *linac radiosurgery*, which uses an adapted linear accelerator to deliver fractionated doses of radiation to the tumor. The beams are adjusted to the shape of the tumor.

- **Code 43** is used for *gamma knife radiosurgery*, which delivers focal radiation adjusted to the tumor shape by using cobalt in a single dose. Because the beams are adjusted to deliver radiation only to the tumor, little or no damage is done to normal tissues surrounding the tumor.

Stereotactic radiosurgery is used to treat acoustic neuromas, craniopharyngiomas, chordomas, hemangioblastomas, pineal tumors, pituitary adenomas, glial tumors, and astrocytoma. These codes are to be used only if stereotactic radiosurgery is the first course of treatment. When radiosurgery is given as subsequent treatment, the procedure should not be coded as part of first course therapy.

Radioactive Implants

Radioactive implants are radiation sources placed directly into the tumor. They are used to treat small tumors and are considered local therapy. For some patients, more surgery is performed after radioactive implants are used to remove the dead tumor cells. Radioactive implants are often a boost modality after regional radiation with external beam. For CNS tumors, radioactive implants are often adjuvant therapy. The radioactive implants are placed in the tumor bed after partial or complete tumor resection.

- **Code 50** is used if therapy is described as *brachytherapy, not otherwise specified*. This includes radiation implants, radiation seeding, radioactive implants, interstitial implants, or intracavitary radiation and not otherwise specified.

- **Code 51** is used if the implants are *intracavitary*, or in a cavity with no direct insertion into tissues, and the application is *low-dose rate* such as with cesium-137 or a Fletcher applicator.

- **Code 52** is also for *intracavitary* implants, but the application is *high-dose rate*.

- **Code 53** is assigned for *low-dose rate interstitial* radiotherapy. Interstitial indicates that the implant is placed in tissue.

- **Code 54** is assigned for *high-dose rate interstitial* radiotherapy.

- **Code 55** is assigned when *radium*, a low-dose interstitial or intracavitary source, is implanted. This is currently used infrequently.

- **Codes 60–62** are used for *radioisotopes*. Code 60 is for radioisotopes, not otherwise specified and includes iodine-131 used for thyroid malignancies and phosphorus-32 used for metastatic bone lesions. Codes 61 and 62 are used for strontium 89 and 90. Strontium 89 is also used to treat metastatic bone disease.

Chemotherapy

These codes are used to record the type of chemotherapy administered as a first course of treatment (Table 5; a full listing of the codes for chemotherapy can be found in the *FORDS* manual, section 2).[6] Chemotherapy can be administered as only one drug (single-agent), or a combination of drugs (multi-agent). A change in the drugs administered can indicate a change in treatment course, and the new drugs might not be the first course of treatment. In such cases, the new drugs

Table 5. Codes for Radiation, Chemotherapy, Hematologic Transplants, and Endocrine Procedures as First Course of Treatment for CNS Tumors*

Radiation Therapy Modality

Code	Description
20	External beam, not otherwise specified
21	Orthovoltage
22	Cobalt; Cesium-137
23–27	Photons
28	Electrons
29	Mixed photon and electron energy
30	Neutrons with or without photons/electrons
31	Intensity-modulated radiation therapy (IMRT)
32	Conformal or 3-D radiation
40	Protons
41	Stereotactic radiosurgery, not otherwise specified
42	Linac radiosurgery
43	Gamma knife radiosurgery
50	Brachytherapy, not otherwise specified
51	Brachytherapy, Intracavitary, low-dose rate
52	Brachytherapy, Intracavitary, high-dose rate
53	Brachytherapy, Interstitial, low-dose rate
54	Brachytherapy, Interstitial, high-dose rate
55	Radium
60	Radioisotopes, not otherwise specified
61	Strontium-89
62	Strontium-90

Chemotherapy

01	Chemotherapy, type and number of agents unknown
02	Single-agent chemotherapy
03	Multi-agent chemotherapy
82–88	Reason no chemotherapy was given

Hematologic Transplants and Endocrine Procedures

10	Bone marrow transplant, type not specified
11	Autologous bone marrow transplant
12	Allogeneic bone marrow transplant
20	Stem cell harvest
30	Endocrine therapy or endocrine radiation therapy
40	Combination endocrine surgery or radiation therapy with a transplant procedure
82–88	Reason no procedures were performed

* For the complete listing of codes, see the *FORDS* manual, section 2: First Course Treatment.[6]

should not be coded in this data field as the first course of treatment. Moreover, chemotherapy for malignant CNS tumors can be administered in combination with ancillary drugs. These ancillary drugs should not be coded as part of the chemotherapy. Several codes are used to describe the reasons chemotherapy was not given.

An obstacle in treating CNS tumors with standard intravenous chemotherapy is the presence of the *blood-brain barrier*. The blood-brain barrier protects the brain from foreign substances that cause infection and functional problems. Tumor cells in the brain are also protected when chemotherapy drugs cannot infiltrate the blood-brain barrier. Synthetic substances called *receptor-mediated permeabilizers* are administered to temporarily open the blood-brain barrier and allow chemotherapy drugs into the brain. The receptor-mediated permeabilizers should not be coded as chemotherapy.

There are two methods of administering chemotherapy to CNS tumors that avoid the blood-brain barrier (note that the method of administration is not included in the chemotherapy codes):

- **Intrathecal** administration of chemotherapy avoids the blood-brain barrier because the drugs are injected directly into CSF. The injection is given in the lower part of the spinal column. Another method of intrathecal administration is through a catheter called an Ommaya reservoir. The Ommaya reservoir is placed on the scalp, chemotherapy drugs are injected into the catheter, and the drugs find their way to CSF. The Ommaya reservoir is often used to administer chemotherapy to children.

- **Interstitial** chemotherapy is administered directly to tissues involved with the tumor. Polymer wafers soaked in a chemotherapeutic agent are inserted in the tumor bed after tumor resection. This method of administration avoids the blood-brain barrier and keeps the medication from affecting normal body tissues.

Hormone Therapy

The codes for hormone therapy are used to record systemic hormonal agents administered as a first course of treatment. (The codes for hormone therapy are in the FORDS manual, section 2.)[6] Hormone therapy may be used to treat nonmalignant CNS tumors, but in most cases, the treatment is for a tumor recurrence. Therefore, registrars should carefully review the patient's records to determine if the treatment was given as first course therapy or to treat tumor recurrence. The codes contain the reason no hormonal therapy was given. Two types of hormonal therapy that are used for CNS tumors are *tamoxifen* and *mifepristone* (formerly known as *RU-486*) that can be used to treat meningiomas.

Steroids can be administered as a first course of treatment for some primary sites but not for CNS tumors. However, steroids are administered to treat intracranial swelling caused by a CNS tumor. This should be coded as a palliative procedure, not as hormone therapy.

Immunotherapy

The codes for immunotherapy are used to record immunotherapeutic agents that are administered as a first course of treatment. (The codes for immunotherapy are in the *FORDS* manual, section 2.)[6] The use of immunotherapy to treat nonmalignant CNS tumors is increasing, but it is not usually a first course of treatment. With immunotherapy, also known as biologic response modifiers, the body's immune system is used to fight cancer by changing the biologic response to the tumor. Codes contain the reason no immunotherapy was given. Examples of immunotherapy used for CNS tumors are angiogenesis inhibitors, interleukins, gene therapy, and tumor vaccines.

- **Angiogenesis inhibitors** block the development of blood vessels. Without new blood vessels, tumors lose their blood supply and starve. The drug thalidomide and interferons that occur naturally in the body are angiogenesis inhibitors.

- **Interleukins** are growth factors. When used for immunotherapy, they manipulate the tumor's ability to grow.

- **Gene therapy** is the treatment of disease either by replacing damaged or abnormal genes with normal ones or by providing new genetic instructions to help fight disease.

- **Specific immunotherapy** or the use of *tumor vaccines* involves immunization to boost the cancer patient's immune response specifically against his or her own tumor. This type of immunotherapy relies on the presence of tumor-associated antigens on the surface of the malignant cells and the ability of those antigens to produce a host immune response.

Hematologic Transplants and Endocrine Procedures

These codes identify hematologic transplant and endocrine procedures administered as a first course of treatment. Registrars should carefully review the patient's health records to determine if the hematologic transplant procedure was given as a first course of treatment.

Hematologic transplant procedures are only used to treat malignant CNS tumors in children. Children with malignant brain tumors, neuroblastoma, or lymphoma can be treated with bone marrow transplant or peripheral blood stem cell transplant. These children receive high-dose chemotherapy or radiation therapy to destroy the tumor cells, but the therapy also destroys the bone marrow and stem cells. The marrow or stem cell transplant is given following the chemotherapy or radiation therapy to replace the destroyed bone marrow or stem cells.

- **Code 10** is used for bone marrow transplant, not otherwise specified.
- **Code 11** is used for documented *autologous* bone marrow transplant. Autologous means the donated marrow came from the patient.
- **Code 12** is used for documented *allogeneic* bone marrow transplant. Allogeneic means the marrow was donated by a person other than the patient.
- **Code 20** is assigned when *peripheral blood stem cell transplantation* or *stem cell harvest* is performed.
- **Code 30** is used when either *surgery or radiation therapy* is used as endocrine therapy. An example would be an orchiectomy for prostate cancer.
- **Code 40** is used when *endocrine surgery or radiation therapy* is used in conjunction *with a transplant* procedure.
- **Codes 82–88** record the reason no procedures were performed**.**

Data Edits

Changes to the data edits were needed to allow for the collection of nonmalignant behavior codes and other rule changes. The NAACCR Edits Committee was responsible for making the needed changes. These data edit changes were needed so that data on nonmalignant CNS tumors can be collected by central and hospital cancer registries that were not already collecting these data before January 1, 2004. In addition, commercial and private software vendors needed to incorporate these changes in their cancer registry software.

A few new edits were added, but other needed changes affected over 30 standard edits. Examples of changes were allowance of behavior codes of 0 and 1, collection of laterality data for CNS sites, and new site/histology combinations(see Appendix D, the *ICD-O-3* Primary Brain and CNS Site and Histology Listing, which lists acceptable site and morphology combinations).

Data Analysis

Currently, no policy exists on how nonmalignant and malignant CNS tumors should be reported and analyzed. However, the NAACCR Registry Operations Subcommittee recommended that nonmalignant CNS tumors be reported and analyzed separately from malignant tumors in general reports. The subcommittee also included a footnote advising that pilocytic astrocytomas be included in analysis for malignant brain tumors for continuity of trends.

Researchers, clinicians, and epidemiologists have special data needs that also should be considered. Recommendations include:

- For special data reports, registrars should determine if the data to be analyzed should include CNS tumors, regardless of their behavior. The clinician or epidemiologist might want to study CNS tumors of all behaviors.

- Historically, cancer registries have displayed lymphoma data separately regardless of the site of origin. For some studies, researchers may want CNS lymphoma data to be included with other CNS data.

- Registries do not usually analyze olfactory tumors of the nasal cavity with CNS tumors. Yet some researchers want these data included with the CNS tumors. Registrars should work closely with the researchers to ensure that the data needed for the study are included.

- The tumor sites and histologies that are to be included in the analysis should be documented clearly.

- A discussion of cancer registry multiple primary rules could also be useful, especially the histology groupings.

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Part IV Exercises
Coding Collaborative Stage and Treatment

For these exercises, assume that if treatment is not mentioned, it was not administered.

1. **The patient was referred to a neurologist because of a right-side hearing loss and possible acoustic tumor. A CT scan on March 1, 2004 was used to diagnose right acoustic neuroma. On March 31, 2004 the patient had a craniotomy. A protrusion of tumor in the vestibule appeared to come from the cochlea. Facial nerve was spared. The tumor was totally resected, with clear margins.**

   CS Extension code
   Surgical procedure of primary site
   Surgical margins of primary site
   Scope of regional lymph node surgery
   Radiation treatment volume
   Regional treatment modality (radiation)
   Chemotherapy
   Hormone therapy
   Immunotherapy
   Hematologic transplant and
   endocrine procedures

2. **A 6-year-old boy had severe headaches and vomiting. An MRI on September 13, 2004 diagnosed medulloblastoma of the cerebellum. An infratentorial craniotomy was performed on September 28, 2004, and the tumor was removed. Macroscopic and microscopic residual medulloblastoma remained. On November 1, 2004 the patient began a course of carmustine. After completion of chemotherapy, the patient had a bone marrow transplant with donor marrow from his older brother.**

   CS Extension code
   Surgical procedure of primary site
   Surgical margins of primary site
   Scope of regional lymph node surgery
   Radiation treatment volume
   Regional treatment modality (radiation)
   Chemotherapy
   Hormone therapy
   Immunotherapy
   Hematologic transplant and
   endocrine procedures

3. **Patient had CT scan of the head on February 24, 2004 showing a large prolactinoma of the pituitary gland. The patient started on bromocriptine to shrink the tumor in March 2004. The patient had gamma knife radiosurgery on October 1, 2004.**

   CS Extension code
   Surgical procedure of primary site
   Surgical margins of primary site
   Scope of regional lymph node surgery
   Radiation treatment volume
   Regional treatment modality (radiation)
   Chemotherapy
   Hormone therapy
   Immunotherapy
   Hematologic transplant and
   endocrine procedures

4. **The patient was referred to a neurologist after reporting symptoms of vomiting, muscle weakness on one side of the face, and several episodes of slurred speech. The patient had an MRI on June 3, 2004, that showed a glioma in the brain stem. Through an infratentorial craniotomy, the tumor was removed on June 30, 2004. The pathology report documented microscopic residual subependymal glioma in the fourth ventricle. On August 1, 2004, the patient had conformal radiation to the fourth ventricle.**

   CS Extension code
   Surgical procedure of primary site
   Surgical margins of primary site
   Scope of regional lymph node surgery
   Radiation treatment volume
   Regional treatment modality (radiation)
   Chemotherapy
   Hormone therapy
   Immunotherapy
   Hematologic transplant and
   endocrine procedures

5. **The patient's symptoms included headaches, double vision, vomiting, and drowsiness. A CT scan on November 1, 2004, showed a growth in the pineal gland. The patient had a biopsy of the pineal gland on November 15, 2004. The tumor pathology was pineocytoma. The patient began beam radiation to the pineal gland on December 8, 2004.**

   CS Extension code
   Surgical procedure of primary site
   Surgical margins of primary site
   Scope of regional lymph node surgery
   Radiation treatment volume
   Regional treatment modality (radiation)
   Chemotherapy
   Hormone therapy
   Immunotherapy
   Hematologic transplant and
   endocrine procedures

Helpful Resources

National Program of Cancer Registries Training Materials

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Helpful Resources

Manuals, Articles, and Reports

- American Brain Tumor Association. A primer of brain tumors. Des Plaines, Illinois: American Brain Tumor Association, 1998. Available at: http://www.abta.org.

- Brenner AV, Linet MS, Selker RG, et al. Polio vaccination and risk of brain tumors in adults: no apparent association. *Cancer Epidemiol Biomark Prev.* 2003;12(2):177-8.

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- American College of Surgeons, Commission on Cancer Information, *Facility Oncology Registry Data Standards (FORDS)*
  http://www.facs.org/cancer/index.html

- American Joint Committee on Cancer, Collaborative Stage Documentation
  http://www.cancerstaging.org

- Brain and Neurosurgery Information Center
  http://www.brain-surgery.com/index.html

- Brain and Spinal Cord Tumors—Hope through Research
  http://www.ninds.nih.gov/health_and_medical/pubs/brain_tumor_hope_through_research.htm

- Brain Tumor Guide
  http://virtualtrials.com/faq/toc.cfm

- Central Brain Tumor Registry of the United States
  http://www.cbtrus.org/page2t.htm

- College of American Pathologists (CAP), Protocol—Brain
  http://www.cap.org/apps/docs/cancer_protocols/Brain04_pw.doc

- Illustrated Glossary of Radiology: Anatomy, Examinations and Procedures; Department of Radiology and Radiological Services, The Uniformed Services University of the Health Sciences
  http://rad.usuhs.mil/glossary.html

- International RadioSurgery Association
  http://www.isra.org

- National Brain Tumor Radiosurgery Association
  http://www.med.jhu.edu/radiosurgery/nbtra

- NCI Brain Tumor Home Page
  http://www.cancer.gov/cancerinformation/cancertype/braintumor

- PDQ Cancer Information Summaries: Adult Treatment
  http://www.cancer.gov/cancerinfo/pdq/adulttreatment

- PDQ Cancer Information Summaries: Pediatric Treatment
  http://www.cancer.gov/cancerinfo/pdq/pediatrictreatment

- The Brain Tumor Foundation
  http://www.braintumorfoundation.org

Additional Terms Associated with Intracranial and Central Nervous System (CNS) Tumors

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Additional Terms Associated with Intracranial and Central Nervous System Tumors

ABTA: American Brain Tumor Association. ABTA is a not-for-profit organization dedicated to the elimination of brain tumors through research and patient education services. ABTA is the oldest organization furthering this effort, begun in 1973 by two mothers struggling to understand brain tumors. Since then, ABTA has funded over $2 million in research awards to the most prestigious institutions in the United States.[1]

ACoS: American College of Surgeons.

ANGIOGENESIS: The growth of new blood vessels from surrounding tissue into growing tissue.[2]

CAUDA EQUINA: The group of spinal roots that descends from the inferior portion of the spinal cord (literally "horse's tail").[3]

CDC: Centers for Disease Control and Prevention.

CEREBELLOPONTINE ANGLE: The angle between the cerebellum and the pons—a common site for the growth of acoustic neuromas.[2]

CoC: Commission on Cancer. Established by the American College of Surgeons (ACoS) in 1922, the multi-disciplinary CoC sets standards for quality multidisciplinary cancer care delivered primarily in hospital settings; surveys hospitals to assess compliance with those standards; collects standardized and quality data from approved hospitals to measure treatment patterns and outcomes; and uses the data to evaluate hospital provider performance and develop effective educational interventions to improve cancer care outcomes at the national and local level.[4]

CORPUS COLLOSUM: Literally "hard body." A large bundle of white matter, found in the longitudinal fissure, forming a "commissure" by interconnecting the two cerebral hemispheres.[3]

CORTEX: The outer layer of a body or organ structure. From the Latin word for "bark."[3]

CRANIECTOMY: Surgery performed on the skull where pieces of bone are removed to gain access to the brain, and the bone pieces are not replaced.[2]

CRANIOTOMY: Surgery performed on the skull where a portion of bone is removed to gain access to the brain, and the bone is put back in place.[2]

CAT SCAN: Computerized Axial Tomography. An X-ray device linked to a computer that produces an image of a predetermined cross-section of the brain. A special dye material may be injected into the patient's vein prior to the scan to help make any abnormal tissue more evident.[2]

EDEMA: Swelling due to an excess of water.[2]

EPIDEMIOLOGY: The study of the distribution of disease and its impact upon a population, using such measures as incidence, prevalence, or mortality.[2]

EXTRACEREBRAL: Located outside the cerebral hemispheres.[2]

EXTRADURAL: External (outside) to the dura mater.[2]

GFAP: Glial Fibrillary Acidic Protein. This protein, found in microfilaments of glial cells, helps distinguish glial from non-glial tumors. A laboratory stain is used to test for its presence.[2]

GLUCOCORTICOSTEROIDS: Medications used to decrease swelling around tumors.[2]

HYPERTHERMIA: The use of heat to kill tumor cells.[2]

HYPOPHYSIS: Pituitary gland.[2]

INTRACEREBRAL: Located within the cerebral hemispheres (cerebrum).[2]

INTRACRANIAL: Within the skull.[2]

INTRADURAL: Beneath the dura mater.[2]

INTRAVENOUS: Injection into a vein.[2]

INTRAVENTRICULAR: Injection into a ventricle.[2]

LASER: An acronym of light amplification by stimulated emission of radiation. A surgical tool that creates intense heat and power when focused at close range, destroying cells by vaporizing them.[2]

NPCR: National Program of Cancer Registries. The Centers for Disease Control and Prevention (CDC) has administered the NPCR since 1994. This program is currently helping states and U.S. territories to improve their cancer registries; meet standards for data completeness, timeliness, and quality; use cancer data to support cancer prevention and control programs; train registry personnel; establish computerized reporting and data-processing systems; and develop laws and regulations that strengthen registry operations.[5]

PHOTODYNAMIC RADIATION THERAPY: A light sensitive drug is given through a vein and concentrates in the tumor. During a surgical procedure, a special light activates the drug, which kills tumor cells.[2]

QUADRIGEMINAL PLATE AND CISTERN: The posterior part of the brainstem at the mesencephalon (midbrain) has four knobby bits: two superior colliculi and two slightly smaller inferior colliculi. The enlarged subarachnoid space posterior is called the Quadrigeminal Plate and Cistern (QP or Kewpee Cistern), and is contiguous with the ambient (circum-mesencephalic) cistern. The QP cistern looks like a smile. During brain herniation, the smile becomes crooked or disappears entirely, as the brainstem shifts and the subarachnoid space is obliterated.[2]

RECURRENCE: The return of symptoms or the tumor itself, as opposed to a remission.

SEER PROGRAM: The Surveillance, Epidemiology, and End Results (SEER) Program of the National Cancer Institute is an authoritative source of information on cancer incidence and survival in the United States. Case ascertainment for SEER began on January 1, 1973, in the states of Connecticut, Iowa, New Mexico, Utah, and Hawaii and the metropolitan areas of Detroit and San Francisco-Oakland. In 1974–1975, the metropolitan area of Atlanta and the 13-county Seattle-Puget Sound area were added. In 1978, 10 predominantly black rural counties in Georgia were added, followed in 1980 by the addition of American Indians residing in Arizona. Three additional geographic areas participated in the SEER program prior to 1990: New Orleans, Louisiana (1974–1977, rejoined 2001); New Jersey (1979–1989, rejoined 2001); and Puerto Rico (1973–1989). The National Cancer Institute also began funding a cancer registry that, with technical assistance from SEER, collects information on cancer cases among Alaska Native populations residing in Alaska. In 1992, the SEER Program was expanded to increase coverage of minority populations, especially Hispanics, by adding Los Angeles County and four counties in the San Jose-Monterey area south of San Francisco. In 2001, the SEER Program expanded coverage to include Kentucky and Greater California, and New Jersey and Louisiana once again became participants.

The SEER Program currently collects and publishes cancer incidence and survival data from 11 population-based cancer registries and three supplemental registries covering approximately 14% of the U.S. population. The expansion registries increase the coverage to approximately 26%. Information on more than 3 million *in situ* and invasive cancer cases is included in the SEER database, and approximately 170,000 new cases are accessioned each year within the SEER areas. The SEER Registries routinely collect data on patient demographics, primary tumor site, morphology, stage at diagnosis, first course of treatment, and follow-up for vital status. The SEER Program is the only comprehensive source of population-based information in the United States that includes stage of cancer at the time of diagnosis and survival rates within each stage. The mortality data reported by SEER are provided by the National Center for Health Statistics.[6]

SHUNT: A drainage system. Spinal fluid flows from a ventricle into a body cavity via a tube. Used to relieve increased intracranial pressure caused by brain tumors that block the flow of spinal fluid.[2]

TRIGONE (Lateral ventricle): The triangular area between the temporal and occipital horns at the junction with the body of the lateral ventricle.[2]

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3. The Department of Radiology and Radiological Services. *Illustrated Glossary of Radiology, Anatomy, Examinations, and Procedures*. Uniformed Services University of the Health Sciences. Available at http://rad.usuhs.mil/glossary.html.

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Answers

National Program of Cancer Registries Training Materials

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Part II Answers
Reportability, Coding Site, and Histology

For these exercises, assume that the patient had no previous nonmalignant or malignant tumors of other sites.

Assign ICD-O-3 codes for site and histology even if the case is not reportable.

1. The patient was seen in the hospital neurology clinic on March 4, 2004 and was prescribed tamoxifen for cerebral meningioma. The patient was first diagnosed with cerebral meningioma in December 2001.

   Reportable: **No**
   Primary site: **C70.0, cerebral meninges**
   Histology: **9530/09, meningioma, NOS**

   Rationale: The original diagnosis was before nonmalignant tumors were required to be reported nationally. (Local reporting rules should be considered separately.)

2. The patient was diagnosed on April 15, 2004 with a chondroma originating in the skull.

   Reportable: **No**
   Primary site: **C41.0, skull**
   Histology: **9220/09, chondroma, NOS**

   Rationale: In this scenario, the primary is stated to have originated in the skull. Benign tumors of bone are not reportable.

   Chondroma is a rare, benign tumor that tends to arise at the base of the skull, especially in the area near the pituitary gland. The chondroma is composed of cartilage formed by the meninges and is usually attached to the dura mater, the outermost layer of the meninges. It can grow to a large size and can occur as single or multiple tumors. (From the American Brain Tumor Association website, www.abta.org.)

3. The patient was diagnosed on December 1, 2004 with a chordoma of the right frontal lobe extending into the skull.

   Reportable: **Yes**
   Primary site: **C71.1, frontal lobe**
   Histology: **9370/39, chordoma, NOS**

   Rationale: Chordoma is a malignant tumor. Reportability requirements for malignant tumors have not changed.

4. On February 2, 2002, the patient was diagnosed with low-grade astrocytoma of the cerebellum, Kernohan grade 2.

   Reportable: **Yes**
   Primary site: **C71.6, cerebellum**
   Histology: **9400/32, astrocytoma, low grade**

   Rationale: The case is reportable because low-grade astrocytoma is a malignant tumor, and reportability requirements have not changed for malignant tumors. The sixth digit of the histology code is 2 because the histology is

low-grade astrocytoma. The reference is *ICD-O-3*, page 39, second paragraph: "If the *ICD-O* sixth digit grade or differentiation code is to be used for central nervous system tumors, coders should give preference to terms from the diagnosis, such as low grade or anaplastic, rather than use the reported WHO grade." Low grade is code 2, and the definition is found in *FORDS*, page 97. Kernohan grade is not coded as part of the histology.

5. The patient had an intracranial biopsy on July 1, 2004, and the tumor pathology was WHO grade I schwannoma.

   Reportable: **Yes**
   Primary site: **C72.5, cranial nerves, NOS**
   Histology: **9560/09, schwannoma, NOS**

   Rationale: Nonmalignant intracranial tumors are reportable for cases diagnosed on January 1, 2004, or later. Intracranial schwannoma with no specific site identified is coded to cranial nerves, NOS. The reference is *ICD-O-3*, page 24, Rule A : "If the diagnosis does not specify the tissue of origin, code the appropriate tissues suggested in the alphabetic index for each ill-defined site in preference to the 'NOS' category." Schwannoma arises from the nerve sheath and consists of Schwann cells in a collagenous matrix.

   The grade for all benign and borderline tumors is 9 (unknown, not applicable). The reference is *ICD-O-3*, page 30, Rule G, paragraph 1: "Only malignant tumors are graded." WHO grade is not coded as part of the histology, but it is coded in a collaborative stage site-specific factor.

6. The final pathologic diagnosis for a procedure performed on January 2, 2004 was well-differentiated pituitary adenoma.

   Reportable: **Yes**
   Primary site: **C75.1, pituitary gland**
   Histology: **8272/09, pituitary adenoma, NOS**

   Rationale: Well-differentiated pituitary adenoma is an intracranial nonmalignant tumor diagnosed after January 1, 2004. The sixth digit of the histology code is 9, even though the tumor is described as well-differentiated, because the grade code for all nonmalignant tumors is 9. The reference is *ICD-O-3*, page 30, Rule G, paragraph 1: "Only malignant tumors are graded."

7. The patient had hearing loss on the right side first documented in 2002. In August 2002, a computerized tomography (CT) scan showed acoustic neuroma, but no treatment was given. On July 25, 2004, the patient had surgical resection of an intracranial tumor. The final pathologic diagnosis was right acoustic neuroma.

   Reportable: **No**
   Primary site: **C72.4, acoustic nerve**
   Histology: **9560/09, acoustic neuroma**

   Rationale: Acoustic neuroma is a nonmalignant tumor and was diagnosed in August 2002, before nonmalignant tumors were required to be reported nationally. (Local reporting rules should be considered separately.)

8. A CT scan in May 2004 identified a lesion in the cerebral meninges. A biopsy of the lesion was used to diagnose cholesteatoma.

   Reportable: **No**
   Primary site: **C70.0, cerebral meninges**
   Histology: **Not applicable**

   Rationale: No histology code exists for cholesteatoma in *ICD-O-3*. Nonmalignant intracranial and CNS reporting requirements include any primary tumor histology with a code defined in *ICD-O-3*.

9. Magnetic Resonance Imaging (MRI) was used to identify a pinealoma on February 20, 2004. The patient had gamma knife radiosurgery on March 15, 2004.

   Reportable: **Yes**
   Primary site: **C75.3, pineal gland**
   Histology: **9360/19, pinealoma**

   Rationale: Pinealoma is a nonmalignant tumor. The site is coded to the pineal gland. The reference is *ICD-O-3*, page 32, Rule H: "Use the topography code provided when a topographic site is not stated in the diagnosis. This topography code should be disregarded if the tumor is known to arise at another site."

10. A CT scan identified a nonglial tumor in the temporal lobe on October 1, 2004. The tumor was removed and final pathologic diagnosis was meningioma of the left temporal dura.

    Reportable: **Yes**
    Primary site: **C70.0, cerebral meninges**
    Histology: **9530/09, meningioma, NOS**

    Rationale: Meningioma, unless stated to be malignant, is a nonmalignant tumor, and this case is reportable, because it was diagnosed after January 1, 2004. The site is assigned to cerebral meninges because meningioma is a tumor of the meninges covering the brain, not of the temporal lobe itself. The reference is *ICD-O-3*, page 32, Rule H: "Use the topography code provided when a topographic site is not stated in the diagnosis. This topography code should be disregarded if the tumor is known to arise at another site."

Part III Answers

Multiple Primaries, Diagnosis Date, Sequence Number, Laterality, Collaborative Stage

For these exercises, assume that the patient had no previous benign or malignant tumors of other sites and nonmalignant CNS tumors were not locally reportable.

1. The patient had a computerized tomography (CT) scan on January 2, 2004, showing an acoustic neuroma. On December 3, 2004, the patient had a craniotomy and removal of the tumor. In the pathology report the final diagnosis was acoustic neuroma.

   What is the date of diagnosis?
   January 2, 2004, the date of the CT scan, is the date of diagnosis because that was the first time the acoustic neuroma was stated to be the diagnosis. The date of diagnosis is the first date the condition was recognized by a medical practitioner, not the first date of pathologic confirmation.

   What is the sequence number?
   60: it is a solitary benign tumor. The information available does not indicate a previous nonmalignant tumor of the CNS.

   Primary site: **C72.4, acoustic nerve**

   Histology: **9560/09, acoustic neuroma**

2. The patient had excisional biopsy on March 1, 2004, and the pathology was WHO grade I gangliocytoma of the basal ganglia. On October 15, 2004, the patient had a re-resection of a tumor of the basal ganglia. The final pathologic diagnosis was anaplastic ganglioglioma, WHO grade III.

   What is the date of diagnosis?
   **For gangliocytoma, the date was March 1, 2004.
   For anaplastic ganglioglioma, the date was October 15, 2004.**

   The original tumor underwent malignant transformation and changed from WHO grade I to WHO grade III. When this occurs, the tumors are considered two primaries and one abstract is completed for the benign tumor and a second abstract is completed for the malignant tumor.

   What is the sequence number?
   **Gangliocytoma, 60 (first benign tumor)
   Anaplastic ganglioglioma, 00 (first malignant tumor)**

   Primary site: **C71.0, basal ganglia**

   Histology: **First tumor: 9492/09, gangliocytoma
   Second tumor: 9505/34, ganglioglioma, anaplastic
   The *ICD-O-3* grade is 4 for the second tumor because it is anaplastic.**

3. The patient is deaf. In 1998, the patient had surgery to remove an acoustic neuroma. A CT scan showed a spinal cord tumor on March 3, 2004. On March 21, 2004, the patient had a laminectomy and partial removal of tumor at T7. The pathology report documented psammomatous meningioma of the dura.

   What is the date of diagnosis? **March 3, 2004, the date of the CT scan.**

   What is the sequence number? **62, meningioma, because it is the second benign tumor, even though the first benign tumor was not reported, because it was diagnosed prior to January 1, 2004.**

   Primary site: **C70.1, spinal meninges**
   The primary site is spinal meninges instead of spinal cord, because the site is stated to be dura, which is one of the layers of meninges.

   Histology: **9533/09, psammomatous meningioma**

4. On April 1, 2004, the patient had a CT scan of the head that showed cholesteatoma. On April 15, 2004, an MRI of the head showed left temporal meningioma. On April 30, 2004, the patient had surgery to remove the meningioma. The final pathologic diagnosis was meningioma of the left inferior temporal dura.

   What is the date of diagnosis? **April 15, 2004, the date of the MRI. The cholesteatoma diagnosed on April 1 is not a reportable condition.**

   What is the sequence number?
   60, first benign tumor

   Primary site: **C70.0, cerebral meninges**
   The meningioma is sited to the cerebral meninges rather than the temporal lobe, because it is stated to be in the temporal dura, which is part of the meninges.

   Histology: **9530/09, meningioma, NOS**

5. An MRI on January 3, 2004, was used to diagnose subependymoma. On January 31, 2004, the patient had a stereotactic craniotomy and removal of the subependymoma from the medulla oblongata. The patient later had a bulge in the lumbar spinal cord and on December 15, 2004, an MRI was used to diagnosed meningioma. On December 30, 2004, a meningioma was removed. The pathology report documented intradural meningioma.

   What is the date of diagnosis? **Subependymoma, January 3, 2004, date of the MRI. Meningioma, December 15, 2004, date of the MRI.**

   What is the sequence number? **Subependymoma, 61, because it is the first of more than one benign tumor. Meningioma, 62, because it is the second of more than one benign tumor.**

   Primary site: **First tumor: C71.7, medulla oblongata. Second tumor: C70.1, spinal meninges**

   The second primary is coded to the spinal meninges, because it is stated to be intradural in the lumbar spine.

   Histology: **First tumor: 9383/19, subependymoma. Second tumor: 9530/09, meningioma, NOS.**

Part IV Answers
Coding Collaborative Stage and Treatment

For these exercises, assume that if a treatment is not mentioned, it was not administered.

1. The patient was referred to a neurologist because of a right-side hearing loss and possible acoustic tumor. A CT scan on March 1, 2004, was used to diagnose right acoustic neuroma. On March 31, 2004, the patient had a craniotomy. A protrusion of tumor in the vestibule appeared to come from the cochlea. Facial nerve was spared. The tumor was totally resected, with clear margins.

   CS Extension code
   Use "Other parts of CNS" schema.
   Code 05, Benign or borderline brain tumor

   Surgical procedure of primary site
   Use surgery codes for Brain.
   Code 55, Gross total resection (All of the tumor was removed, with no evidence of tumor.)

   Surgical margins of primary site
   Code 0, No residual tumor

   Scope of regional lymph node surgery
   Code 9, Not applicable

   Radiation treatment volume
   Code 00, No radiation treatment

   Regional treatment modality (radiation)
   Code 00, No radiation treatment

   Chemotherapy
   Code 00, None

   Hormone therapy
   Code 00, None

   Immunotherapy
   Code 00, None

   Hematologic transplant and endocrine procedures
   Code 00, None

2. A 6-year-old boy had severe headaches and vomiting. An MRI on September 13, 2004, was used to diagnose medulloblastoma of the cerebellum. An infratentorial craniotomy was performed on September 28, 2004, and the tumor was removed. Macroscopic and microscopic residual medulloblastoma remained. On November 1, 2004, the patient began a course of carmustine. After completion of chemotherapy, the patient had a bone marrow transplant with donor marrow from his older brother.

   CS Extension code
   Use "Brain & Cerebral Meninges" schema.
   Code 11, Infratentorial tumor, confined to cerebellum

Surgical procedure of primary site
Use surgery codes for Brain.
Code 40, partial resection (Tumor was removed, but visible and microscopic tumor remained.)

Surgical margins of primary site
Code 3, Macroscopic residual tumor (When both microscopic and macroscopic residual tumor remain, use the higher code.)

Scope of regional lymph node surgery
Code 9, Not applicable

Radiation treatment volume
Code 00, No radiation treatment

Regional treatment modality (radiation)
Code 00, No radiation treatment

Chemotherapy
Code 02, Single agent chemotherapy

Hormone therapy
Code 00, None

Immunotherapy
Code 00, None

Hematologic transplant and endocrine procedures
Code 12, Allogeneic bone marrow transplant (Patient's brother donated the bone marrow.)

3. Patient had CT scan of the head on February 24, 2004, showing a large prolactinoma of the pituitary gland. The patient started on bromocriptine to shrink the tumor in March 2004. The patient had gamma knife radiosurgery on October 1, 2004.

CS Extension code
Use "Thymus, Adrenal Gland and Other Endocrine Glands" schema.
Code 05, Benign or borderline tumors

Surgical procedure of primary site
Use Surgery codes for "All Other Sites."
Code 00, None (Gamma knife radiosurgery is radiation.)

Surgical margins of primary site
Code 8, No primary site surgery

Scope of regional lymph node surgery
Code 0, None (None instead of not applicable, because pituitary is not one of the sites listed in *FORDS* as "not applicable" for lymph node surgery.)

Radiation treatment volume
Code 02, Pituitary

Regional treatment modality (radiation)
Code 43, Gamma knife

Chemotherapy
Code 00, None

Hormone therapy
Code 00, None

Immunotherapy
Code 82, Immunotherapy as first course of therapy (Bromocriptine is a biologic response modifier or immunotherapy.)

Hematologic transplant and endocrine procedures
Code 00, None

4. The patient was referred to a neurologist after reporting symptoms of vomiting, muscle weakness on one side of the face, and several episodes of slurred speech. The patient had an MRI on June 3, 2004, that showed a glioma in the brain stem. Through an infratentorial craniotomy, the tumor was removed on June 30, 2004. The pathology report documented microscopic residual subependymal glioma in the fourth ventricle (9383/1). On August 1, 2004 the patient had conformal radiation to the fourth ventricle.

   CS Extension code
   Use "Brain & Cerebral Meninges" schema.
   Code 05, Benign

   Surgical procedure of primary site
   Use surgery codes for Brain.
   Code 55, Gross total resection (When tumor is removed and only microscopic residual tumor remains, surgery is still considered total resection.)

   Surgical margins of primary site
   Code 2, Microscopic residual tumor

   Scope of regional lymph node surgery
   Code 9, Not applicable

   Radiation treatment volume
   Code 04, Brain limited (The radiation was given to only part of the brain, fourth ventricle.)

   Regional treatment modality (radiation)
   Code 32, Conformal therapy

   Chemotherapy
   Code 00, None

   Hormone therapy
   Code 00, None

   Immunotherapy
   Code 00, None

   Hematologic transplant and endocrine procedures
   Code 00, None

5. The patient's symptoms included headaches, double vision, vomiting, and drowsiness. A CT scan on November 1, 2004, showed a growth confined to the pineal gland. The patient had a biopsy of the pineal gland on November 15, 2004, and the tumor pathology was pineocytoma. The patient began beam radiation to the pineal gland on December 8, 2004.

   CS Extension code
   Use "Thymus Gland and Other Endocrine Glands" schema.
   Code 05, Benign and borderline tumors

   Surgical procedure of primary site
   Use surgery codes for All Other Sites.
   Code 00, None, no surgery of primary site (This is not stated to be an "excisional biopsy" which would be coded "27." Stereotactic biopsy of tumors of the pineal region has recently become popular, particularly for those patients who do not benefit from open surgery. Biopsy can also be performed endoscopically using a flexible ventriculoscope.)

   Surgical margins of primary site
   Code 7, Margins not evaluable

   Scope of regional lymph node surgery
   Code 0, None (None instead of not applicable, because pituitary is not one of the sites listed in *FORDS* to use "not applicable" for lymph node surgery.)

   Radiation treatment volume
   Code 04, Brain limited

Regional treatment modality (radiation)
Code 20, External beam, NOS

Chemotherapy
Code 00, None

Hormone therapy
Code 00, None

Immunotherapy
Code 00, None

Hematologic transplant and endocrine procedures
Code 00, None