# Accessioning Primary Intracranial and Central Nervous System Tumors
# General Reporting Rules

**Prepared by the NAACCR Registry Operations Committee Benign Brain Tumor Subcommittee**
**Reviewed and Approved by NAACCR Uniform Data Standards Committee July 2, 2003**

## Introduction

In the early 1900's, the neurosurgeon Harvey Cushing made the observation that some brain tumors are malignant because of their histology, and some are malignant because of their location. By this he meant that in the early 1900's some tumors were not resectable and would result in the death of the patient because of mass effects on vital areas of the brain. In the past 100 years, with advances in microsurgery, radiation therapy, and earlier diagnosis, the maxim of Dr. Cushing still stands, although at a greatly diminished number. The tumors, whether benign or malignant, produce clinical effects by similar mechanisms of mass effect, hemorrhage, seizure activity, and edema. Although these tumors are individually rare, patients with benign brain tumors represent an under-appreciated financial and health burden in the United States. These cases include those tumors arising in families with an inherited tendency to develop benign and malignant brain tumors, tumors arising from developmental abnormalities, morbidity from ruptured benign brain tumors, and eventual malignant transformation in a subgroup of patients with optic nerve gliomas.

Existing coding rules for brain and CNS tumors have been guided by the behavior of these tumors. With the change to a site definition to guide their collection, the ROC Benign Brain Tumor Subcommittee reviewed coding rules applicable to both nonmalignant and malignant brain and CNS tumors. Recommendations applicable to the current rules guiding multiple primaries for malignant brain and CNS tumors are contained in a separate document and have been forwarded to the SEER Histology Coding Committee for review in 2003.

**Rules for Benign Brain Tumors**

Effective with cases diagnosed January 2004 and after

(Note: the rules for malignant brain tumors follow the same rules for multiple primaries that have been in effect, but are presented with those for non-malignant brain tumors for ease of use.)

Beginning with tumors diagnosed on or after January 1, 2004, reportable tumors required to be abstracted include non-malignant primary intracranial and central nervous system tumors in ICD-O-3 with a behavior code of /0 or /1 (benign and borderline, or "non-malignant") regardless of histologic type, for the following ICD-O-3 topography codes.

**Table 1. Topography Codes for Benign Brain Tumors**

| Codes | Description |
|---|---|
| | Meninges |
| C70.0 | Cerebral meninges |
| C70.1 | Spinal meninges |
| C70.9 | Meninges, NOS |
| | Brain |
| C71.0 | Cerebrum |
| C71.1 | Frontal lobe |
| C71.2 | Temporal lobe |
| C71.3 | Parietal lobe |
| C71.4 | Occipital lobe |
| C71.5 | Ventricle, NOS |
| C71.6 | Cerebellum, NOS |
| C71.7 | Brain stem |
| C71.8 | Overlapping lesion of brain |
| C71.9 | Brain, NOS |
| | Spinal Cord, Cranial Nerves and Other Parts of the 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 |
| | Other Endocrine Glands and Related Structures |
| C75.1 | Pituitary gland |
| C75.2 | Craniopharyngeal duct |
| C75.3 | Pineal gland |

- For non-malignant primary intracranial and central nervous system tumors (C70.0 – C72.9, C75.1 – C75.3), the terms "tumor" and "neoplasm" are considered diagnostic for the purpose of case reporting, in addition to the terms generally applicable to malignant tumors.

**I. Definitions:**

A. Non-malignant: behavior code of /0 or /1.

B. Malignant: behavior code of /2 or /3.

C. Same Site
   1. Non-malignant: same 4 digit site
      *Exception:* 4 digit NOS site code (C70.9, C71.9, C72.9) with specific 4-digit site code in same rubric
      *Example:* meninges, NOS (C70.9) with spinal meninges (C70.1) or cerebral meninges (C70.0) is the same site

   2. Malignant: same 3 digit site

D. Different site
   1. Non-malignant: different 4 digit site code
      *Exception:* 4 digit NOS site code (C70.9, C71.9, C72.9) with specific 4-digit site code in same rubric
      *Example of exception:* Brain stem (C71.7) with intracranial site (C71.9) is the *same* site.

   2. Malignant: different 3 digit site

E. Same histology
   1. Non-malignant (in priority order):
      a) Use Table 2 listed under II.D. in this document – if both histologies are in the same histologic group, then same histology
      b) If same first 3 digits as any histology in Table 2, then same histology
      c) If same first 3 digits but neither in Table 2, then same histology

   2. Malignant (current rule): same at 3 digit level

F. Different histology
   1. Non-malignant:
      a. If 2 different histologic groups in Table 2
      b. If different at 3 digit level and not in same group in Table 2
      c. If different at 3 digit level and neither in Table 2, then different histology

   2. Malignant (current rule): different at 3 digit level

G. Timing
   1. Non-malignant: current 2-month timing rule does not apply.

   2. Malignant:
      a. Within 2 months
      b. 2+ months

H. Laterality:
   1. Single side (SS): involves only one side of sites listed in Section III, A.
   2. Both sides (BS): involves both sides of sites listed in Section III, A.
   3. Laterality unknown (LX): Site does not have laterality coded or laterality is not coded for site

**II. General Rules for Determining Multiple Primaries: The following rules apply for defining multiple primaries for non-malignant and malignant primary intracranial and central nervous system tumors (C70.0 – C72.9, C75.1 – C75.3).**

*Rationales for multiple primaries rules*:

1. The natural biology of non-malignant tumors is that of expansive, localized growth, with local recurrences common, and metastasis uncommon or unusual.
2. Non-malignant tumors of the same histology, same site, and same side will recur in the same location. If they recur, even after 20 years, they are still the same tumor.
3. The corollary to statement 2 is that multiple non-malignant tumors of the same histology identified in different locations or sides of the CNS should be considered separate primaries.

A. Multiple lesions in which all are non-malignant tumors
   1. If different sites, then separate primaries
   2. If different histologies, then separate primaries
   3. If same site and same histology*:
      a. and laterality is same side, one side unknown or not applicable, then single primary
      b. and laterality is both sides, then separate primaries
         * Note: if two histologies are in the same group in Table 2, code the more specific histology

B. Multiple tumors in which one was non-malignant and the other was a malignant lesion
   1. Non-malignant tumor followed by malignant tumor: separate primaries regardless of timing
   2. Malignant tumor followed by a non-malignant tumor : separate primaries regardless of timing

C. Multiple malignant tumors
   1. If same histology:
      a. < 2 months:
         i. 1 if same site
         ii. 2 if different site and not stated to be a recurrence or metastases
      b. 2+ months (site does not matter):
         i. 2 unless stated to be a recurrence or metastases
   2. If different histologies:
      a. <2 months:
         i. 2 if same site unless one is more specific histology
         ii. 2 if different site
      b. 2+ months:
         i. always 2 primaries

D. **Table 2. Histologic groupings to determine same histology for non-malignant brain tumors**

| | |
|---|---|
| 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 neuronal-glial neoplasms | 9412, 9413, 9505, 9506 |
| Oligodendrogliomas | 9450, 9451, 9460 |

* includes gliomas, astrocytomas, astroblastomas, and glioblastomas

*Rationale:* Brain tumor histologies grouped in Table 2 do not follow the standard 3-digit histology difference rule because they represent a progression, differentiation or subtype of a single histologic category.

In a review of the ICD-O histology codes, applying the current 3 digit histology rule to non-malignant tumors would combine tumors that are no longer considered to be biologically related.

## III. Collection of additional data

A. Laterality

Beginning with malignant and benign/borderline tumors diagnosed in 2004, the following sites require a laterality code of 1-4, or 9 (The NAACCR Uniform Data Standards Committee has approved this coding change.):

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
C72.5 Cranial nerve, NOS

The research community has indicated that the location and laterality for primary CNS tumors is of significant interest in determining causation and assessing the impact on quality of life. With respect to CNS tumors diagnosed prior to 2004: Primary brain and CNS tumors were traditionally reported with a laterality code 0, although some registries recorded laterality codes "by agreement" for these sites. Therefore, EDITS will allow CNS tumors diagnosed prior to 2004 to have laterality codes of 0, 1-4, or 9.

*Rationale:* a. Laterality is needed to determine multiple primaries for benign brain tumors.

b. Researchers, including epidemiologists, have requested the collection of laterality *(Inskip PD, Neuroepidemiology 2003; 22;130-138*). The location of certain tumors might help in determining causation. Certain investigations such as those involving cell phone usage would benefit from having this variable routinely available.

c. Non-treatment-related factors such as location of tumor by hemisphere can be predictive factors for cognitive outcome. Brown PD, Buckner, JC, Uhm JH, and Shaw EG (2003) The neurocognitive effects of radiation in adult low-grade glioma patients. *Neuro-Oncology* 5, 161-167, 2003.

B. WHO Grade Code

This item is to be coded in Site Specific Factor 1 of the Collaborative Staging System for Brain and other Central Nervous System sites.

WHO Grade I - Code 010 in Collaborative Staging System
WHO Grade II - Code 020
WHO Grade III - Code 030
WHO Grade IV - Code 040
WHO Grade unknown - Code 999

WHO grade I generally describes non-malignant or benign tumors; however, non-malignant tumors should not be coded as Grade I unless WHO grade is specifically stated in the source document.
WHO grade II generally describes a malignant tumor but it can describe a non-malignant tumor depending on histologic type.
WHO grade III and IV describe malignant tumors.
For certain types of CNS tumors, no WHO grade is assigned.

C. Reportability/Sequence number

1. Non-malignant: a primary non-malignant tumor of any of the sites specified diagnosed *on or after* January 1, 2004, is reportable. The sequence number for the tumor is in the range 60 – 87.

   Non-malignant tumors diagnosed before January 1, 2004 should be included in the lifetime sequence of non-malignant and borderline tumors in the range 60-87.

   A primary non-malignant tumor of any of the sites specified diagnosed *before* January 1, 2004, is not reportable unless there are specific preexisting regional or state reporting requirements.

   *Rationale:* To clarify reporting implementation date and sequence rules for non-malignant tumors.

2. Malignant: the sequence number for the malignancy is in the range 00-35.

3. The sequencing of non-malignant tumors does not affect the sequencing of malignant tumors, and vice versa. For example, a first malignancy (sequence 00) will remain sequence 00 if followed by a non-malignant tumor (sequence 60-87)

## IV. Analysis/Reporting of Brain and CNS Tumors:

The ROC Benign Brain Tumor Subcommittee **recommends** that non-malignant and malignant brain tumors be reported separately with a footnote that pilocytic astrocytomas are included in analysis for malignant brain tumors for continuity of trends.

**We recommend** reviewing the standard site and histology groupings for tabulating estimates of these tumors to allow comparability of information across registries.

**We recommend** that training for reporting and tabulating primary intracranial and CNS tumors be offered on a regular basis.

### Registry Operations Committee Benign Brain Tumor Subcommittee

**Members:**

| | |
|---|---|
| Susan Bolick-Aldrich (Chair) | SC Central Cancer Registry, Registry Operations Committee Co-Chair |
| Trista Aarnes-Leong | St. Vincent Medical Center, Los Angeles |
| Gayle Clutter | National Program of Cancer Registries, CDC |
| April Fritz | SEER Program, NCI |
| Susan Gershman, PhD | Massachusetts Cancer Registry, Registry Operations Committee Co-Chair |
| Bette Smith | Ohio Cancer Incidence Surveillance System |
| Carol Kruchko | Central Brain Tumor Registry of the US |
| Bridget McCarthy, PhD | Central Brain Tumor Registry of the US |
| Roger McLendon, MD | Neuropathologist, Duke University Medical Center, Durham, NC |
| Fran Michaud | National Program of Cancer Registries, CDC |
| Eileen Morgan | Duke University Medical Center Cancer Registry, Durham, NC |
| Donna Morrell | USC School of Medicine Cancer Surveillance Program, Los Angeles |
| Jerri Linn Phillips | American College of Surgeons, Commission on Cancer |
| Katheryne Vance | California Cancer Registry |
| Shannon Vann | Independent contractor, NAACCR |
| Claudia Feight | Oregon State Cancer Registry |
| James Gurney, PhD | University of Minnesota Division of Pediatric Epidemiology |
| Elaine Hamlyn | Canadian Cancer Registry, Statistics Canada |
| Linda Mulvihill | North Carolina Central Cancer Registry |
| James Smirniotopoulos, MD | Dept of Radiology & Radiological Sciences, Uniform Services University of the Health Sciences |
| Valerie Vesich | American College of Surgeons, Commission on Cancer |