Benign Brain Tumor Reporting:
2004 Data Changes
2004 Data Changes for Benign/Borderline Brain Tumors

• Casefinding Lists
• Definitions
• Timing
• Laterality
• Multiple Primaries
  – Transformation
• Sequencing
Public Law 107-260

- Signed 10/29/2002
- Effective with 01/01/2004 diagnosis
- Benign Brain Tumor Cancer Registries Amendment Act requires that state-wide cancer registries funded by NPCR collect data for benign and borderline tumors.
- SEER will also begin collecting benign and borderline CNS tumors.
PL-107-260

• CoC has added benign and borderline CNS tumors to the case eligibility definition in the FORDS 2004 manual.
How Does this Change State Reporting?

- CCR has been collecting benign and borderline CNS tumors since 1/1/2001
- CCR was represented on the NAACCR Benign Brain Tumor Subcommittee charged with defining the national reportability rules
Changes in Reporting

• Expanded definition of reportable histologies
• Definition of multiple primaries
• Coding laterality
ICD-9 Casefinding Codes

225 Benign neoplasms of brain and other parts of central nervous system
  225.0 Brain
  225.1 Cranial nerves
  225.2 Cerebral meninges; meninges, NOS; meningioma
  225.3 Spinal Cord; cauda equina
  225.4 Spinal meninges
  225.8 Other specified sites of nervous system
  225.9 Nervous system part unspecified
ICD-9 Casefinding Codes

227.3 Benign neoplasm of pituitary gland and craniopharyngeal duct (pouch)
227.4 Benign neoplasm of pineal gland/body
237 Neoplasm of uncertain behavior of endocrine glands and nervous system
237.0 Pituitary gland and craniopharyngeal duct
237.1 Pineal gland
237.5 Brain and spinal cord
237.6 Meninges (NOS, cerebral, spinal)
ICD-9 Casefinding Codes

237.7 Neurofibromatosis
  237.70 Unspecified
  237.71 Type I (von Recklinghausen’s disease)
  237.72 Type II (acoustic neurofibromatosis)
  237.9 Other and unspecified parts of nervous system;
    cranial nerves
ICD-10 Casefinding Codes
(death certificates)

D32    Benign neoplasm of meninges
D32.0 Cerebral meninges
D32.1 Spinal meninges
D32.9 Meninges, unspecified/NOS
D33    Benign neoplasm of brain and other parts of central nervous system
D33.0 Brain, supratentorial
D33.1 Brain, infratentorial
D33.2 Brain, unspecified
D33.3 Cranial nerves
D33.4 Spinal cord
<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>D33.7</td>
<td>Other specified sites of nervous system</td>
</tr>
<tr>
<td>D33.9</td>
<td>Central nervous system, part unspecified</td>
</tr>
<tr>
<td>D35</td>
<td>Benign neoplasm of other and unspecified endocrine glands</td>
</tr>
<tr>
<td>D35.2</td>
<td>Pituitary gland</td>
</tr>
<tr>
<td>D35.3</td>
<td>Craniopharyngeal duct</td>
</tr>
<tr>
<td>D35.4</td>
<td>Pineal gland</td>
</tr>
<tr>
<td>D42</td>
<td>Benign neoplasm of meninges</td>
</tr>
<tr>
<td>D42.0</td>
<td>Cerebral meninges</td>
</tr>
<tr>
<td>D42.1</td>
<td>Spinal meninges</td>
</tr>
<tr>
<td>D42.9</td>
<td>Meninges, unspecified/NOS</td>
</tr>
</tbody>
</table>
ICD-10 Casefinding Codes
(death certificates)

Q85.1 Neurofibromatosis (non-malignant); (von Recklinghausen’s disease)
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 specified sites of nervous system
ICD-10 Casefinding Codes
(death certificate)

D43.9 Central nervous system, part unspecified
D44 Neoplasm of uncertain or unknown behavior of endocrine glands
D44.3 Pituitary gland
D44.4 Craniopharyngeal duct
D44.5 Pineal gland
Glial Tumors

- Astrocytic tumors
- Ependymal tumors
- Oligodendroglial tumors
- Mixed Tumors
- Other Gliomas
  >Ganglioneuromas
  >Optic Nerve Gliomas
Non-Glial Tumors

- Pineal region tumors
  1. Parenchymal tumors
  2. Germ cell tumors
- Meningioma
- Choroid plexus tumors
Other CNS Tumors

• Craniopharyngioma
  Rathke Pouch Tumor

• Chordomas
  *Chordomas that arise in the bone are not reportable*

• Schwannoma
  Acoustic Schwannoma/Neuroma
  aka “vestibular schwannoma”
Other CNS Tumors

- Embryonal tumors
- Lymphomas
- Vascular tumors
- Cysts and Tumor-like lesions

Reportable: Dermoid Cyst
Granular cell tumor
Rathke pouch tumor
Other CNS Tumors

- Cysts and Tumor-like lesions
  
  Not-reportable: Epidermoid cyst
  Colloid cyst
  Enterogenous cyst
  Neuroglial cyst
  Plasma cell granuloma
  Nasal glial herterotopia
  Rathke cleft cyst
ICD-0-3 Coding

• Some histologies may be difficult to determine if the primary site is intracranial or the skull (C41.0)
• Non-malignant tumors of the skull are not reportable.
• Chondroma must originate in a CNS related site to be reportable
ICD-0-3 Coding

• Continue to assign histology code 9421/3 to pilocytic astrocytoma

• When the primary site for intracranial schwannoma (9560/0) is not documented in source documents, site should be coded to cranial nerves, NOS (C72.5)
ICD-0-3 Coding

9530/1

Multiple Meningiomas

• Does NOT apply to multiple or sequential malignant meningiomas with behavior /3
• Multiple meningiomas or meningiomatosis is a rare condition strongly associated with neurofibromatosis type 2 and other genetic disorders. 1-2% of all meningiomas
Ambiguous Terminology

- If the final pathologic diagnosis is a CNS neoplasm or mass, there must be an ICD-0-3 histology code for the case to be reportable.
- “Hypodense mass” or “cystic neoplasm” are not reportable even for CNS sites.
Coding Grade for Non-Malignant CNS Tumors

- Always assign code 9 for non-malignant tumors
- Not the same as WHO grade
Determining Multiple Primaries

- Malignant CNS tumors - NO CHANGE
- Non-malignant CNS tumors - NEW RULES
Determining Multiple Primaries

• Requires review of:
  – Sites
  – Histologies
  – Timing
  – Laterality
Determining Multiple Primaries
Definitions

• Different site: Non-malignant
  - different at the subsite level

  Example: Meningioma of cervical spine dura (C70.1) and separate meningioma overlying occipital lobe (C70.0, cerebral meninges). Count and abstract as separate primaries.
Determining Multiple Primaries

Definitions

Exception: NOS (C___.9) with specific 3-digit site code in same rubric

*Example*: Meninges, NOS (C70.9) with spinal meninges (C70.1) or cerebral meninges (C70.0)
Determining Multiple Primaries
Definitions

- Same site- Malignant
  - Different at the site level

*Example*: A malignant tumor in the parietal lobe (C71.3) and a separate malignant tumor in the frontal lobe (C71.1). Abstract as one primary.
# Determining Multiple Primaries: Histology – Non-Malignant

## Table 2

<table>
<thead>
<tr>
<th>Histologic Type</th>
<th>ICD-O-3 Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Choroid plexus neoplasms</td>
<td>9390/0, 9390/1</td>
</tr>
<tr>
<td>Ependymomas</td>
<td>9383, 9394, 9444</td>
</tr>
<tr>
<td>Neuronal and neuronal-glial neoplasms</td>
<td>9384, 9412, 9413, 9442, 9505/1, 9506</td>
</tr>
<tr>
<td>Neurofibromas</td>
<td>9540/0, 9540/1, 9541, 9550, 9560/0</td>
</tr>
<tr>
<td>Neurinomatosis</td>
<td>9560/1</td>
</tr>
<tr>
<td>Neurothekeoma</td>
<td>9562</td>
</tr>
<tr>
<td>Neuroma</td>
<td>9570</td>
</tr>
<tr>
<td>Perineurioma, NOS</td>
<td>9571/0</td>
</tr>
</tbody>
</table>
Determining Multiple Primaries: Histology – Non-Malignant

Same Histology: Non-malignant

- Refer to Table 2 and use the following rules in priority order:

  a) If all histologies are in the same histologic group in Table 2, then the histology is the same.

  Example: 9384 and 9442 = same histology. One Primary
Determining Multiple Primaries: Histology – Non-Malignant cont’d

b) If the first 3 digits are the same as the first 3 digits of any histology group in Table 2, then the histology is the same.

Example: A ganglioma (9505/1) of the cerebrum (C71.6) and a neurocytoma (9506/1) of the cerebellopontine angle (C71.6). One primary
c) If the first 3 digits are the same but the codes are not found in Table 2, then the histology is the same.

Example: Clear cell meningioma (9538/1) of the cerebral meninges and a separate transitional meningioma (9537/0) in another part of the same hemisphere. One primary
Determining Multiple Primaries
Non-Malignant – Histology

Summary

Apply these rules in order:

1. If histology is in any histologic group of Table 2 – same histology
2. If first 3 digits match any histology group in Table 2 – same histology
3. If first 3 digits of histology same, but not in Table 2 – same histology
Determining Multiple Primaries: Histology - Malignant

Different Histology: Malignant

Current rule: Differences in histologic type refer to differences in the FIRST THREE digits of the morphology code
Determining Multiple Primaries

Timing

- Non-malignant CNS tumors - no timing rule

If a new non-malignant tumor of the same histology as an earlier one and diagnosed in the same site is diagnosed subsequently at any time, it is considered to be the same primary
Determining Multiple Primaries Timing

- Primary malignant CNS tumors
  1. Diagnosed in the same site within 2 months: tumors are counted as one primary
  2. Malignant tumors of the same site, diagnosed more than 2 months apart: tumors are counted as separate primaries
Determining Multiple Primaries: Laterality

- Brain is not a paired organ
- Laterality collected on both non-malignant and malignant tumors
- Used to determine if multiple non-malignant CNS tumors are counted as multiple primary tumors
Determining Multiple Primaries: Laterality

• Laterality is collected for malignant tumors. However, it is NOT used to determine if multiple malignant tumors of the same intracranial or CNS site are multiple primary tumors.
CNS sites to be coded with laterality:
Cerebral meninges, NOS (C70.0)
Cerebrum (C71.0)
Frontal lobe (C71.1)
Temporal lobe (C71.2)
Parietal lobe (C71.3)
Occipital lobe (C71.4)
Coding Laterality (continued)

Olfactory nerve (C72.2)
Optic nerve (C72.3)
Acoustic nerve (C72.4)
Cranial nerve, NOS (C72.5)
General Rules for Determining Multiple Primaries of CNS Sites

REVIEW
A. Multiple lesions in which all are non-malignant
   1. If different site (subsites), then separate primaries
   2. If different histologies, then separate primaries
General Rules for Determining Multiple Primaries of CNS Sites

- 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 same group in Table 2, code the 1st or more specific histology
General Rules for Determining Multiple Primaries of CNS Sites

• B. Multiple tumors 1 non-malignant and 1 malignant

  1. Non-malignant tumor followed by malignant tumor: separate primary regardless of timing

  2. Malignant tumor followed by a non-malignant tumor: separate primary regardless of timing

CCR 2004 Data Changes
### Counting Non-malignant Primaries

#### Same Histology

<table>
<thead>
<tr>
<th>Tumor</th>
<th>Timing (months)</th>
<th>1st</th>
<th>2nd</th>
<th>Same Site</th>
<th>Different Site</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>1st</td>
<td>2nd</td>
<td>Same side</td>
<td>Other side</td>
</tr>
<tr>
<td>B B</td>
<td>NA</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>B M</td>
<td>&lt; 2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>B M</td>
<td>2 +</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
</tbody>
</table>

#### Different Histology

<table>
<thead>
<tr>
<th>Tumor</th>
<th>Timing</th>
<th>1st</th>
<th>2nd</th>
<th>Same Site</th>
<th>Different Site</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>1st</td>
<td>2nd</td>
<td>Same side</td>
<td>Other side</td>
</tr>
<tr>
<td>B B</td>
<td>NA</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>B M</td>
<td>&lt; 2</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
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<tr>
<td>B M</td>
<td>2 +</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
</tbody>
</table>
# Counting Malignant Primaries

**Same Histology**  * unless stated to be metastatic or recurrent

<table>
<thead>
<tr>
<th>Tumor</th>
<th>Timing (months)</th>
<th>Same Site</th>
<th>Different Site</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Same side</td>
<td>Other side</td>
</tr>
<tr>
<td>1st</td>
<td>2nd</td>
<td></td>
<td></td>
</tr>
<tr>
<td>M</td>
<td>M</td>
<td>&lt;2</td>
<td>1</td>
</tr>
<tr>
<td>M</td>
<td>M</td>
<td>2 +</td>
<td>2*</td>
</tr>
<tr>
<td>M</td>
<td>B</td>
<td>NA</td>
<td>2</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Tumor</th>
<th>Timing</th>
<th>Same Site</th>
<th>Different Site</th>
</tr>
</thead>
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<tr>
<td></td>
<td></td>
<td>Same side</td>
<td>Other side</td>
</tr>
<tr>
<td>1st</td>
<td>2nd</td>
<td></td>
<td></td>
</tr>
<tr>
<td>M</td>
<td>M</td>
<td>&lt;2</td>
<td>2**</td>
</tr>
<tr>
<td>M</td>
<td>M</td>
<td>2 +</td>
<td>2</td>
</tr>
<tr>
<td>M</td>
<td>B</td>
<td>NA</td>
<td>2</td>
</tr>
</tbody>
</table>

CCR 2004 Data Changes
Malignant Transformation

- If a malignant CNS tumor recurs (transforms) as a higher grade tumor
  - SAME tumor
  - Do not change histology or grade
  - Do not abstract as new primary
Malignant Transformation

- Transformation of a non-malignant tumor to a malignant tumor is a rare occurrence.
- A change from a WHO grade I to a WHO grade II, III, or IV indicates malignant transformation.
- Complete 2 abstracts:
  - 1 for the non-malignant tumor
  - 1 for the malignant tumor
Malignant Transformation
Sequence Number

- Non-malignant tumors: assigned sequence numbers from the reportable-by-agreement series (60-87)

- Malignant tumors: assigned sequence numbers from the malignant series (00-35)
Malignant Transformation
Sequence Number

• The sequencing of non-malignant tumors does not affect the sequencing of malignant tumors, and vice versa.
  – Example: A first malignancy (sequence 00) will remain sequence 00 if followed by a non-malignant tumor (sequence 60-87)
Malignant Transformation Sequence Number

- Example: Patient has non-malignant CNS tumor that progressed into a malignant CNS tumor
- Non-malignant tumor is sequenced 60,
- Malignant tumor is sequenced 00
Malignant Transformation

• Diagnosis dates for non-malignant tumors:
  1st date that a medical practitioner
diagnosed the non-malignant tumor either
  clinically or histologically

• Diagnosis dates for malignant tumors:
  1st date that a medical practitioner
diagnosed the malignant transformation
  either clinically or histologically
## Malignant Transformation

<table>
<thead>
<tr>
<th>Situation</th>
<th>Create new abstract?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign /0 to borderline /1</td>
<td>No*</td>
</tr>
<tr>
<td>Benign /0 to malignant /3</td>
<td>Yes</td>
</tr>
<tr>
<td>Borderline /1 to malignant /3</td>
<td>Yes</td>
</tr>
<tr>
<td>Malignant /3 to malignant /3</td>
<td>No*</td>
</tr>
<tr>
<td>WHO Grade I to Grade II, III, or IV</td>
<td>Yes</td>
</tr>
<tr>
<td>WHO Grade II to III or IV</td>
<td>No*</td>
</tr>
<tr>
<td>WHO Grade III to IV</td>
<td>No*</td>
</tr>
</tbody>
</table>

* Abstract as one primary using original histology and note progression in remarks.
Collaborative Stage Extension

- For all Benign or Borderline Brain Tumors code CS Ext to Code 05
Collaborative Stage
Lymph Nodes

- Brain, Cerebral Meninges, and Other CNS (C70.0, C70.1, C70.9, C71_, C72.0, C72.5, C72.8, C72.9): Code 88/ Not applicable

- Pituitary gland, craniopharyngeal duct, and pineal gland (C75.1, C75.2, C75.3): Code 99/ Not applicable
Collaborative Stage
Mets at Dx

- Brain and cerebral meninges use a specific set of codes
- Pituitary gland, craniopharyngeal duct, pineal gland, and Other Parts of the CNS use the same site-specific codes
**CS Site-Specific Factor 1**  
**WHO Grade Classification**  
**Brain/Other CNS ONLY**

- Code the WHO Grade Classification as documented in the medical record.

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>010</td>
<td>Grade I</td>
</tr>
<tr>
<td>020</td>
<td>Grade II</td>
</tr>
<tr>
<td>030</td>
<td>Grade III</td>
</tr>
<tr>
<td>040</td>
<td>Grade IV</td>
</tr>
<tr>
<td>999</td>
<td>Clinical dx/unknown. Not documented in medical record, etc.</td>
</tr>
</tbody>
</table>
CS Site-Specific Factor 1

- Do not code WHO grade in the 6th digit histology data field
- Applies only to C70.0-C70.9, C71.0-C71.9, C72.0-C72.9
- There is no CS Site-Specific Factor 1 data field for pituitary gland, craniopharyngeal duct, or pineal gland
Questions

• Questions should be directed to your Quality Control Coordinator

• Regional QC Coordinators will forward to the appropriate staff member at CCR
Acknowledgments

• NAACCR Registry Operations Benign Brain Tumor Subcommittee
• CDC National Program for Cancer Registries
• NCI/SEER Program
• Collaborative Stage Task Force