FLORIDA CANCER DATA SYSTEM

# JANUARY/FEBRUARY 2004 MONTHLY MEMO

HAPPY VALENTINES DAY

## WHAT'S NEW:

**The Following newsletters and reports are currently available from the FCDS website:**

- THE DECEMBER 2003 MONTHLY MEMO
- COC CONVERSION ERRATA
- DATA ACCESS MANUAL ERRATA
- THE FCDS REGISTER, VOL. 22

## On the Web:

- **2001 FCDS Monograph of Cancer in Florida** **https://fcds.med.miami.edu/stats/monograph/2001/FCDS_Monograph2001_Vol1_Incidence.pdf**
- **Cancer in North America, 1996-2000** **http://www.naaccr.org/index.asp?Col_SectionKey=11&Col_ContentID=50**
- **Summary Stage: Data Effects of the Changes in 2000** **http://www.naaccr.org/filesystem/pdf/Summary%20Stage%20Report%201-21-04b.pdf**

# New Reporting Requirements For 2004

By: Stuart Herna, CTR

Beginning with cases diagnosed on or after January 1, 2004, implementation of new rules, regulations and reporting requirements will be employed that will dramatically change not only how we collect and stage our cases, but the very nature of the type of cases that we collect as well. These new reporting requirements, effective July 1, 2004, will focus on reporting of benign brain tumors and a change to the staging schema used by FCDS.

Benign Brain: New legislation passed by both the House and the Senate and signed by President Bush in October of 2002 enacted the **Benign Brain Tumor Cancer Registries Act** (Public Law 107-260). This act requires the abstracting and reporting of non-malignant primary intracranial and Central Nervous System tumors by the National Program of Cancer Registries (NPCR). The Commission on Cancer (CoC) and the Surveillance, Epidemiology, and End Results Program (SEER) added benign and borderline intracranial and Central Nervous System tumors to their case definitions soon thereafter.

Collaborative Staging: This new staging system was designed to eliminate duplicate data collection by registrars reporting to facility based and central registries, to address the concerns of clinicians for more accurate and complete data, and to provide greater parity and reduce discrepancies between the three major staging systems used in the United States (SEER, TNM and EOD). Beginning with cases diagnosed January 1, 2004 and forward, FCDS will collect all 15 items of the Collaborative Staging schema. Because of the change in the record layout, no 2004 cases will be accepted prior to July 1, 2004. And as with all previous conversions, any 2003 cases not reported by the FCDS deadline of June 30, 2004 must be submitted to FCDS in the new NAACCR v10.1 format and must include all 15 items of the Collaborative Staging schema.

**Benign and Borderline Intracranial and CNS Reporting Requirements**

Any tumor diagnosed on or after January 1, 2004 with a behavior code of '0' or '1' will be collected for the following sites based upon the International Classification of Diseases for Oncology, 3rd Edition (ICD-O 3): Meninges (C70.0-C70.9), Brain (C71.0-C71.9), Spinal Cord/Cranial Nerves/ Other CNS (C72.0-C72.9), Pituitary Gland (C75.1), Craniopharyngeal Duct (C75.2) and Pineal Gland (C75.3). Morphology Codes will also be based upon ICD-O 3.

Juvenile Astrocytomas should continue to be coded as 9421/3.

With the implementation of collection of benign and borderline intracranial and CNS tumors, facilities will need to add the ICD-9-CM

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codes for these diseases to their casefinding list.

**ICD-9-CM Casefinding Codes for Benign and Borderline Intracranial and CNS Tumors**

| ICD-9 CM Code | Description of Neoplasm |
|---|---|
| **225.0** | Benign neoplasm of brain |
| **225.1** | Benign neoplasm of cranial nerves |
| **225.2** | Benign neoplasm of cerebral meninges; 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, craniabuccal pouch, hypophsis, Rathke's pouch, sella turcica |

**ICD-9-CM Casefinding Codes for Benign and Borderline Intracranial and CNS Tumors (Cont'd)**

| | |
|---|---|
| 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: NOS, cerebral, spinal |
| **237.70** | Neurofibromatosis, Unspecific 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 |

Researchers have indicated that the laterality of primary intracranial and CNS tumors is pertinent in determining their cause and assessing quality of life. Beginning with January 1, 2004, reporting laterality for the following sites will be required for primary malignant and non-malignant tumors: C70.0 Cerebral Meninges, NOS, C71.0 Cerebrum, C71.1 Frontal Lobe, C71.2 Temporal Lobe, C71.3 Parietal Lobe, C71.4 Occipital Lobe, C72.2 Olfactory Nerve, C72.3 Optic Nerve, C72.4 Acoustic Nerve, and C72.5 Cranial Nerve NOS.

Traditionally, primary intracranial and CNS tumors were reported with a laterality code of '0'. Laterality codes will now be expanded to allow for codes that depict the side of the brain in which the reportable tumor originates. Laterality codes to be used are as follows:
Sequence numbers for **primary non-malignant** intracranial and CNS tumors are to be assigned in the range from 60 – 87. All **primary malignant** tumors will continue to be sequenced in the range from 00 – 35.

| | |
|---|---|
| 0 | Not a paired site |
| 1 | Right: origin of primary |
| 2 | Left: origin if primary |
| 3 | Only one side involved, right or left origin unspecified |
| 4 | Bilateral involvement, lateral origin unknown; stated to be single primary |
| 9 | Paired site, but no information regarding laterality, midline tumor |

New rules for determining multiple primaries for non-malignant tumors have been created. They are as follows:

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Differences in histologic type refer to differences in the **First Three Digits** of the histology code.

If multiple non-malignant tumors occur in the same site and the first three digits of the histology code are the same they are considered the same primary.

If multiple non-malignant tumors occur in the same site and the first three digits of the histology codes are different, they are considered separate primaries.

Each topographic sub-site (4th digit level) as delineated in ICD-O 3 is considered to be a separate site.

If there are multiple tumors of the same site and the same histology and both sides of the site are involved, the tumors should be counted as separate primaries.

If non-malignant tumors of the same histology, same site, and the same side recur in the same location, regardless of the time frame, they are considered the same primary.

The 6th digit of the ICD-O 3 Histology code refers to tumor grade or differentiation. This field should always be coded '9' (not applicable) for non-malignant intracranial and CNS tumors. The 6th digit Grade Code is not the same as the WHO Grade Code. WHO Grade is coded in the Collaborative Staging data field entitled Site Specific Factor 1 for Brain and CNS tumors. The WHO Grade codes are as follows:

WHO Grade I tumors are slow growing, have long-term survival and are non-malignant.

WHO Grade II tumors are relatively slow growing but sometimes recur as a higher grade tumor. They may be non-malignant or malignant.

WHO Grade III tumors are by definition malignant and often recur as higher grade tumors.

WHO Grade IV tumors reproduce rapidly and are very aggressive malignant tumors.
SEER Summary Stage 2000 code '8' (not applicable) will be derived from the Collaborative Staging algorithm for non-malignant intracranial and CNS tumors.

A Benign Brain Tumor Reporting web based training module is currently available on SEER's training web site at www.training.seer.cancer.gov.

**Collaborative Staging Reporting Requirements**

Collaborative Staging is effective beginning with cancer cases diagnosed on or after January 1, 2004. For each cancer case, the abstractor extracts the required Collaborative Staging information from the medical record and codes the information into the corresponding Collaborative Staging System fields. When data collection is complete, a computer algorithm **derives** the AJCC 6th Ed TNM Stage, STAGE Group, SEER Summary Stage 1977, SEER Summary Stage 2000, AJCC TNM Descriptors, AJCC Flag, Summary Stage 1977 Flag and Summary Stage 2000 Flag. Derived Collaborative Staging fields should be viewable only and the ability to edit should not be available.

General rules and guidelines that apply to the Collaborative Staging system are as follows:

Collaborative Staging is collected on all sites and histologies regardless of whether or not they are microscopically confirmed. Cases not microscopically confirmed should be coded from the schema for the site and histology that the physician determines to be the primary.

Cases should be microscopically confirmed for Collaborative Staging to effectively assign TNM, Stage Group and SEER Summary Stage. Summary Stage 1977 and Summary Stage 2000 are generated for all sites and histologies. The TNM Stage and Stage Group are only generated for those sites and histologies that meet TNM Stage and Stage Group criteria. Cases not meeting TNM Stage and Stage Group criteria receive a derived value of "Not Applicable" once the computer algorithm is generated.

Site specific and histology specific guidelines take precedence over general guidelines. Some malignancies that can develop in different parts of the body (i.e. lymphoma) are coded to the histology of the case, not to the site. A case with one of these histologic types must be coded using the shema for the histologic type group: Melanoma, Kaposi Sarcoma, Retinoblastoma, Lymphoma, Mycosis Fungoides, and Hematopoietic and Reticuloendothelial system.

Data collected in the Collaborative Staging system are limited to all information gathered through the completion of surgery in the first course of treatment or all information available within four months of the date of diagnosis, whichever is longer.

For each field, code the highest applicable number. The codes are hierarchal except for the codes for Unknown, Not Applicable and NOS. More specific codes with lower numbers take priority over these.
Record the greatest extent of disease based upon combined clinical and pathologic assessment.

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If the patient does not receive pre-operative treatment, pathology information takes priority over clinical information.

If the patient does receive pre-operative treatment, extent of disease should be based upon either clinical or pathologic findings, whichever is greater.

Metastases that develop after the initial extent of disease is established (disease progression) should be excluded when determining extent of disease at diagnosis.

The Collaborative Staging system applies the same rules for inclusion and exclusion to Autopsy reports as it does to pathology reports.

If the extent of disease is described by the physician in terms of TNM elements, code the TNM information in the appropriate fields. If there is a discrepancy between documentation in the medical record and the physician assigned TNM Stage, the documentation takes precedence.

In all, there are fifteen data items collected in the Collaborative Staging System. These data items and their descriptions are as follows:

**CS Tumor Size** records the largest lateral dimension of the primary tumor in millimeters. Information previously collected in this field such as depth of invasion for malignant melanoma has been moved to Site Specific Factors.

**New Rules**: If no pre-operative treatment was performed, always code the largest pathologic tumor size recorded. If pre-operative treatment was performed, code the pre-operative clinical tumor size. Imaging takes precedence over physical examination findings for determining tumor size. Code the size of the invasive component of the tumor if it is available. Site Specific code 998 takes precedence over a stated tumor size for the following sites: esophagus (entire circumference), stomach (diffuse, widespread ¾ or more, linitis plastica), colorectal (familial/ multiple polyposis), lung and main stem bronchus (diffuse, entire lobe or lung) and breast (diffuse, widespread ¾ or more, inflammatory carcinoma). Site Specific Code 990 (microscopic focus) can only be used when tumor is identified microscopically.

**CS Extension** identifies contiguous growth of the primary tumor within the organ of origin or by direct extension into surrounding tissues.

**New Rules**: Record the farthest extension of the primary tumor. Extension must be direct or contiguous (except for corpus and ovary). Disregard discontinuous metastases to distant sites. If no pre-operative treatment was performed, use information from the path report. If pre-operative treatment was performed, use pre-operative clinical extension. If post op path is more extensive, use the more extensive code. Imaging takes priority over physical examination. Disregard microscopic residual tumor or positive margins when coding extension. If involved organ is not listed, approximate the extension and code with similar tissues (consult a physician). If there is any lymph node or metastatic involvement, Tumor Extension cannot be In Situ. Code Extension as Local NOS if no other information is available.

**CS Tumor Size/Extent Evaluation** records how the codes for Collaborative Staging Tumor size and Collaborative Staging Extension were determined and whether the patient received pre-operative treatment.

*Code 0* = physical examination, imaging or other non-invasive clinical evidence.
*Code 1* = endoscopy, biopsy or other invasive techniques including surgical observation without biopsy.
*Code 2* = no surgery. Based on autopsy.
*Code 3* = based on surgical findings. No pre-surgical systemic treatment or radiation therapy **OR** it is unknown if patient had pre-surgical systemic treatment or radiation therapy.
*Code 5* = surgery with pre-surgical systemic treatment or RT (clinical evidence).
*Code 6* = surgery with pre-surgical systemic treatment or RT (pathologic evidence).
*Code 8* = autopsy only (tumor not suspected or diagnosed prior to autopsy).
*Code 9* = unknown if surgical resection performed; not assessed.

**CS Lymph Nodes** identifies the regional lymph nodes involved with cancer at the time of diagnosis.

**New Rules**: Record the farthest involved regional lymph node chain; disregard distant lymph nodes. If no pre-operative treatment, use path report. If pre-operative treatment, use pre-operative clinical information. Code as much detail as possible. The size of mets in a lymph node is coded, not the size of the lymph node. For low stage inaccessible primaries, code as clinically negative if there is no mention of regional lymph node involvement either on physical exam, imaging or surgical exploration **AND** patient receives standard treatment to the primary site. If tumor not local, code lymph nodes as unknown. For accessible sites, look for statement of negativity such as "remainder of exam negative".

**CS Regional Lymph Nodes Evaluated** records how the code for Collaborative Staging Lymph Nodes was determined based upon the

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# DEADLINES AND REMINDERS

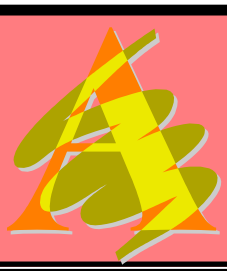
## RADIATION THERAPY CENTERS CANCER CASE IDENTIFICATION PROGRAM

Beginning with the 2003 patient encounters, Radiation Therapy Centers are responsible for identifying and reporting all of their cancer cases to the Florida Cancer Data System using the FCDS-IDEA Single Entry or the File Upload modules. The deadline to submit the cases is June 30, 2004. Please log on to the FCDS website fcds.med.miami.edu or call Betty Hallo at (305) 243-2627 for additional information.

## PATH LAB REPORTING

Every anatomic pathology laboratory that reads biopsy and surgical resection specimens collected from patient encounters within the state of Florida MUST electronically submit the specified data for every malignant cancer case. **Specimens read between July 1, 2003 and December 31, 2003 must be submitted to FCDS on or before June 30, 2004.**

# FCDS Q & A



## GRADE--BRAIN AND CNS

References
ICD-O-3 ;pgs 67 Brief

**Q:**
Should "high grade" be coded for a Brain/CNS malignancy?

**A:**
Grade, ICD-O-3 morphology 6th digit, is usualy not specified for CNS malignancies. Code grade only when differentiation is specified, such as "well," "moderately," "poorly," "anaplastic." Do not code WHO grade 1, 2, 3 or 4 in the 6th digit of the morphology code.

Do not code "high grade" for the glioblastoma multiforme in the example above; code grade 9 [Not determined, not stated or not applicable].

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diagnostic methods employed and whether the patient received pre-operative treatment.

*Code 0* = physical examination, imaging or other non-invasive clinical evidence.
*Code 1* = endoscopy, biopsy or other invasive techniques including surgical observation without biopsy. No LN's removed.
*Code 2* = no LN's removed. Based on autopsy.
*Code 3* = Lymph node removed. No pre-surgical systemic treatment, RT, LN removed **OR** unknown if patient had pre-surgical systemic treatment or RT.
*Code 5* = LN removed with pre-surgical systemic treatment or RT (clinical evidence).
*Code 6* = LN removed with pre-surgical systemic treatment or RT (pathologic evidence).
*Code 8* = autopsy only (tumor not suspected or diagnosed prior to autopsy).
*Code 9* = unknown if surgical resection performed; not assessed.

**Regional Lymph Nodes Examined** records the total number of regional lymph nodes that were removed and examined by the Pathologist.

**New Rules**: If no LN's examined code 00. If no LN's in specimen code 00. Aspiration of LN is coded 95. Any combination of aspirated/biopsied/sampled/dissected LN's is coded 98.

**Regional Lymph Nodes Positive** records the number of regional lymph nodes examined by the Pathologist and found to be positive.

**New Rules**: Minor changes from current rules. If no LN's examined, code 98. Positive aspiration is coded 95. Any combination of positive aspirated/biopsied/sampled/dissected LN's is coded 97 (LN's positive but number unknown).

**CS Mets At Diagnosis** identifies the distant site or sites of metastatic involvement at the time of diagnosis.

**New Rules**: Record only blood borne or implantation discontinuous mets or distant LN involvement. Code structures, nodes and tissues not listed in Extension or LN fields. Disregard mets that develop after extent of disease was established. For low stage inaccessible primary sites code as clinically negative if there is no mention of distant mets on PE/imaging/surgical exploration **and** patient receives standard treatment to primary site. If tumor no longer local, Mets at Diagnosis may be coded unknown. For accessible sites, look for statement of negativity.

**CS Mets Eval** records how the code for data item Collaborative Staging Mets At Diagnosis was determined based upon the diagnostic methods employed and whether the patient received pre-operative treatment.

*Code 0* = physical examination, imaging or other non-invasive clinical evidence.
*Code 1* = endoscopy, biopsy or other invasive techniques including surgical observation without biopsy.
*Code 2* = no metastatic tissue examined prior to death. Based on autopsy.
*Code 3* = metastatic tissue examined. No pre-surgical systemic treatment, RT or metastatic tissue examined **OR** it is unknown if patient had pre-surgical systemic treatment or RT.
*Code 5* = metastatic tissue examined with pre-surgical systemic treatment or RT (clinical evidence).
*Code 6* = metastatic tissue examined with pre-surgical systemic treatment or RT (pathologic evidence).
*Code 8* = autopsy only (tumor not suspected or diagnosed prior to autopsy).
*Code 9* = unknown if surgical resection performed; not assessed.

**CS Site Specific Factors 1 through 6** identify additional information necessary to generate stage or prognostic factors that effect stage or survival. The six Site Specific Factors are used to collect information including HIV status. They replace existing tumor marker fields. These six fields are only used as needed by primary site. For some sites where Site Specific factors are not used, they are coded 888 (not applicable).

Once data collection is complete, the computer algorithm is generated that **derives** the following twelve data items:

**Derived AJCC 'T'** is determined by the combination of the Collaborative Staging Tumor Size field, the Tumor Extension field, the Method of Evaluation field and any required Site Specific Factor fields.

**Derived AJCC 'T' Descriptor** indicates how the data was derived, either clinically, pathologically, at autopsy, or based upon pathologic evidence of extension after pre-surgical systemic treatment or radiation.

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**Derived AJCC 'N'** is determined by the combination of the Collaborative Staging LN field, the LN Evaluation field, the Number of LN's Examined and the Number of LN's Positive fields.

**Derived AJCC 'N' Descriptor** indicates how the data was derived, either clinically, pathologically, at autopsy, or based upon nodal involvement after pre-surgical systemic treatment or radiation.

**Derived AJCC 'M'** is determined by the combination of the Collaborative Staging Metastases at Diagnosis and the Mets Evaluation fields.

**Derived AJCC 'M' Descriptor** indicates how the data was derived, either clinically, pathologically, at autopsy, or based upon metastatic evaluation after pre-surgical systemic treatment or radiation.

**Derived AJCC Stage Group** represents the combination of the Collaborative Staging T, the N and the M elements into one group.

**Derived Summary Stage 1977** indicates the anatomic extent of disease at diagnosis for cases diagnosed prior to January 1st, 2001.

**Derived Summary Stage 2000** indicates the anatomic extent of disease at diagnosis for cases diagnosed on or after January 1st, 2001.

**Derived AJCC Flag** indicates whether stage was coded directly or derived from Collaborative Staging.

**Derived Summary Stage 1977 Flag** indicates whether stage was coded directly or derived from Collaborative Staging.

**Derived Summary Stage 2000 Flag** indicates whether stage was coded directly or derived from Collaborative Staging.

## Impact On Registries Of New Reporting Requirements For 2004

**Benign and borderline** intracranial and CNS tumor incidence is estimated to be equivalent to that of **malignant** intracranial and CNS tumors. Cancer Registries can get a rough estimate of the increase in volume that will result from collection of these tumors by doubling the number of their reported malignant intracranial and CNS tumors for a given year.

Software will need to be modified to allow for the collection of new or revised data fields.

The Site/Histology Validation List, the Case Finding List, the Reportable List and the Policy and Procedure Manual all need to be updated to include benign and borderline intracranial and CNS tumors, Collaborative Staging data elements and the new rules and regulations for data collection and reporting.

Edits need to be modified to accommodate non-malignant behavior codes and sequence numbers and to handle the Collaborative Staging data items.

Changes to the criteria for determining the data reported to Central Registries will be necessary to allow for the transmission of benign and borderline CNS and intracranial tumors with a Behavior Code of 0 or 1 and the Collaborative Staging data elements.

A slight learning curve is anticipated for new data fields and codes.

Time will be saved since the Registrar will no longer need to reference different staging manuals, separate books, documents or help screens in order to make decisions to assign stage.

## Training and Educational Resources

Training and educational resources, including the Collaborative Staging And Coding Manual Volumes 1 and 2, are available on the web at www.cancerstaging.org. The CoC will distribute one copy of the Collaborative Staging And Coding Manual Volumes 1 and 2 to each CoC approved cancer program. FCDS will provide one copy of the manuals to all non-approved facilities.

FCDS will be presenting a series of teleconferences covering both Collaborative Staging and reporting of benign and borderline CNS and intracranial tumors. The first, on Thursday February 25th, 2004 from 2:00 – 3:00 P.M., will provide a general overview and discussion of new reporting requirements for 2004. The second, on Wednesday March 24th, 2004 from 2:00 – 4:00 P.M., will cover Part I of Collaborative Staging. The third, on Wednesday April 14th, 2004 from 2:00 – 4:00 P.M., will cover Part II of Collaborative Staging. All are welcome to participate. For additional information please access the FCDS website at http://fcds.med.Miami.edu and click on '**What's New'.**

*Some information contained in this article was originally presented in the*

# Benign Brain Tumor Reporting

**SEER Training Website:** http://training.seer.cancer.gov/module_bbt/00_bbtr_home.html

## BRAIN AND CNS TUMORS

It is expected that more than 190,600 brain tumors will be diagnosed in the United States during 2003. Among these cases, there are about 40,600 primary brain tumors and 150,000 secondary (metastatic) brain tumors. In the United States, approximately 3,100 children younger than age 20 are diagnosed annually with brain tumors. Brain and CNS cancer is the leading cause of cancer-related death in patients younger than age 35 in this country.

Primary tumors are tumors that begin in the brain and tend to stay in the brain. Metastatic brain tumors begin as a cancer elsewhere in the body and spread, or metastasize, to the brain. Metastatic brain tumors are the most common brain tumor, with an annual incidence more than four times greater than that of primary brain tumors. The cancers that most commonly metastasize to the brain are breast and lung cancer.

Brain tumors are different from other cancers in several ways. One important factor is that brain tumors develop within the confined space of the skull where there is little extra room into which they may grow. Thus, even a small tumor can seriously affect normal brain function.

Also, brain tumors have relatively little tendency to metastasize outside the nervous system regardless of histologic type. Therefore, the terms "benign" and "malignant" have different meanings from those referring to abnormal growths elsewhere in the body. For brain tumor, benign means the tumor is relatively slow-growing; malignant means the tumor is aggressive or fast-growing. Most histologic types of CNS tumors can be either benign or malignant. Remember that a benign CNS tumor can become just as dangerous as a malignant one if the tumor presses on a vital area of brain tissue.
Go to Types of Brain and CNS Tumors on the Seer Training Website. to learn more about types of brain and CNS tumors.

Anatomy

The CNS consists of the brain and spinal cord, which are located in the dorsal body cavity. The brain is surrounded by the cranium, and the spinal cord is protected by the vertebrae. The brain is continuous with the spinal cord at the foramen magnum. In addition to bone, the CNS is surrounded by connective tissue membranes, called meninges, and by cerebrospinal fluid. The following are the major components of the brain and CNS.

- Structure of neurons and glial cells
- Brain
- Meninges
- Spinal Cord
- Cranial Nerves
- Pineal and Pituitary Glands

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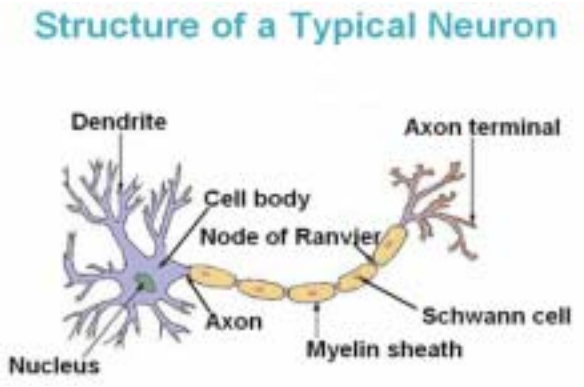
Structure of Neurons and Glial Cells

Neurons are the conducting cells of the nervous system. A typical neuron consists of a cell body, containing the nucleus and the surrounding cytoplasm; several short radiating processes (dendrites); and one long process (the axon), which terminates in twiglike branches and may have branches projecting along its course.




Cell Body

In many ways, the cell body is similar to other types of cells. It has a nucleus with at least one nucleolus and contains many of the typical cytoplasmic organelles. It lacks centrioles, however. Because centrioles function in cell division, the fact that neurons lack these organelles is consistent with the amitotic nature of the cell.

Dendrites and Axons

An axon is a long, hair-like extension of a nerve cell that carries a message to another nerve cell.

Dendrites are thread-like extensions of the cytoplasm of a neuron that receive signals from other neurons. Typically, as in multipolar neurons, dendrites branch into treelike processes, but in unipolar and bipolar neurons, dendrites resemble axons.

Glial Cells

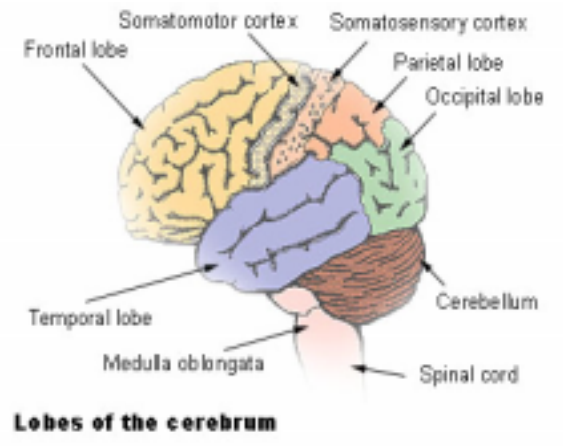
Glial (Neuroglial) cells do not conduct nerve impulses, but instead, support, nourish, and protect the neurons. Glial cells are far more numerous than neurons and, unlike neurons, are capable of mitosis.

Brain

Cerebrum

Cerebrum is the part of the brain that receives and processes conscious sensation, generates thought, and controls conscious activity. It is the uppermost and largest part of the brain, and is divided into left and right hemispheres, which are joined by and communicate through the corpus callosum.

Each cerebral hemisphere is divided into five lobes, four of which have the same name as the bone over them: the frontal lobe, the parietal lobe, the occipital lobe, and the temporal lobe. A fifth lobe, the insula or Island of Reil, lies deep within the lateral sulcus.



**Lobes of the cerebrum**

Cerebellum

The Cerebellum is a cauliflower-shaped section of the brain located in the hindbrain, at the bottom rear of the head, directly behind the pons. The cerebellum is a complex system mostly dedicated to the intricate coordination of voluntary movement, including walking and balance. Damage to the cerebellum leaves the sufferer with a gait that appears drunken and is difficult to control.

Ventricles and Cerebrospinal Fluid

A series of interconnected, fluid-filled cavities called ventricles lie within the brain. The fluid is cerebrospinal fluid (CSF), which also circulates over the outside of the brain and spinal cord.

Brain Stem

The brain stem is the part of the brain continuous with the spinal

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cord and comprising the medulla oblongata and pons and midbrain and parts of the hypothalamus.

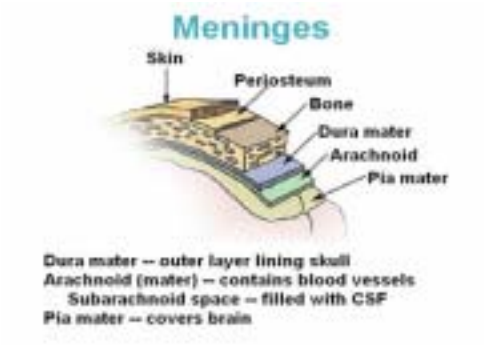
Tentorium

The tentorium is a fold of the dura mater which separates the cerebellum from the cerebrum and often encloses a process or plate of the skull called the bony tentorium.


Meninges

There are three layers of meninges around the brain and spinal cord. The outer layer, the dura mater, is tough white fibrous connective tissue. The middle layer of meninges is arachnoid, a thin layer resembling a cobweb with numerous threadlike strands attaching it to the innermost layer. The space under the arachnoid, the subarachnoid space, is filled with cerebrospinal fluid and contains blood vessels. The pia mater is the innermost layer of meninges. This thin, delicate membrane is tightly bound to the surface of the brain and spinal cord and cannot be dissected away without damaging the surface.

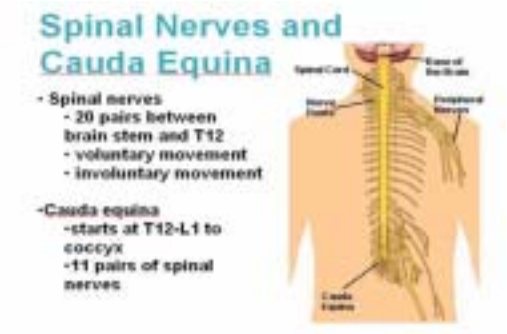
Meningiomas are tumors of the nerve tissue covering the brain and spinal cord. Although meningiomas are unlikely to spread, physicians often treat them as though they were malignant because symptoms that may develop when a tumor applies pressure to the brain.




Spinal Cord

The spinal cord extends from the foramen magnum at the base of the skull to the level of the first lumbar vertebra. The cord is continuous with the medulla oblongata at the foramen magnum. Like the brain, the spinal cord is surrounded by bone, meninges, and cerebrospinal fluid.

The spinal cord is divided into 31 segments with each segment giving rise to a pair of spinal nerves. At the distal end of the cord, many spinal nerves extend beyond the conus medullaris to form a collection that resembles a horse's tail. This is the cauda equina. In cross section, the spinal cord appears oval in shape.

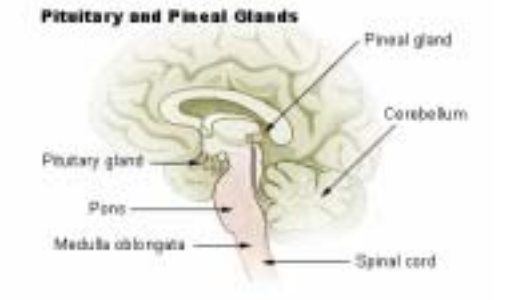



Cranial Nerves

The cranial nerves are composed of twelve pairs of nerves that emanate from the nervous tissue of the brain. In order to reach their targets they must ultimately exit/enter the cranium through openings in the skull. Hence, their name is derived from their association with the cranium. See list of cranial nerves, their functions, and tumor examples on page 5.

Pineal and Pituitary Glands

The pineal gland is a small endocrine gland in the brain, situated beneath the back part of the corpus callosum, and secretes melatonin. The pituitary gland is located at the base of the brain that secretes hormones and regulates and controls other hormone-secreting glands and many body processes, including reproduction.




*(Continued on page 11)*

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## Cranial Nerves (Function and Tumor Examples)

| # | Name | Function | Tumor Example |
|---|---|---|---|
| **I** | olfactory | The olfactory nerve carries impulses for the sense of smell. | Esthesioneuronblastoma |
| **II** | optic | The optic nerve carries impulses for the sense of sight. | Optic nerve glioma |
| **III** | occulomotor | The occulomotor nerve is responsible for motor enervation of upper eyelid muscle | Schwannoma |
| **IV** | trochlear | The trochlear nerve controls an extraocular muscle. | Schwannoma |
| **V** | trigeminal | The trigeminal nerve is responsible for sensory enervation of the face and motor enervation to muscles of mastication (chewing). | Malignant peripheral nerve sheath tumor (MPNST) |
| **VI** | abducent | The abducent nerve enervates a muscle which moves the eyeball. | Schwannoma |
| **VII** | facial | The facial nerve enervates the muscles of the face (facial expression). | Schwannoma (rare) |
| **VIII** | vestibulocochlear | The vestibulocochlear nerve is responsible for the sense of hearing and balance (body position sense). | Vestibular Schwannoma |
| **IX** | glossopharyngeal | The glossopharyngeal nerve enervates muscles involved in swallowing and taste. Lesions of the ninth nerve result in difficulty swallowing and disturbance of taste. | Glomus tumor |
| **X** | vagus | The vagus nerve enervates the gut (gastrointestinal tract) | MPNST, paraganglioma |
| **XI** | accessory | The accessory nerve enervates the sternocleidomastoid muscles and the trapezius muscles. | Schwannoma |
| **XII** | hypoglossal | The hypoglossal nerve enervates the muscles of the tongue. | Schwannoma |

Abstracting, Coding, and Staging Brain and CNS Tumors

Topographic Sites

| CEREBRAL MENINGES | |
|---|---|
| **ICD-O-3** | **Term** |
| **C70.0** | Cerebral meninges |
| **C70.1** | Spinal meninges |
| **C70.9** | Meninges, NOS |

*(Continued on page 13)*

# EDUCATION AND TRAINING

## FCDS 2004 EDUCATIONAL TELEPHONE CONFERENCE SERIES

COLLABORATIVE STAGING PART I

MARCH 24, 2004 2PM-4PM
Dial-in number: 888-476-3762
Host Code: 819551
Participant Code: 359957

COLLABORATIVE STAGING PART II

APRIL 14, 2004 2PM-4PM
Dial-in number: 888-422-7137
Host Code: 113138
Participant Code: 175525

## FCRA REGIONAL WORKSHOP

"DATA COLLECTION OF PRIMARY CENTRAL NERVOUS SYSTEM TUMORS"

**Date:** March 20, 2004

**Contact:** Patricia Bentley, CTR, Program Chair
patbentley@cfl.rr.com.

## EMORY UNIVERSITY TRAINING PROGRAMS

### ADVANCED CANCER REGISTRY TRAINING PROGRAM

The Advanced Cancer Registry Program will be held at the Holiday Inn Select Hotel and Suites, 2183 North Decatur Road, Decatur, Georgia 30033, located in the Atlanta-Emory University area on March 29-31, 2004.

Participants **must** be familiar with (at a minimum) the contents of the *Self-Instructional Manual for Tumor Registrars, Book One (Objectives and Functions of a Tumor Registry)* and *Book Three (Tumor Registrar Vocabulary: the Composition of Medical Terms)*. These *Self-Instruction Manuals* are available free from the National Cancer Institute's SEER Program (phone 1-301-496-8510 or fax 1-301-496-9949).

***Registration fee:*** $500 for the full 3 day training

## PRINCIPLES AND PRACTICE OF CANCER REGISTRATION, SURVEILLANCE, AND CONTROL

The Principles and Practice of Cancer Registration, Surveillance and Control will be held at the Holiday Inn Select Hotel and Suites, 2183 North Decatur Road, Decatur, Georgia 30033, located in the Atlanta-Emory University area on April 5-9, 2004.

***Registration fee:*** $1000 for the full week training


Mark your calendars for National Health Information Privacy and Security Week, April 11 through 17, 2004. The week is designed to raise awareness among healthcare professionals, their employers, and the public of the importance of protecting the privacy, confidentiality, and security of personal health information. During the week, AHIMA and its members will work to educate and inform these groups of their rights and responsibilities related to the use and disclosure of personal h

## AHIMA CODING WORKSHOP

Plan now for acquiring the latest and most expert coding knowledge:

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For further information on upcoming AHIMA Coding Workshops visit the AHIMA website at: http://www.ahima.org/coding/coding_meetings.cfm

*(Continued from page 11)*

| BRAIN | | | |
|---|---|---|---|
| | | Supratentorial | Infratentorial |
| C71.0 | Cerebrum | X | |
| | Basal ganglia | X | |
| | Central white matte | X | |
| | Cerebral cortex | X | |
| | Cerebral hemisphere | X | |
| | Corpus striatum | X | |
| | Globus pallidus | X | |
| | Hypothalamus | | X |
| | Insula | X | |
| | Internal capsule | X | |
| | Island of Reil | X | |
| | Operculum | X | |
| | Pallium | | X |
| | Putamen | X | |
| | Rhinencephalon | X | |
| | Supratentorial brain, NOS | X | |
| | Thalamus | | X |
| C71.1 | Frontal lobe | | |
| C71.2 | Temporal lobe | | |
| | Hippocampus | | |
| | Uncus | | |
| C71.3 | Parietal lobe | | |
| C71.4 | Occipital lobe | | |
| C71.5 | Ventricle, NOS* | | |
| | Cerebral ventricle | | |
| | Choroid plexus, NOS* | | |
| | Choroid plexus of lateral ventricle | X | |
| | Choroid plexus third ventricle | X | |

*More anatomical information is needed before determining whether a lesion at this location is infra- or supratentorial.
*More anatomical information is needed before determining whether a lesion at this location is infra- or supratentorial.

*(Continued on page 14)*

*(Continued from page 13)*

| BRAIN | | | |
|---|---|---|---|
| | | Supratentorial | Infratentorial |
| C71.5 | Ependyma* | | |
| | Lateral ventricle, NOS | X | |
| C71.6 | Cerebellum, NOS | | X |
| | Cerebellopontine angle | | X |
| | Vermis of cerebellum | | X |
| C71.7 | Brain stem | | X |
| | Cerebral pedunculi | | X |
| | Basis pedunculi | | X |
| | Choroid plexus of fourth ventricle | | X |
| | Fourth ventricle, NOS | | X |
| | Infratentorial brain, NOS | | X |
| | Medulla oblongata | | X |
| | Midbrain | | X |
| | Olive | | X |
| | Pons | | X |
| | Pyramid | | X |
| C71.8 | Overlapping lesion of brain | | |
| | Corpus callosum | X | |
| | Tapetum | X | |
| C71.9 | Brain, NOS* | | |
| | Intracranial site* | | |
| | Cranial fossa, NOS* | | |
| | Anterior cranial fossa | X | |
| | Posterior cranial fossa | | X |
| | suprasellar | X | |

*(Continued on page 15)*

*(Continued from page 14)*

| SPINAL CORD AND OTHER CENTRAL NERVOUS SYSTEM | |
|---|---|
| C72.0 | Spinal cord |
| C72.1 | Cauda equina |
| C72.2 | Olfactory nerve |
| C72.3 | Optic nerve |
| C72.4 | Acoustic nerve |
| C72.5 | Cranial nerve, NOS |
| C72.8 | Overlapping lesion of brain and central nervous system |
| C72.9 | Nervous system, NOS |
| **NEUROENDEOCRINE AND RELATED STRUCTURES** | |
| C75.1 | Pituitary gland |
| C75.2 | Craniopharyngeal duct (suprasellar region) |
| C75.3 | Pineal gland |

## ICD-O-3 Coding Issues

For some histologic types, it is difficult to determine if the primary site is intracranial or the skull (C41.0). Tumors that originate in the skull are not intracranial. The malignant tumors are reportable regardless of origin, but nonmalignant tumors that originate in the skull are not reportable.

- Chondroma (9220/0) is a benign tumor of cartilage cells. The ICD-O-3 Manual shows the code for bone in parentheses next to the morphology. Review the record carefully to determine if the tumor originated in bone or in an intracranial site. Because chondroma is a benign tumor, only complete an abstract if the primary tumor is in an intracranial site. A chondroma of the skull is not reportable.
- Chordoma is a malignant tumor arising from the embryonic notochord, and chondrosarcoma (9220/3) is a malignant tumor of cartilage cells. These tumor histologies are reportable, but you must determine if the primary site is bone or an intracranial site because the intracranial tumors are analyzed separately.

When ICD-O-3 was published, the code for pilocytic astrocytoma changed from malignant behavior (3) to borderline behavior (1). Registrars were instructed to continue to assign the code with malignant behavior. For data consistency, continue to assign the malignant behavior code (3) to pilocytic astrocytoma even after nonmalignant CNS cases are collected.

Intracranial schwannoma (9560/0) is reportable for cases diagnosed January 1, 2004 and later. It is difficult to determine the intracranial site of a schwannoma. Assign the primary site for intracranial schwannoma to cranial nerves NOS (C72.5) when the site is not documented in the health record.

### Histological types of Brain Tumors

The 2000 revision of the WHO classification of tumors of the nervous system dropped the terms glial and non-glial as major categories. Instead, tumors are grouped by their tissue of origin. The majority of tumors arise in neuroepithelial tissue, the largest category, that includes astrocytomas and ependymomas. Listed following are the major categories of brain tumor based on WHO classification:

- Tumors of neuroepithelial tissue.

*(Continued on page 16)*

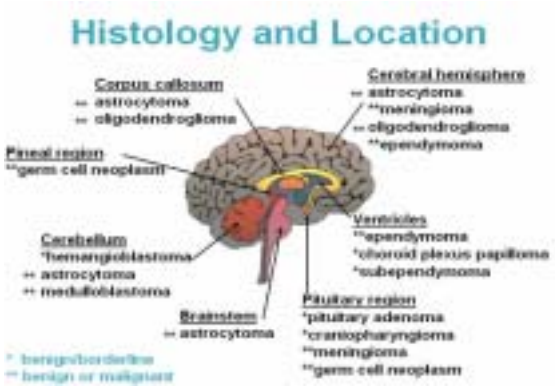
*(Continued from page 15)*

- Tumors of peripheral nerves.
- Tumors of meninges.
- Tumors of sellar region.
- Germ cell tumors.
- Lymphomas.
- Metastatic tumors.

The following table shows some of the brain tumors categoried by their histological types:
Various types of brain tumors can arise in various places in the

| **NEUROEPITHELIAL TYPE** | |
|---|---|
| | **ICD-O Codes** |
| Astrocytic | |
| Pilocytic (juvenile) astrocytoma | M-9421/3 |
| Well-diff. low grade astrocytoma | M-9400/3 |
| Anaplastic astrocytoma | M-9401/3 |
| Glioblastoma multiforme | M-9440/3 |
| **NEUROEPITHELIAL TYPE (CONT'D)** | |
| Ependymoma | M-9391/3 |
| Choroid plexus papilloma | M-9390/3 |
| Oligodendroglioma | M-9450/3 |
| Mixed gliomas | M-9382/3 |
| Medulloblastoma | M-9470/3 |
| Anaplastic angliogliomа | M-9505/3 |
| Pineal parenchymal tumor of intermediate diff. | M-9362/3 |
| **NON-NEUROEPITHELIAL TYPE** | |
| Craniopharyngioma | M-9350/1 |
| Meningioma | M-9539/3 |
| Schwannoma | M-9560/3 |
| Lymphoma | M-9590/3 |

central nervous system. The image below shows most of those sites and their histologies.




Brain

*(Continued on page 17)*

*(Continued from page 16)*

Tumor Grading

The sixth digit of the ICD-O-3 morphology code describes the histologic grade or differentiation of the tumor. The ICD-O-3 grade or differentiation is not always described by pathologists for CNS tumors. When it is not described, assign code 9 (not determined, not stated, or not applicable). The ICD-O-3 grade or differentiation code for nonmalignant CNS tumors is always assigned code 9. This is documented in ICD-O-3, page 30, Rule G, paragraph 1.

Other grading systems used to describe CNS tumors are WHO grade, Kernohan grade, and St. Anne/Mayo grade.

- Kernohan's original description of a four grade system for astrocytomas was published in 1949. It formed the basis for the atypia, necrosis, vascularity, and other factors now used in the WHO classification, although Kernohan grades I and II became WHO grade II.
- The Ringertz classification first published in 1950 is a three grade system where grade I is the same as the WHO classification, but WHO grades II, III and IV are split between Ringertz grades II and III.
- The St Anne/Mayo system from 1988 is also known by the names of its authors, Daumas and Duport. This system is based on four criteria--atypia, necrosis, mitosis and endothelial proliferation. There is a potential score of 0 - 4 in this system.
- The WHO grading system was initially published in 1979, but was modified in 1993 and again most recently in 2000. The WHO system includes all types of central nervous system tumors. It is not criteria based.

WHO grades are not the same as the ICD-O-3 grade or differentiation and are not recorded in the sixth digit histology code data field for grade. The grade is used by the clinician to plan treatment and predict prognosis.

The most important thing for a registrar to understand about the WHO grade for central nervous system tumors is that it does not parallel the ICD-O-3 grade code in the 6th digit of the morphology code. As this graphic shows, Grade I in the WHO system is roughly equivalent to a behavior code of benign in ICD-O-3.

The following tables shows both WHO histologic typing and ICD-O 6th digit Grade.

**WHO Histologic Typing vs. ICD-O 6th Digit Grade**

| Histology | WHO grade | ICD-O 6th digit |
|---|---|---|
| Pilocytic astrocytoma | 1 | 9 |
| W-d low grade astrocytoma | 2 | 1 |
| Anaplastic astrocytoma | 3 | 4 |
| Glioblastoma multiforme | 4 | 9 |

Brain Tumor Staging

Basically, all brain tumors are considered localized unless they cross the midline or the tentorium or unless they are described as having "drop" metastases in the spinal cord.

There was a TNM staging for brain tumors in the fourth edition of the AJCC Manual for Staging of Cancer based heavily on the tumor grade, but this was withdrawn in subsequent editions.

Summary Staging 2000

Local

confined to:
one hemisphere in one part of brain (infra/supratentorial); meninges; invading/encroaching on ventricular system

Regional

crossing midline or tentorium invades bone, blood vessel, nerves, spinal cord

Distant

circulating cells in CSF; extension to nasal cavity, nasopharynx, posterior pharynx; outside CNS

## TNM

None in 5th or 6th editions.

## Collaborative Staging

The Collaborative Staging System uses two fields to code the extent of disease for tumors of the brain and cerebral meninges. The extension field includes a code for benign brain tumors to meet the needs of cases that will be reported as of 2004. The Collaborative Staging code structure is based on what area

*(Continued on page 18)*

*(Continued from page 17)*

of the brain is involved and how far the tumor has spread.

Collaborative Staging -- Extension

| CNS (C70.0, C71._) | |
|---|---|
| 05 | Benign brain tumors |
| 10 | Supratentorial tumor confined to:<br>Cerebral hemisphere (cerebrum) or meninges of cerebral hemisphere on one side:<br>Lobe(s): Frontal -- Occipital -- Parietal -- Temporal |
| 11 | Infratentorial tumor confined to:<br>Cerebellum or meninges of cerebellum on one side:<br>Vermis: Lateral lobes -- Median lobe of cerebellum |
| 12 | Infratentorial tumor confined to:<br>Brain stem or meninges of brain stem on one side:<br>Medulla oblongata -- Midbrain (mesencephalon) --<br>Pons -- Hypothalamus -- Thalamus |
| 15 | Confined to brain, NOS— Confined to meninges, NOS |
| 20 | Infratentorial tumor:<br>Both cerebellum and brain stem involved with tumor on one side |
| 30 | Confined to ventricles<br>Tumor invades or encroaches upon ventricular system |
| 40 | Tumor crosses the midline<br>Tumor involves contralateral hemisphere<br>Tumor 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) |

| CNS (C70.0, C71._) | |
|---|---|
| 60 | Tumor invades:<br>Bone (skull) -- Major blood vessel(s) -- Meninges (dura) -- Nerves, NOS {Cranial nerves} — Spinal cord/canal |
| 70 | Circulating cells in cerebral spinal fluid (CSF)<br>Nasal cavity<br>Nasopharynx<br>Posterior pharynx<br>Outside central nervous system (CSF) |
| 80 | Further contiguous extension |
| 95 | No evidence of primary tumor |
| 99 | Unknown extension<br>Primary tumor cannot be assessed<br>Not documented in patient record |

Determining and Counting Multiple Primaries

| Other CNS (C71.1, C71.9, C72.0-C72.5, C72.8, C72.9) | |
|---|---|
| 05 | Benign brain tumors |
| 10 | Tumor confined to tissue or site of origin |
| 30 | Localized, NOS |
| 40 | Meningeal tumor infiltrates nerve<br>Nerve tumor infiltrates meninges (dura) |
| 50 | Adjacent connective/soft tissue<br>Adjacent muscle |
| 60 | Brain, for cranial nerve tumors<br>Major blood vessel(s)<br>Sphenoid and frontal sinuses (skull) |
| 70 | Brain except for cranial nerve tumors<br>Bone, other than skull — Eye |
| 80 | Further contiguous extention |
| 95 | No evidence of primary tumor |
| 99 | Unknown extension; Primary tumor cannot be assessed; Not documented in record |

In determining multiple primaries, separate rules are used for

*(Continued on page 19)*

*(Continued from page 18)*

nonmalignant and malignant brain tumors. Rules are based on the specific data elements.

**CNS (Endocrine) C75.1-C75.3 (from the Collab. Stage schema for Thymus, Adrenal (Suprarenal) Gland, and Other Endocrine Glands)**

| | |
|---|---|
| 00 | In situ; non-invasive; intraepithelial |
| 05 | For C75.1 pituitary gland, C75.2 craniopharyngeal duct, and C75.3 pineal gland ONLY: Benign |
| 10 | Invasive carcinoma confined to gland of origin |
| 30 | Localized, NOS |
| 40 | Adjacent connective tissue<br>(see definition in General Instructions)<br>Adjacent organs/structures<br><br>Thymus and aortic body:<br>Organs/structures in mediastinum<br><br>Adrenal (suprarenal):<br>Kidney<br>Retroperitoneal structures<br><br>Parathyroid<br>Thyroid<br>Thyroid cartilage |
| 60 | Pituitay and carniopharyngeal duct:<br>Cavernous sinus<br>Infundibulum<br>Pons<br>Sphenoid body and sinuses<br><br>Pineal:<br>Infratentorial and central brain<br><br>Carotid body:<br>Upper neck |
| 80 | Further contiguous extension |
| 95 | No evidence of primary tumor<br>Unknown extension |
| 99 | Primary tumor cannot be assessed<br>Not documented in patient record |

Timing (assuming same site and same histology)

- Malignant: 2 months
- Nonmalignant: no limitation

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

The current 2 month rule does not apply to nonmalignant CNS tumors. Nonmalignant tumors may recur in the same location. If they recur, even after 20 years, they are still the same tumor.

Site

- Malignant: 3 character level
- Nonmalignant: 4 character level

For nonmalignant CNS tumors, sites are different when there is a difference in the 4th digit of the site code, but the first 3 digits are the same. For example, spinal meninges (C70.1) and cerebral meninges (C70.0) are considered different primaries. (The exception is when the difference in the 4th digit occurs because the 4th digit of one site code is 9, which indicates a non-specific code.)

Malignant tumors remain as they are currently defined with differences only at the 3 digit level. Therefore separate malignant tumors occurring in the cerebral meninges and in the spinal meninges (both C70) are not considered different primaries.

Laterality

- Applies to nonmalignant only

Laterality will be used to determine if multiple nonmalignant CNS tumors are counted as multiple primary tumors. Laterality is not used to determine if multiple malignant tumors of the same CNS site are multiple primary tumors.

CNS sites to be coded with laterality:

- Cerebral meninges, NOS (C70.0)
- Cerebrum (C71.0)

*(Continued on page 20)*

*(Continued from page 19)*

- 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, NOS (C72.5)

Code laterality using codes 1 through 4 or 9 (paired site but lateral origin unknown; midline tumor). The laterality for all other CNS sites is coded 0 (not a paired organ).

Histology

- Malignant: 3 digit level
- Nonmalignant: Histology Groups Table

The histology rules for counting multiple primaries have to be modified to count tumors at a level other than the first three digits of the morphology code.

The evolution and grading of brain tumors is such that a tumor may recur at a higher grade which has a different ICD-O-3 code number. In such cases, the new tumor is not counted as a new primary (except if it progress or transforms from benign or borderline to malignant). The various four-digit histologies within each of these histologic groups will be counted as one primary. Thus the patient could have one glioma and one ependymoma, but not a low grade astrocytoma followed by a glioblastoma multiforme at the same site.

For counting nonmalignant primaries, each of the following groups is considered ONE primary.

B = Benign
M = Malignant
* includes gliomas, astrocytomas, astroblastomas and glioblastomas
Please use the two reference tables below to count malignant and nonmalignant primaries.

| | |
|---|---|
| Gliomas* | 9380, 9381, 9382, 9400, 9401, 9410,9411, 9420, 9421, 9423, 9424, 9430, 9440,9441, 9442 |
| Subependymomas | 9383, 9384 |
| Choroid plexus neoplasms | 9390 |
| Ependymomas | 9391, 9392, 9393, 9394, 9444 |
| Neuronal and neuronalglial neoplasms | 9412, 9413, 9505, 9506 |
| Oligodendrogliomas | 9450, 9451, 9460. |

Counting Non-malignant Primaries

Same Histology

| Tumor | | Timing (months) | Same Site | | | Different Site | | |
|---|---|---|---|---|---|---|---|---|
| 1st | 2nd | | Same side | Other side | Unkn side | Same side | Other side | Unkn side |
| B | B | NA | 1 | 2 | 1 | 2 | 2 | 2 |
| B | M | < 2 | 2 | 2 | 2 | 2 | 2 | 2 |
| B | M | 2 + | 2 | 2 | 2 | 2 | 2 | 2 |

Different Histology

| Tumor | | Timing | Same Site | | | Different Site | | |
|---|---|---|---|---|---|---|---|---|
| 1st | 2nd | | Same side | Other side | Unkn side | Same side | Other side | Unkn side |
| B | B | NA | 2 | 2 | 2 | 2 | 2 | 2 |
| B | M | < 2 | 2 | 2 | 2 | 2 | 2 | 2 |
| B | M | 2 + | 2 | 2 | 2 | 2 | 2 | 2 |

Counting Malignant Primaries

Same Histology *unless stated to be metastatic or recurrent

| Tumor | | Timing (months) | Same Site | | | Different Site | | |
|---|---|---|---|---|---|---|---|---|
| 1st | 2nd | | Same side | Other side | Unkn side | Same side | Other side | Unkn side |
| M | M | < 2 | 1 | 1 | 1 | 2* | 2* | 2* |
| M | M | 2 + | 2* | 2* | 2* | 2* | 2* | 2* |
| M | B | NA | 2 | 2 | 2 | 2 | 2 | 2 |

Different Histology **unless one histology is a specific subtype of the other

| Tumor | | Timing | Same Site | | | Different Site | | |
|---|---|---|---|---|---|---|---|---|
| 1st | 2nd | | Same side | Other side | Unkn side | Same side | Other side | Unkn side |
| M | M | < 2 | 2** | 2** | 2** | 2 | 2 | 2 |
| M | M | 2 + | 2 | 2 | 2 | 2 | 2 | 2 |
| M | B | NA | 2 | 2 | 2 | 2 | 2 | 2 |

Malignant Transformation

In some patients, a diagnosed nonmalignant tumor will transform into a malignant tumor, which is a rare occurrence.

Malignant transformation of a nonmalignant tumor or progression of a nonmalignant primary CNS tumor to a malignant tumor can be determined from physician statements in the patient medical record or by pathological review.

Pathologists develop a final diagnosis of malignant transformation or progression from a nonmalignant tumor by review of old slides from previous biopsies, excisions of the nonmalignant tumor, or by review of the newly-biopsied or -resected malignant brain tumor.

Malignant transformation and progression to malignancy mean the same thing. A change in morphology in a tumor from WHO grade I to WHO grade II, III, or IV indicates malignant transformation. When malignant transformation occurs, the tumors are considered separate primaries and two abstracts are completed. Recording these as separate primaries will allow researchers investigating these specific conditions to identify cases.

*(Continued on page 21)*

*(Continued from page 20)*

Use the following table as a reference for determining whether a new abstraction is needed.

| Situation | Create new abstract? |
|---|---|
| Benign /0 to borderline /1 | No* |
| Benign /0 to malignant /3 | Yes |
| Borderline /1 to malignant /3 | Yes |
| Malignant /3 to malignant /3 | No* |
| WHO Grade I to Grade II, III, or IV | No* |
| * Abstract as one primary using original histology and note progression in remarks | |

Coding Sequence Numbers

The sequence number indicates the sequence of all reportable neoplasms over the lifetime of the patient and it is recorded in the sequence number data field.
Malignant and in situ neoplasms, including malignant CNS neoplasms, are assigned codes 00 through 35. If a patient has only one primary malignant or in situ neoplasm, the sequence number assigned is 00. If a patient has multiple primary neoplasms during a 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 60 to 87 range. The first nonmalignant tumor will be coded 60 and a subsequent nonmalignant tumor would be coded 61.

If a patient was diagnosed with a nonmalignant CNS neoplasm before reporting was required (January 1, 2004 and earlier) and is diagnosed with a second nonmalignant CNS neoplasm in 2004, the first neoplasm is assigned sequence number 61 and the second neoplasm is assigned sequence number 62 even though an abstract is required only for the second neoplasm.

When a nonmalignant CNS tumor progresses into a malignant CNS tumor, sequencing of nonmalignant does not affect sequencing of malignant. For example, if a patient had a nonmalignant CNS tumor that progressed into a malignant CNS tumor, the sequence number for the benign tumor is 60, and the sequence number for the malignant tumor is 00.

Date of Diagnosis

The same rules are used to assign diagnosis date for both nonmalignant and malignant CNS tumors.

The rules are found in Commission on Cancer's Facility Oncology Data Standards (FORDS), Section 2: Coding Instructions, pages 89 and 90. It is not unusual for a patient with a nonmalignant CNS tumor to be diagnosed in a physician's office and treated with watchful waiting. Several years may go by before the patient receives subsequent treatment at a health care facility in the form of surgery or radiation therapy or some type of systemic therapy. Also, nonmalignant CNS tumors, especially meningiomas, often recur. The date of initial diagnosis should be recorded in the abstract, not the date of subsequent treatment or date of recurrence. Health records must be reviewed carefully to determine the initial date of diagnosis by a medical practitioner, regardless if the initial diagnosis was clinical or histologic.

Standard Treatment

Generally the treatment of choice is surgery unless the tumor is in an inaccessible or delicate area, such as in speech, vision, or motor control area. Some tumors are so aggressive that they also need radiation therapy.

S = Surgery
R = Radiation therapy
C = Chemotherapy
( ) = Optional treatments

Surgery Codes

| | |
|---|---|
| Astrocytoma, noninfil | **S(+R)** |
| Astrocytoma, anaplastic | **S+R** |
| Astrocytoma, high grade | **S+R(+C)** |
| Glioblastoma multiforme | **S+R+C** |
| Brain stem glioma | **R** |
| Ependymoma, w-d | **S(+R)** |
| Ependymoma, anaplastic | **S+R** |
| Oligodendroglioma | **S(+R)** |
| Oligodendroglioma, anaplastic | **S+R(+C)** |
| Mixed glioma | **S+R(+C)** |
| Medulloblastoma | **S(+R)** |
| Pineal parenchymal tumor | **S+R(+C)** |
| CNS germ cell tumor | **S(+R)** |
| Craniopharyngioma | **S(+R)** |
| Meningioma | **S(+R)** |
| Meningioma, malignant | **S+R** |

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CNS sites included in brain related sites fall under 2 separate surgery schemes. BRAIN and ALL Others. The Brain codes include the brain and spinal cord as well as the meninges. The Other Sites include the pitutitary and pineal glands & the craniopharyngeal duct.

The following Surgical Procedure of Primary Site codes are used when the primary site is meninges, brain, spinal cord, cranial nerves, or other parts of the CNS. (C70-0-C70.9, C71.0-C71.9, C72.0-C72.9)

- 10 -- Tumor destruction NOS
- 20 -- Local Tumor Excision, NOS
- 40 -- Partial resection
- 55 -- Gross total resection
- 90 -- Surgery, NOS

The following Surgical Procedure of Primary Site codes are used when the site is pituitary gland (C75.1), craniopharyngeal duct (C75.2), or pineal gland (C75.3). They are the surgery codes used for all other sites.

- 10 -- Local tumor destruction, NOS
- 20 -- Local tumor excision, NOS
- 40 -- total surgical removal of the primary site
- 50 -- Surgery stated to be "debulking"
- 60 --Radical surgery 90 -- Surgery, NOS

Other surgical data fields are completed for CNS tumors as they are for other malignant primaries. Directions for coding the surgical data fields can be found in FORDS manual.

Other Treatments

Cutting Edge Treatment

There have been enormous advances in improving the success and reducing the morbidity of surgery and radiation therapy thanks to computerized tomography.

The terms gamma knife, cyber knife and x-knife are used by different companies for their products. When a tumor is identified, treatment is planned so that Cobalt 60 beams will converge on a small area (usually less than 3 cm) from many directions, thereby minimizing the damage to normal tissue surrounding the tumor and maximizing the radiation dose to the tumor itself. The patient is placed in a helmet-like head holder called a collimater and then slides into the treatment machine. Usually this is a one-time outpatient treatment.

Investigational Treatment

- Radiosensitizers
- Chemotherapy
  angiogenesis inhibitors
  growth factor inhibitors
- Immunotherapy
  monoclonal antibodies
  radioisotope-tagged monoclonal antibodies

The above types of therapies are under clinical investigation as part of clinical trials. Most of these therapies do not kill cells by

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themselves, but work with other therapies.

Radiosensitizers enhance the effect of radiotherapy. Angiogenesis inhibitors attempt to block the tumor's ability to create its own blood vessels to bring extra nourishment to the tumor. Growth factor inhibitors work in a similar manner to stop certain parts of the tumor from growing. Immunotherapy helps deliver antineoplastic agents to the tumor itself minimizing damage to surrounding normal tissue.

Systemic Agents

Systemic agents rather than chemotherapy are used because some of these drugs do not fit the registry definition of chemotherapy.

- BCNU (Carmustine)
- CCNU (Lomustine)
- CPT-11 (Camptosar)
- Melphalan
- Methotrexate
- Procarbazine (Matulane)
- Tamoxifen (Nolvadex)
- Temodar (Temozolomide)
- Thalidomide
- Thiotepa
- Vincristine

Standard chemotherapy has not been terribly successful due to the brain's natural blood-brain barrier which blocks large molecules from entering the vascular system in the brain.

Some drugs have proven effective against neural tumors, particularly the nitrosoureas BCNU and CCNU. Newer drugs under clinical investigation include Camptosar, Temodar, and even Tamoxifen. Older drugs like melphalan, vincristine and procarbazine have been given second opportunities to prove their effectiveness in combination therapies. Thalidomide has made a comeback as an antiangiogenesis agent in clinical trials. Tamoxifen has been used to treat recurrent meningioma.

## FDA Approves Erbitux for Colorectal Cancer

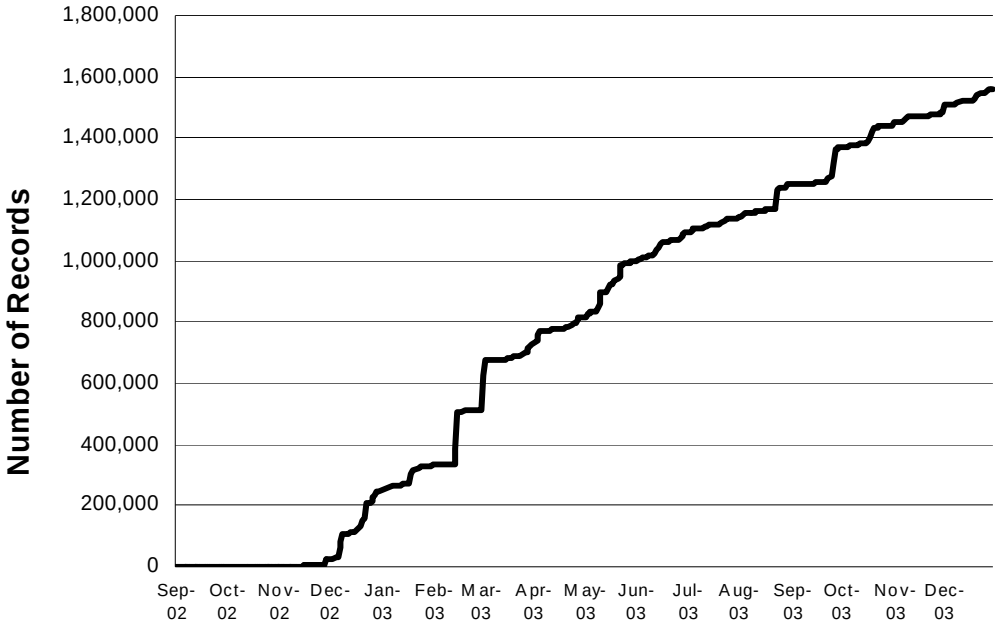
http://www.fda.gov/bbs/topics/NEWS/2004/NEW01024.html

FDA approved Erbitux (cetuximab), February 12, 2004 to treat patients with advanced colorectal cancer that has spread to other parts of the body. Erbitux is the first monoclonal antibody approved to treat this type of cancer and is indicated as a combination treatment to be given intravenously with irinotecan, another drug approved to fight colorectal cancer, or alone if patients cannot tolerate irinotecan.

FLORIDA CANCER DATA SYSTEM

# Path Reporting

## Cumulative Path Data Received

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