# Multiple Primary and Histology Coding Rules
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- i. Head and Neck – Multiple primary
  - Head and Neck – Histology
- ii. Colon – Multiple primary
  - Colon – Histology
- iii. Lung – Multiple primary
  - Lung – Histology
- iv. Melanoma – MP
  - Melanoma – Histology
- v. Breast – MP
  - Breast – Histology
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  - Kidney - Histology
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Benign and Borderline Intracranial and CNS Tumors
Equivalent Terms, Definitions, Charts and Illustrations
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753

Note: Malignant intracranial and CNS tumors have a separate set of rules.

Do not change the behavior code when during the lifetime of the patient when a tumor(s) progresses from a benign /0 to an uncertain whether benign or malignant /1 behavior.

These rules apply to tumors that occur within the cranial vault or within the spinal canal (reportable)

Note: Non-malignant peripheral nerve tumors are not reportable

Equivalent or Equal Terms (Terms that can be used interchangeably)
- Tumor, mass, lesion, neoplasm
- Type, subtype, variant

Definitions

**Benign:** ICD-O-3 behavior code of /0.

**Borderline:** ICD-O-3 behavior code of /1.

**Cerebellum:** The part of the brain below the back of the cerebrum. It regulates balance, posture, movement, and muscle coordination.

**Corpus Callosum:** A large bundle of nerve fibers that connect the left and right cerebral hemispheres. In the lateral section, it looks a bit like a "C" on its side.

**Different lateralities:** The right side of a site and the left side of a site are different lateralities.

**Frontal Lobe of the Cerebrum:** The top, front region of each of the cerebral hemispheres. Used for reasoning, emotions, judgment, and voluntary movement.

**Infratentorial:** Tumors located in the posterior fossa, cerebellum, or fourth ventricle.

**Invasive:** ICD-O-3 behavior code of /3.

**Medulla Oblongata:** The lowest section of the brainstem (at the top end of the spinal cord). It controls automatic functions including heartbeat, breathing, etc.
Benign and Borderline Intracranial and CNS Tumors
Equivalent Terms, Definitions, Charts and Illustrations
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753

**Meninges:** The three membranes that cover the brain and spinal cord. The outside layer is the dura mater and is the most resilient. The center layer is the arachnoid membrane. The thin innermost layer is the pia mater.

**Mesencephalon:** The region of the brainstem located above the pons.

**Nerve sheath:** A protective covering around nerves.

**Occipital Lobe of the Cerebrum:** The region at the back of each cerebral hemisphere that contains the centers of vision and reading ability (located at the back of the head).

**Parietal Lobe of the Cerebrum:** The middle lobe of each cerebral hemisphere between the frontal and occipital lobes. It contains important sensory centers (located at the upper rear of the head).

**Pituitary Gland:** A gland attached to the base of the brain that secretes hormones. It is located between the Pons and the Corpus Callosum, above the Medulla Oblongata. Synonym: Hypophysis.

**Pons:** The region of the brainstem located below the mesencephalon and above the medulla oblongata.

**Progression of disease:** For the purposes of these rules, progression is defined as a change to a more aggressive behavior (Example: a change from /0 to /1).

**Spinal Cord:** A thick bundle of nerve fibers that runs from the base of the brain to the hip area, running through the spine (vertebrae).

**Supratentorial:** Tumors located in the sellar or suprasellar region or in other areas of the cerebrum.

**Temporal Lobe of the Cerebrum:** The region at the lower side of each cerebral hemisphere; contains centers of hearing and memory (located at the sides of the head).

**Timing:** The amount of time between the original and subsequent tumors is not used to determine multiple primaries because the natural biology of non-malignant tumors is that of expansive, localized growth.

**Transformation:** The histology of a disease process may change over time.
Table 1 – Paired Sites

*Table Instructions:* Use this table to identify paired sites (Rule M5).

<table>
<thead>
<tr>
<th>Column 1: Paired Sites</th>
<th>Column 2: Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cerebral meninges, NOS</td>
<td>C700</td>
</tr>
<tr>
<td>Cerebrum</td>
<td>C710</td>
</tr>
<tr>
<td>Frontal lobe</td>
<td>C711</td>
</tr>
<tr>
<td>Temporal lobe</td>
<td>C712</td>
</tr>
<tr>
<td>Parietal lobe</td>
<td>C713</td>
</tr>
<tr>
<td>Occipital lobe</td>
<td>C714</td>
</tr>
<tr>
<td>Olfactory nerve</td>
<td>C722</td>
</tr>
<tr>
<td>Optic nerve</td>
<td>C723</td>
</tr>
<tr>
<td>Acoustic nerve</td>
<td>C724</td>
</tr>
<tr>
<td>Cranial nerve</td>
<td>C725</td>
</tr>
</tbody>
</table>
Chart 1: Benign and Borderline Intracranial and CNS Tumors

Note: This chart is based on the WHO Classification of Tumors of the Benign Brain. Use this chart to determine multiple primaries and to code histology as instructed in the coding rules.

- **Glial Tumors**
  - **Ependymomas**
    - Subependymoma (9383/1)
    - Myxopapillary Ependymoma (9394/1)
    - Choroid glioma (9444/1)
  - **Neuronal and neuronal-glial neoplasms**
    - Subependymal giant cell astrocytoma (9384/1)
    - Desmoplastic infantile astrocytoma (9412/1)
    - Dysembryoplastic neuroepithelial tumor (9413/0)
    - Glioblastoma (9442/1)
    - Ganglioglioma (9505/1)
    - Central neurocytoma (9506/1)

- **Nerve Sheath Tumors**
  - **Neurofibromas**
    - Neurofibroma (9540/0), Neurofibromatosis, NOS (9540/1)
    - Melanotic neurofibroma (9541/0)
    - Plexiform neurofibroma (9550/0)
  - **Neurinomatosis (9556/1)**
  - **Neurothekeoma (9570)**
  - **Neuroma (9571/0)**
  - **Perineurioma, NOS (9571/0)**

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Benign and Borderline Intracranial and CNS Tumors
Equivalent Terms, Definitions, Charts and Illustrations
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753

www.gender.org.uk/about/07neur/74_brain.htm
Benign and Borderline Intracranial and CNS Tumors
Equivalent Terms, Definitions, Charts and Illustrations
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Meninges

URL: www.cardioliving.com/consumer/Stroke/Hemorrhagic_Stroke.sht 7/18/03

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Benign and Borderline Intracranial and CNS Tumors Multiple Primary Rules - Flowchart
(C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753)

Note: Malignant intracranial and CNS tumors have a separate set of rules.
* Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.
** Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.

<table>
<thead>
<tr>
<th>UNKNOWN IF SINGLE OR MULTIPLE TUMORS</th>
<th>DECISION</th>
<th>NOTES</th>
</tr>
</thead>
<tbody>
<tr>
<td>M1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Is it impossible to determine if there is a single tumor or multiple tumors?</td>
<td>YES</td>
<td></td>
</tr>
<tr>
<td>NO</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Error: Choose appropriate module</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>SINGLE TUMOR</th>
<th>DECISION</th>
<th>NOTES</th>
</tr>
</thead>
<tbody>
<tr>
<td>M2</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Is there a single tumor?</td>
<td>YES</td>
<td></td>
</tr>
<tr>
<td>NO</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Error: Choose appropriate module</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Tumor(s) not described as metastasis.
Use this rule only after all information sources have been exhausted.
End of instructions for Unknown if Single or Multiple Tumors.
Tumor not described as metastasis.
The tumor may overlap onto or extend into adjacent/contiguous site or subsite.
End of instructions for Single Tumor.

Revised April 30, 2008
Benign and Borderline Intracranial and CNS Tumors Multiple Primary Rules - Flowchart
(C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753)

Note: Malignant intracranial and CNS tumors have a separate set of rules.

* Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.
** Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.

**MULTIPLE TUMORS**
Multiple tumors may be a single primary or multiple primaries.

<table>
<thead>
<tr>
<th>DECISION</th>
<th>NOTES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tumors not described as metastases.</td>
<td></td>
</tr>
</tbody>
</table>

**M3**
Is there an invasive tumor (/3) and either a benign brain tumor (/0) or an uncertain/borderline brain tumor (/1)?

**M4**
Are there tumors in sites with ICD-O-3

topography codes that are different
at the second (Cxx), third character (Cxxx) and/or
fourth character (Cxxxx)?

**M5**
Are there tumors on both sides (left and right) of a paired site (See Table 1)?
Benign and Borderline Intracranial and CNS Tumors Multiple Primary Rules - Flowchart

(C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753)

Note: Malignant intracranial and CNS tumors have a separate set of rules.
* Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.
** Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.

**MULTIPLE TUMORS, continued**

**M6**
Is there an atypical choroid plexus papilloma (9390/1) following a choroid plexus papilloma, NOS (9390/0)?

**YES**

SINGLE Primary*

**NOTES**
Tumors not described as metastases.

**NO**

**M7**
Is there a neurofibromatosis, NOS (9540/1) following a neurofibroma, NOS (9540/0)?

**YES**

SINGLE Primary*

**NOTES**
Do not code progression of disease as multiple primaries.

**NO**

**M8**
Do the tumors have two or more histologic types on the same branch in Chart 1?

**YES**

SINGLE Primary*

**NOTES**
Do not code progression of disease as multiple primaries.

**NO**

Next Page
Benign and Borderline Intracranial and CNS Tumors Multiple Primary Rules - Flowchart
(C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753)

Note: Malignant intracranial and CNS tumors have a separate set of rules.

* Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.
** Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.

M9
Do the tumors have multiple histologic types on different branches in Chart 1?

M10
Do the tumors have two or more histologic types and at least one of the histologies is not listed in Chart 1?

M11
Do the tumors have ICD-O-3 histology codes that are different at the first (xxx), second (xx), or third (x) number?

NOTES
Tumors not described as metastases.

Flowchart Key
- Question
- Yes
- No
- Program
- Notes
- Flow/Direction

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Benign and Borderline Intracranial and CNS Tumors Multiple Primary Rules - Flowchart
(C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753)

Note: Malignant intracranial and CNS tumors have a separate set of rules.

* Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.

** Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.

** Rule M12 Examples: The following are examples of cases that use Rule M12. This is NOT intended to be an exhaustive set of examples; there are other cases that may be classified as a single primary.

---

**Warning:** Using only these case examples to determine the number of primaries can result in major errors.

<table>
<thead>
<tr>
<th>Example 1</th>
<th>Example 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tumors in the same site with the same histology (Chart 1) and the same laterality as the original tumor are a single primary.</td>
<td>Tumors in the same site with the same histology (Chart 1) and it is unknown if laterality is the same as the original tumor are a single primary.</td>
</tr>
</tbody>
</table>

---

**End of instructions for Multiple Tumors.**

Timings is not used to determine multiple primaries for benign and borderline intracranial and CNS tumors.
Benign and Borderline Intracranial and CNS Tumors Histology Coding Rules - Flowchart
(C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753)

Note: Malignant intracranial and CNS tumors have a separate set of rules.

### SINGLE TUMOR

<table>
<thead>
<tr>
<th>Rule</th>
<th>Action</th>
<th>Notes and Examples</th>
</tr>
</thead>
</table>
| H1   | Code the histology documented by the physician. | 1. Priority for using documents to code the histology  
  - Documentation in the medical record that refers to pathologic or cytologic findings  
  - Physician’s reference to type of tumor (histology) in the medical record  
  - PET, CT or MRI scans  
  2. Code the specific histology when documented.  
  3. Code the histology to 8000 (neoplasm, NOS) as stated by the physician when nothing more specific is documented. |
|     | YES    |        |
|     | NO     |        |
| H2   | Code the histology. |  |
|     | YES    |        |
|     | NO     |        |

**Flowchart Key**
- **Question**
- **End Program**
- **Notes**
- **Flow Direction**

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Benign and Borderline Intracranial and CNS Tumors Histology Coding Rules - Flowchart
(C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753)

Note: Malignant intracranial and CNS tumors have a separate set of rules.

**SINGLE TUMOR**

<table>
<thead>
<tr>
<th>Rule</th>
<th>Action</th>
<th>Notes and Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>H3</td>
<td>Are there multiple histologies and all histologies are in the same branch on Chart 1?</td>
<td>Code the more specific histology.</td>
</tr>
<tr>
<td></td>
<td>YES</td>
<td></td>
</tr>
<tr>
<td></td>
<td>NO</td>
<td></td>
</tr>
<tr>
<td>H4</td>
<td>Code the numerically higher ICD-O-3 code.</td>
<td></td>
</tr>
</tbody>
</table>

This is the end of instructions for Single Tumor.
Code the histology according to the rule that fits the case.
Benign and Borderline Intracranial and CNS Tumors Histology Coding Rules - Flowchart
(C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753)

Note: Malignant intracranial and CNS tumors have a separate set of rules.

MULTIPLE TUMORS ABSTRACTED AS A SINGLE PRIMARY

<table>
<thead>
<tr>
<th>Rule</th>
<th>Action</th>
<th>Notes and Examples</th>
</tr>
</thead>
</table>
| H5 | **Is there no pathology/cytology specimen or is the pathology/cytology report unavailable?** | 1. Priority for using documents to code the histology
   o Documentation in the medical record that refers to pathologic or cytologic findings
   o Physician's reference to type of tumor (histology) in the medical record
   o PET, CT or MRI scans
   2. Code the specific histology when documented.
   3. Code the histology to 8000 (neoplasm, NOS) or as stated by the physician when nothing more specific is documented. |
| H6 | **Are there multiple meningiomas of uncertain behavior?** | 1. This is a rare condition that is usually associated with neurofibromatosis type 2 and other genetic disorders.
   2. Use this code only for meningiomas with uncertain behavior; do not use this code for multiple benign or malignant meningiomas. |

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Benign and Borderline Intracranial and CNS Tumors Histology Coding Rules - Flowchart
(C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753)

Note: Malignant intracranial and CNS tumors have a separate set of rules.

MULTIPLE TUMORS ABSTRACTED AS A SINGLE PRIMARY

<table>
<thead>
<tr>
<th>Rule</th>
<th>Action</th>
<th>Notes and Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>H7</td>
<td>Code the histology.</td>
<td></td>
</tr>
<tr>
<td>H8</td>
<td>Code the histology from the original diagnosis.</td>
<td>Do not change the behavior code when a later tumor(s) shows progression of disease.</td>
</tr>
<tr>
<td>H9</td>
<td>Code the more specific histology.</td>
<td></td>
</tr>
<tr>
<td>H10</td>
<td>Code the numerically higher ICD-O-3 histology code.</td>
<td></td>
</tr>
</tbody>
</table>

This is the end of instructions for Multiple Tumors Abstracted as a Single Primary.

Code the histology according to the rule that fits the case.
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Benign and Borderline Intracranial and CNS Tumors
Multiple Primary Rules – Matrix
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753

* Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.
** Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.

<table>
<thead>
<tr>
<th>Rule</th>
<th>Site</th>
<th>Histology</th>
<th>Laterality</th>
<th>Behavior</th>
<th>Notes/Examples</th>
<th>Primary</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>UNKNOWN IF SINGLE OR MULTIPLE TUMOR</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Tumor(s) not described as metastasis</td>
<td></td>
</tr>
<tr>
<td>M1</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Use this rule only after all information sources have been exhausted</td>
<td>Single*</td>
</tr>
<tr>
<td><strong>SINGLE TUMOR</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Tumor not described as metastasis</td>
<td></td>
</tr>
<tr>
<td>M2</td>
<td>Single</td>
<td></td>
<td></td>
<td></td>
<td>The tumor may overlap onto or extend into adjacent/contiguous site or subsite</td>
<td>Single*</td>
</tr>
<tr>
<td><strong>MULTIPLE TUMORS</strong> Multiple tumors may be a single primary or multiple primaries</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Tumors not described as metastases</td>
<td></td>
</tr>
<tr>
<td>M3</td>
<td>Brain</td>
<td></td>
<td></td>
<td>Invasive (/3) and either a benign (/0) or uncertain/borderline (/1)</td>
<td></td>
<td>Multiple**</td>
</tr>
<tr>
<td>M4</td>
<td>Topography codes different at the second (Cxxx) and/or third (Cxxx) character, ), or fourth (Cxxx) are multiple primaries.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Multiple**</td>
</tr>
<tr>
<td>M5</td>
<td></td>
<td></td>
<td></td>
<td>Both sides (left and right) of a paired site (Table 1)</td>
<td></td>
<td>Multiple**</td>
</tr>
<tr>
<td>M6</td>
<td>Atypical choroid plexus papilloma (9390/1) following Choroid plexus papilloma, NOS (9390/0)</td>
<td></td>
<td></td>
<td>Do not code progression of disease as multiple primaries</td>
<td>Single*</td>
<td></td>
</tr>
</tbody>
</table>

Note: Malignant intracranial and CNS tumors have a separate set of rules.

Revised April 30, 2008
### Benign and Borderline Intracranial and CNS Tumors

#### Multiple Primary Rules – Matrix

**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**

<table>
<thead>
<tr>
<th>Rule</th>
<th>Site</th>
<th>Histology</th>
<th>Laterality</th>
<th>Behavior</th>
<th>Notes/Examples</th>
<th>Primary</th>
</tr>
</thead>
<tbody>
<tr>
<td>M7</td>
<td></td>
<td>Neurofibromatosis, NOS (9540/1) Following Neurofibroma, NOS (9540/0)</td>
<td></td>
<td></td>
<td>Do not code progression of disease as multiple primaries</td>
<td>Single*</td>
</tr>
<tr>
<td>M8</td>
<td></td>
<td>Multiple types on the same branch in Chart 1</td>
<td></td>
<td></td>
<td></td>
<td>Single*</td>
</tr>
<tr>
<td>M9</td>
<td></td>
<td>Multiple types on different branches in Chart 1</td>
<td></td>
<td></td>
<td></td>
<td>Multiple**</td>
</tr>
<tr>
<td>M10</td>
<td></td>
<td>Multiple types, at least one not listed in Chart 1</td>
<td></td>
<td></td>
<td></td>
<td>Multiple**</td>
</tr>
<tr>
<td>M11</td>
<td></td>
<td>Codes are different at the first (xxxx), second (xxxxx) or third (xxxxx) number</td>
<td></td>
<td></td>
<td>Use this rule when none of the histology codes are listed in Chart 1</td>
<td>Multiple**</td>
</tr>
</tbody>
</table>

Revised April 30, 2008
## Multiple Primary Rules – Matrix

**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**

<table>
<thead>
<tr>
<th>Rule</th>
<th>Site</th>
<th>Histology</th>
<th>Laterality</th>
<th>Behavior</th>
<th>Notes/Examples</th>
<th>Primary</th>
</tr>
</thead>
<tbody>
<tr>
<td>M12</td>
<td>Does not meet any of the above criteria</td>
<td></td>
<td></td>
<td></td>
<td>Timing is not used to determine multiple primaries for benign and borderline intracranial and CNS tumors. <strong>Examples:</strong> The following are examples of cases that use Rule M12. This is NOT intended to be an exhaustive set of examples; there are other cases that may be classified as a single primary. <strong>Warning:</strong> <em>Using only these case examples to determine the number of primaries can result in major errors.</em></td>
<td>Single*</td>
</tr>
</tbody>
</table>

**Example 1:** Tumors in the same site with the same histology (Chart 1) and the same laterality as the original tumor are a single primary.

**Example 2:** Tumors in the same site with the same histology (Chart 1) and it is unknown if laterality is the same as the original tumor are a single primary.

**Example 3:** Tumors in the same site and same laterality with histology codes not listed in Chart 1 that have the same first three numbers are a single primary.
Benign and Borderline Intracranial and CNS Tumors
Histology Coding Rules – Matrix
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753

*Note:* Malignant intracranial and CNS tumors have a separate set of rules.

<table>
<thead>
<tr>
<th>Rule</th>
<th>Pathology/Cytology Specimen</th>
<th>Histology</th>
<th>Behavior</th>
<th>Notes and Examples</th>
<th>Code</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>SINGLE TUMOR</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
| H1 | No specimen or report available | | | 1: Priority for using documents to code the histology  
● Documentation in the medical record that refers to pathologic or cytologic findings  
● Physician’s reference to type of tumor (histology) in the medical record  
● PET, CT or MRI scans  
2: Code the specific histology when documented  
3: Code the histology to 8000 (neoplasm, NOS) as stated by the physician when nothing more specific is documented | | Histology documented by the physician |
| H2 | | One type | | | The histology |
| H3 | | Multiple, all in the same branch on Chart 1 | | | The more specific histology |
| H4 | | None of the above conditions are met | | | The histology with the numerically higher ICD-O-3 code |

| **MULTIPLE TUMORS ABSTRACTED AS A SINGLE PRIMARY** | | | | | |
| H5 | No specimen or report available | | | 1: Priority for using documents to code the histology  
● Documentation in the medical record that refers to pathologic or cytologic findings  
● Physician’s reference to type of tumor (histology) in the medical record  
● PET, CT or MRI scans  
2: Code the specific histology when documented  
3: Code the histology to 8000 (neoplasm, NOS) as stated by the physician when nothing more specific is documented | | Histology documented by the physician |

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<table>
<thead>
<tr>
<th>Rule</th>
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<th>Histology</th>
<th>Behavior</th>
<th>Notes and Examples</th>
<th>Code</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>H6</strong></td>
<td>Multiple meningiomas</td>
<td>Uncertain behavior (/1)</td>
<td>1: This is a rare condition that is usually associated with neurofibromatosis type 2 and other genetic disorders 2: Use this code only for meningiomas with uncertain behavior; do not use this code for multiple benign or malignant meningiomas</td>
<td>9530/1</td>
<td></td>
</tr>
<tr>
<td><strong>H7</strong></td>
<td>One type</td>
<td></td>
<td></td>
<td>The histology</td>
<td></td>
</tr>
<tr>
<td><strong>H8</strong></td>
<td>Original diagnosis</td>
<td></td>
<td>Do not change the histology code when a later tumor(s) shows progression of disease</td>
<td>The histology from the original diagnosis.</td>
<td></td>
</tr>
<tr>
<td><strong>H9</strong></td>
<td>Multiple, all in the same branch on Chart 1</td>
<td></td>
<td></td>
<td>The more specific histology</td>
<td></td>
</tr>
<tr>
<td><strong>H10</strong></td>
<td>None of the above conditions are met</td>
<td></td>
<td></td>
<td>The histology with the numerically higher ICD-O-3 code</td>
<td></td>
</tr>
</tbody>
</table>

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Benign and Borderline Intracranial and CNS Tumors
Multiple Primary Rules – Text
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753

Note: Malignant intracranial and CNS tumors have a separate set of rules.

### UNKNOWN IF SINGLE OR MULTIPLE TUMORS

**Note:** Tumor(s) not described as metastasis

**Rule M1**
When it is not possible to determine if there is a **single** tumor **or multiple** tumors, opt for a single tumor and abstract as a single primary.*

* Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.

This is the end of instructions for Unknown if Single or Multiple Tumors.

### SINGLE TUMOR

**Note:** Tumor not described as metastasis

**Rule M2**
A **single tumor** is always a single primary. *

* Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.

This is the end of instructions for Single Tumor.

### MULTIPLE TUMORS

Multiple tumors may be a single primary or multiple primaries.

**Note:** Tumors not described as metastases

**Rule M3**
An **invasive** brain tumor (/3) **and either** a **benign** brain tumor (/0) **or an uncertain/borderline** brain tumor (/1) are always multiple primaries. **

**Rule M4**
Tumors with ICD-O-3 **topography** codes that are **different** at the second (Cxx) and/or third characters (Cxx), or fourth (Cxxx) are multiple primaries. **

**Rule M5**
Tumors on **both sides** (left and right) of a **paired site** (Table 1) are multiple primaries. **

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Rule M6  An atypical choroid plexus papilloma (9390/1) following a choroid plexus papilloma, NOS (9390/0) is a single primary. *
  Note: Do not code progression of disease as multiple primaries.

Rule M7  A neurofibromatosis, NOS (9540/1) following a neurofibroma, NOS (9540/0) is a single primary. *
  Note: Do not code progression of disease as multiple primaries.

Rule M8  Tumors with two or more histologic types on the same branch in Chart 1 are a single primary. *

Rule M9  Tumors with multiple histologic types on different branches in Chart 1 are multiple primaries. **

Rule M10 Tumors with two or more histologic types and at least one of the histologies is not listed in Chart 1 are multiple primaries. **

Rule M11 Tumors with ICD-O-3 histology codes that are different at the first (xxxx), second (xxxx) or third (xxxx) number are multiple primaries. **
  Note: Use this rule when none of the histology codes are listed in Chart 1.

Rule M12 Tumors that do not meet any of the above criteria are a single primary. *
  Note: Timing is not used to determine multiple primaries for benign and borderline intracranial and CNS tumors.

Rule M12 Examples: The following are examples of cases that use Rule M12. This is NOT intended to be an exhaustive set of examples; there are other cases that may be classified as a single primary. Warning: Using only these case examples to determine the number of primaries can result in major errors.

| Example 1: Tumors in the same site with the same histology (Chart 1) and the same laterality as the original tumor are a single primary. |
| Example 2: Tumors in the same site with the same histology (Chart 1) and it is unknown if laterality is the same as the original tumor are a single primary. |
| Example 3: Tumors in the same site and same laterality with histology codes not listed in Chart 1 that have the same first three numbers are a single primary. |

** Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.
This is the end of instructions for Multiple Tumors.

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Benign and Borderline Intracranial and CNS Tumors
Histology Coding Rules – Text
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Note: Malignant intracranial and CNS tumors have a separate set of rules.

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**SINGLE TUMOR**

**Rule H1** Code the histology documented by the physician when there is **no pathology/cytology specimen** or the **pathology/cytology report** is **not available**.

*Note 1:* Priority for using documents to code the histology
- Documentation in the medical record that refers to pathologic or cytologic findings
- Physician’s reference to type of tumor (histology) in the medical record
- PET, CT or MRI scans

*Note 2:* Code the specific histology when documented.
*Note 3:* Code the histology to 8000 (neoplasm, NOS) or as stated by the physician when nothing more specific is documented.

**Rule H2** Code the histology when only **one histologic type** is identified.

**Rule H3** When there are **multiple histologies** and all histologies are in the **same branch** on Chart 1, code the more specific histology.

**Rule H4** Code the histology with the **numerically higher** ICD-O-3 code.

This is the end of instructions for Single Tumor.
Code the histology according to the rule that fits the case.

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**MULTIPLE TUMORS ABSTRACTED AS A SINGLE PRIMARY**

**Rule H5** Code the histology documented by the physician when there is **no pathology/cytology specimen** or the **pathology/cytology report** is **not available**.

*Note 1:* Priority for using documents to code the histology
- Documentation in the medical record that refers to pathologic or cytologic findings
- Physician’s reference to type of tumor (histology) in the medical record
- PET, CT or MRI scans

*Note 2:* Code the specific histology when documented.
*Note 3:* Code the histology to 8000 (neoplasm, NOS) or as stated by the physician when nothing more specific is documented.
Rule H6  Code multiple meningiomas of uncertain behavior to 9530/1  
*Note 1:* This is a rare condition that is usually associated with neurofibromatosis type 2 and other genetic disorders  
*Note 2:* Use this code only for meningiomas with uncertain behavior; do not use this code for multiple benign or malignant meningiomas

Rule H7  Code the histology when only **one histologic type** is identified.

Rule H8  Code the histology from the original diagnosis.  
*Note:* Do not change the behavior code when a later tumor(s) shows progression of disease.

Rule H9  When there are **multiple histologies** and all histologies are in the **same branch** on Chart 1, code the more specific histology

Rule H10  Code the histology with the **numerically higher** ICD-O-3 code.

This is the end of instructions for Multiple Tumors Abstracted as a Single Primary.  
Code the histology according to the rule that fits the case.