

Multiple Primary and Histology Coding Rules

Table of Contents

I.	Cover Page	
II.	Table of Contents	2
III.	Preface	5
IV.	Multiple Primary and Histology Rules General Instructions	7
V.	Terms & Definitions - Multiple Primary and Histology Coding Rules	15
	i.	Head and Neck 17
	ii.	Colon 29
	iii.	Lung 33
	iv.	Melanoma 41
	v.	Breast 47
	vi.	Kidney 55
	vii.	Ureter/Renal Pelvis/Bladder 61
	viii.	Benign Brain 68.1
	ix.	Malignant Brain 69
	x.	Other Sites 77
VI.	Flowchart Format - Multiple Primary and Histology Coding Rules	83
	i.	Head and Neck – Multiple primary 85
		Head and Neck – Histology 89
	ii.	Colon – Multiple primary 95
		Colon – Histology 100
	iii.	Lung – Multiple primary 111
		Lung – Histology 116
	iv.	Melanoma – MP 123
		Melanoma - Histology 126
	v.	Breast – MP 131
		Breast - Histology 135
	vi.	Kidney – MP 147
		Kidney - Histology 152
	vii.	Ureter/Renal Pelvis/Bladder – MP 159
		Ureter/Renal Pelvis/Bladder – Histo 163
	viii.	Benign Brain – MP 168.1
		Benign Brain – Histo 168.6
	ix.	Malignant Brain – MP 169
		Malignant Brain – Histo 173
	x.	Other Sites – Multiple primary 177
		Other Sites – Histology 184

Multiple Primary and Histology Coding Rules

Table of Contents

VII. Table Matrix Format - Multiple Primary and Histology Coding Rules	201
i. Head and Neck – Multiple primary	203
Head and Neck – Histology	207
ii. Colon – Multiple primary	211
Colon – Histology	213
iii. Lung – Multiple primary	221
Lung – Histology	224
iv. Melanoma – MP	229
Melanoma – Histology	231
v. Breast – MP	233
Breast – Histology	235
vi. Kidney – MP	243
Kidney - Histology	246
vii. Ureter/Renal Pelvis/Bladder – MP	249
Ureter/Renal Pelvis/Bladder – Histo	251
viii. Benign Brain – MP	254.1
Benign Brain – Histo	254.5
ix. Malignant Brain – MP	255
Malignant Brain – Histo	257
x. Other Sites – Multiple primary	259
Other Sites – Histology	263
VIII. Text Format - Multiple Primary and Histology Coding Rules	275
i. Head and Neck – Multiple primary	277
Head and Neck – Histology	279
ii. Colon – Multiple primary	283
Colon – Histology	285
iii. Lung – Multiple primary	289
Lung – Histology	291
iv. Melanoma – MP	295
Melanoma – Histology	297
v. Breast – MP	299
Breast – Histology	301
vi. Kidney – MP	307
Kidney - Histology	309
vii. Ureter/Renal Pelvis/Bladder – MP	313
Ureter/Renal Pelvis/Bladder – Histo	315

Multiple Primary and Histology Coding Rules

Table of Contents

viii.	Benign Brain – MP	318.1
	Benign Brain – Histo	318.3
ix.	Malignant Brain – MP	319
	Malignant Brain – Histo	321
x.	Other Sites – Multiple primary	323
	Other Sites – Histology	326
IX.	New Data Items	333
Appendix A	Histology Coding Committee and Project Roster	345
Appendix B	Commission on Cancer Site Teams	349

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.

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

Table 1 –Paired Sites

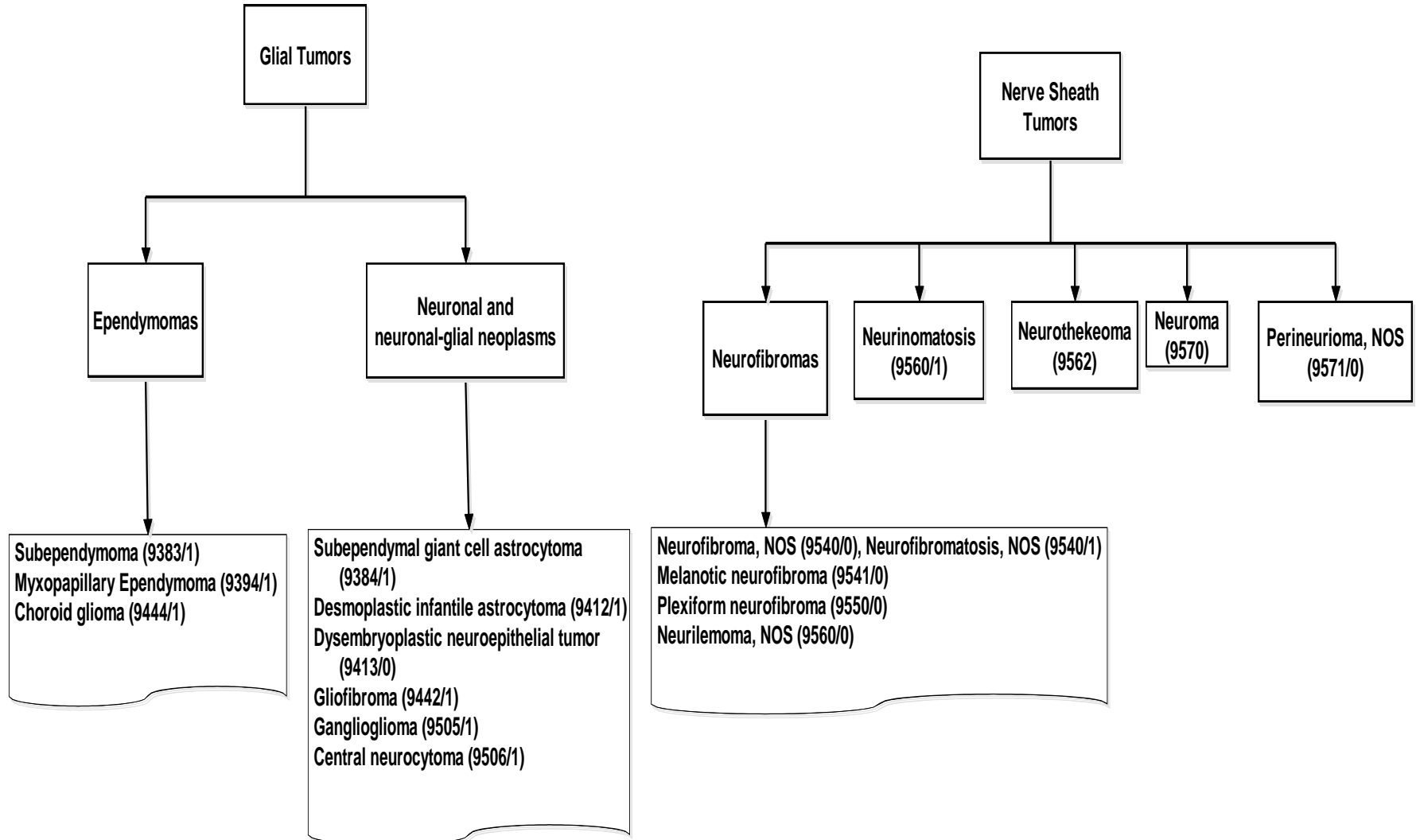
Table Instructions: Use this table to Identify paired sites (Rule M5).

Column 1: Paired Sites	Column 2: Code
Cerebral meninges, NOS	C700
Cerebrum	C710
Frontal lobe	C711
Temporal lobe	C712
Parietal lobe	C713
Occipital lobe	C714
Olfactory nerve	C722
Optic nerve	C723
Acoustic nerve	C724
Cranial nerve	C725

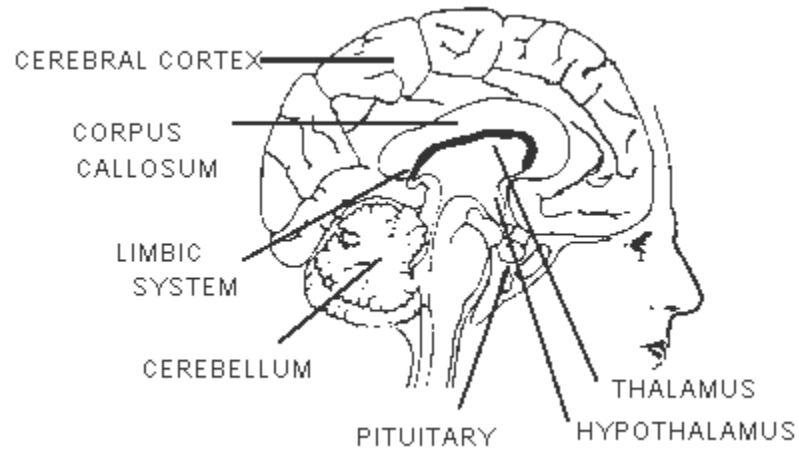
Benign and Borderline Intracranial and CNS Tumors
Equivalent Terms, Definitions, Charts and Illustrations
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753

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.



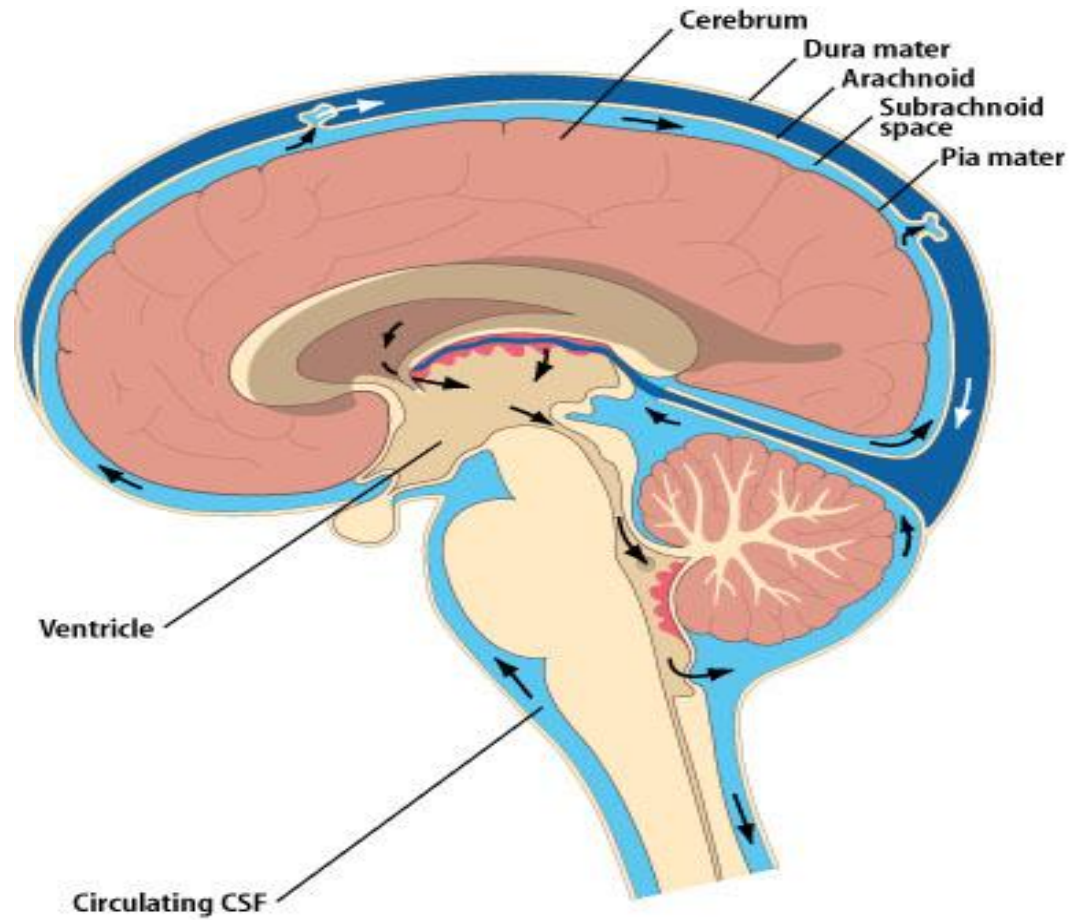
**Benign and Borderline Intracranial and CNS Tumors
Equivalent Terms, Definitions, Charts and Illustrations
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**



www.gender.org.uk/about/07neur/74_brain.htm

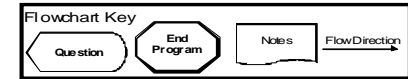
**Benign and Borderline Intracranial and CNS Tumors
Equivalent Terms, Definitions, Charts and Illustrations
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Meninges



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

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



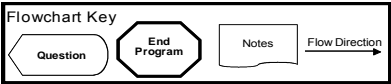
Note: Malignant intracranial and CNS tumors have a separate set of rules.

* 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.

UNKNOWN IF SINGLE OR MULTIPLE TUMORS	DECISION	NOTES
<p>M1</p> <pre> graph TD Q1{{Is it impossible to determine if there is a single tumor or multiple tumors?}} D1{{SINGLE Primary*}} E1[Error: Choose appropriate module] Q1 -- YES --> D1 Q1 -- NO --> E1 </pre>	<p>End of instructions for Unknown if Single or Multiple Tumors</p>	<p>Tumor(s) not described as metastasis.</p> <p>Use this rule only after all information sources have been exhausted.</p>
SINGLE TUMOR	DECISION	NOTES
<p>M2</p> <pre> graph TD Q2{{Is there a single tumor?}} D2{{SINGLE Primary*}} E2[Error: Choose appropriate module] Q2 -- YES --> D2 Q2 -- NO --> E2 </pre>	<p>End of instructions for Single Tumor.</p>	<p>Tumor not described as metastasis.</p> <p>The tumor may overlap onto or extend into adjacent/contiguous site or subsite.</p>

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



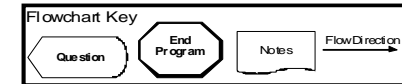
Note: Malignant intracranial and CNS tumors have a separate set of rules.

- * 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	DECISION	NOTES
<p>Multiple tumors may be a single primary or multiple primaries.</p>		<p>Tumors not described as metastases.</p>
<p>M3</p> <p>Is there an invasive tumor (/3) and either a benign brain tumor (/0) or an uncertain/borderline brain tumor (/1)?</p>	<p>YES → MULTIPLE Primaries**</p>	
<p>NO ↓</p> <p>M4</p> <p>Are there tumors in sites with ICD-O-3 topography codes that are different at the second (Cxxx), third character (Cxxx) and/or fourth character (Cxxx)?</p>	<p>YES → MULTIPLE Primaries**</p>	
<p>NO ↓</p> <p>M5</p> <p>Are there tumors on both sides (left and right) of a paired site (See Table 1) ?</p>	<p>YES → MULTIPLE Primaries**</p>	
<p>NO ↓</p> <p>Next Page</p>		

Benign and Borderline Intracranial and CNS Tumors Multiple Primary Rules - Flowchart

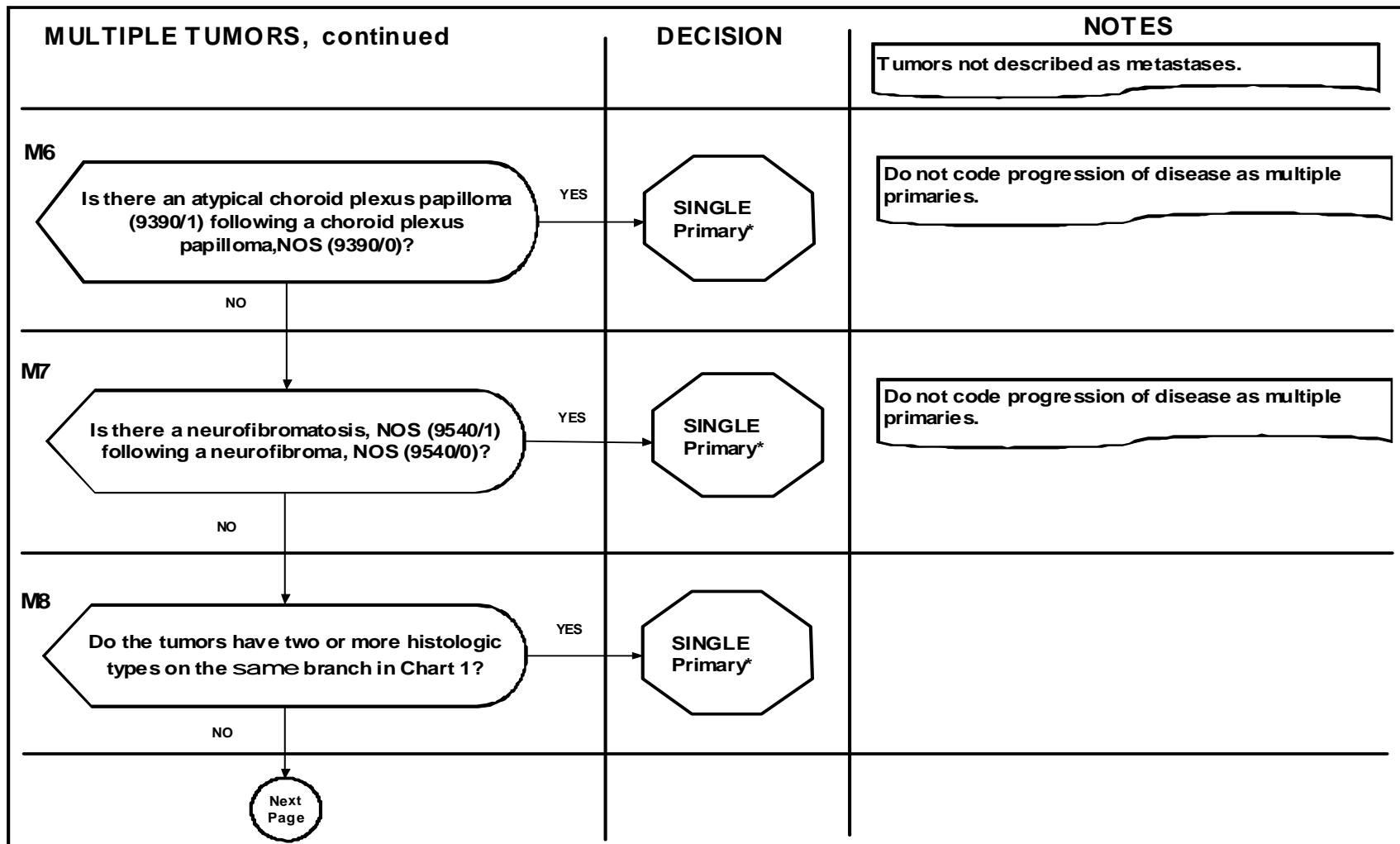
(C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753)



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

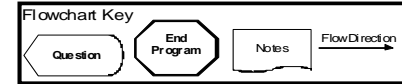
* Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.

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



Benign and Borderline Intracranial and CNS Tumors Multiple Primary Rules - Flowchart

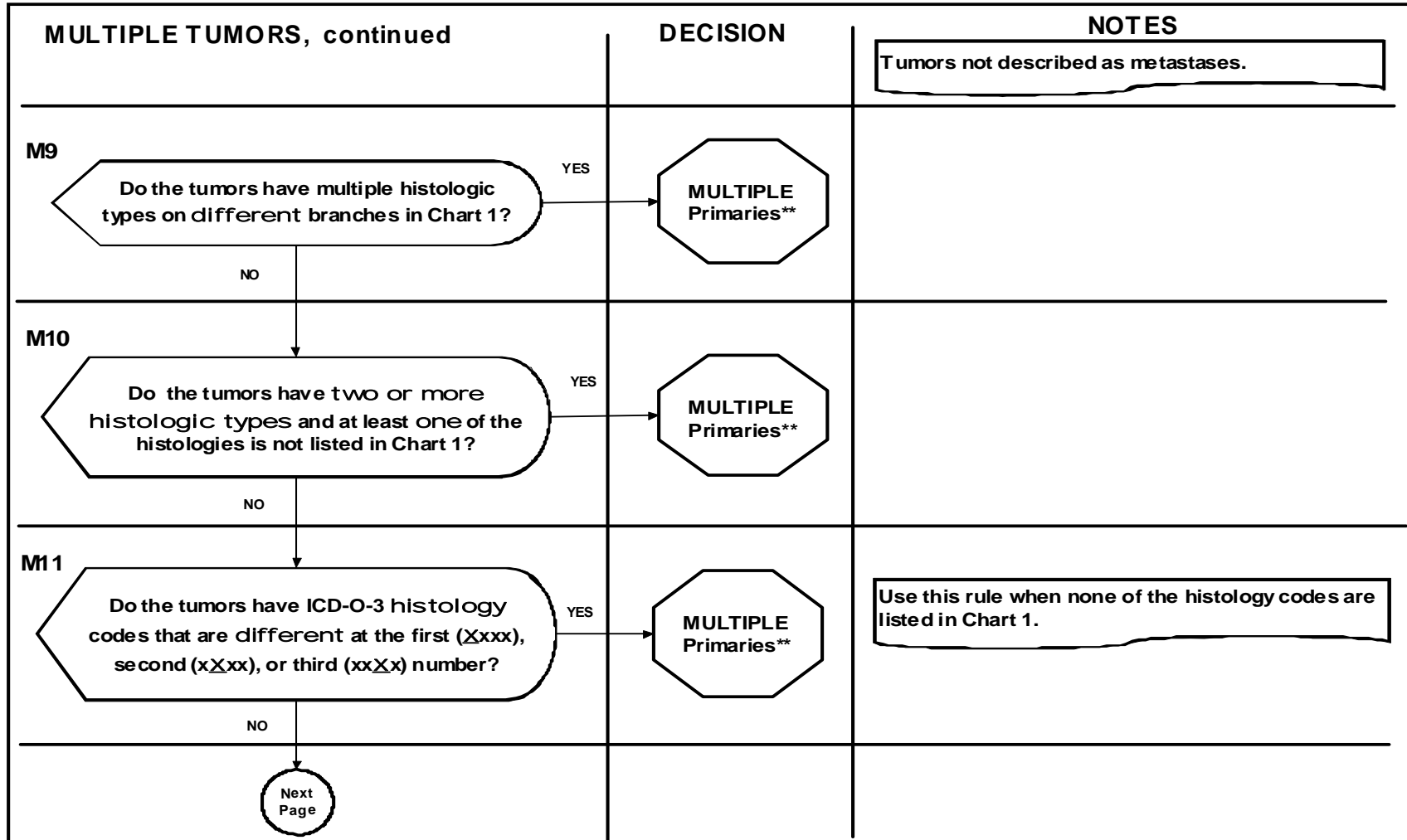
(C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753)



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

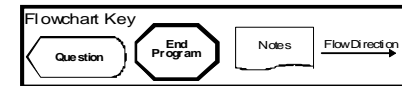
* Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.

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



Benign and Borderline Intracranial and CNS Tumors Multiple Primary Rules - Flowchart

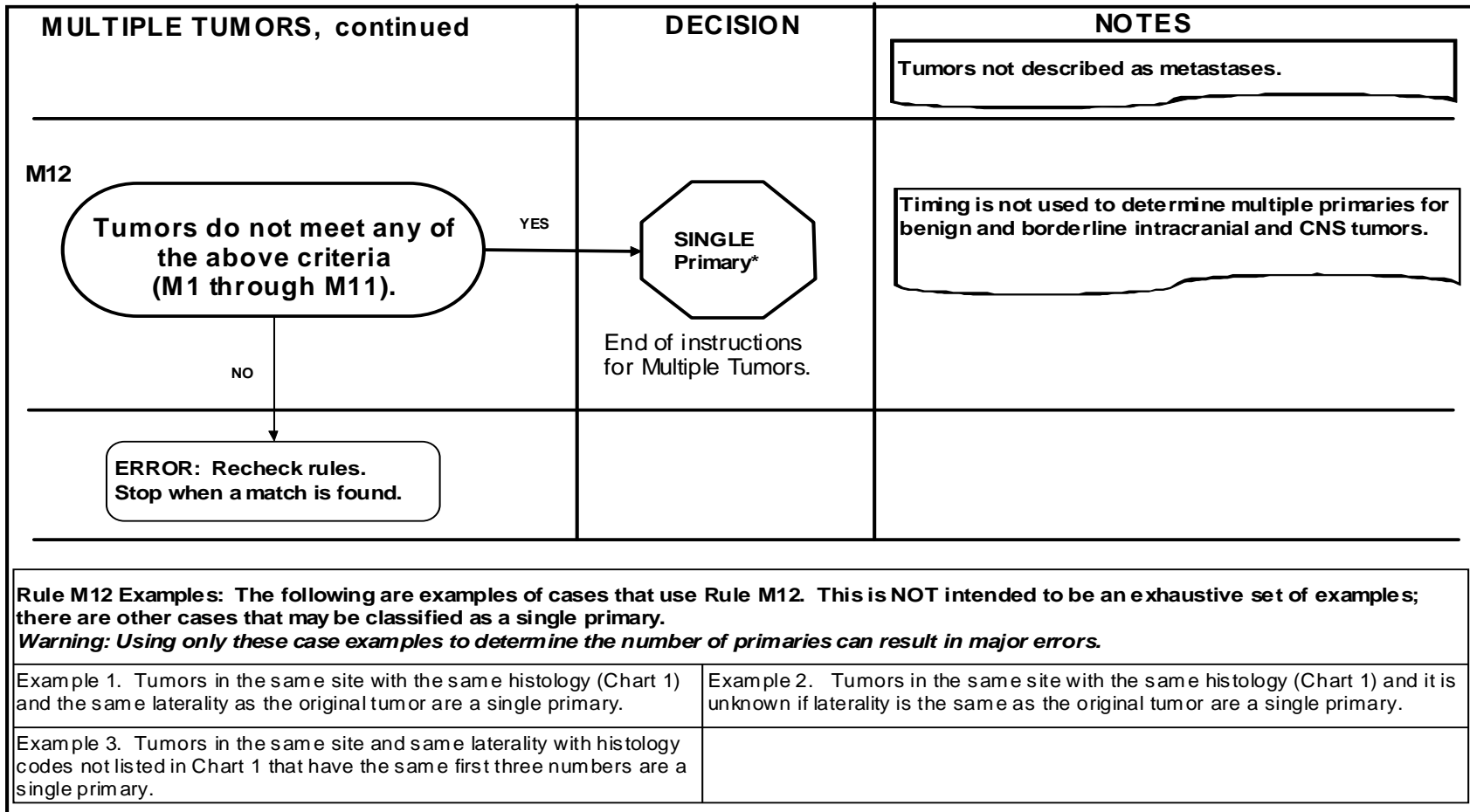
(C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753)



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

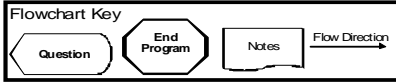
* Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.

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



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.

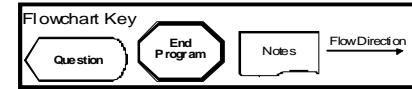


SINGLE TUMOR

Rule	Action	Notes and Examples
<p>H1</p> <p>YES</p> <p>NO</p>		<ol style="list-style-type: none"> 1. Priority for using documents to code the histology <ul style="list-style-type: none"> 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) as stated by the physician when nothing more specific is documented.
<p>H2</p> <p>YES</p> <p>NO</p> <p>Next Page</p>		

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.



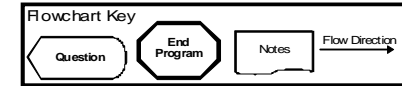
SINGLE TUMOR

Rule	Action	Notes and Examples
<p>H3</p> <p>YES</p>		
<p>NO</p> <p>H4</p>		

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.



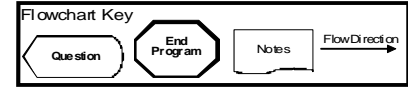
MULTIPLE TUMORS ABSTRACTED AS A SINGLE PRIMARY

Rule	Action	Notes and Examples
<p>H5</p>		<ol style="list-style-type: none"> 1. Priority for using documents to code the histology <ul style="list-style-type: none"> 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.
<p>H6</p>		<ol style="list-style-type: none"> 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.

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

Rule	Action	Notes and Examples
H7 <p>Is only one histologic type identified?</p>	<p>Code the histology.</p>	
H8 <p>Was there a previous tumor(s)?</p>	<p>Code the histology from the original diagnosis.</p>	<p>Do not change the behavior code when a later tumor(s) shows progression of disease.</p>
H9 <p>Are there multiple histologies and all histologies are in the same branch on Chart 1?</p>	<p>Code the more specific histology.</p>	
H10	<p>Code the numerically higher ICD-O-3 histology code.</p>	

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

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

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

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

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

Rule	Site	Histology	Laterality	Behavior	Notes/Examples	Primary
M12	Does not meet any of the above criteria				<p>Timing is not used to determine multiple primaries for benign and borderline intracranial and CNS tumors.</p> <p>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.</p> <p>Warning: <i>Using only these case examples to determine the number of primaries can result in major errors.</i></p> <p>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</p> <p>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.</p> <p>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.</p>	Single*

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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.

Rule	Pathology/Cytology Specimen	Histology	Behavior	Notes and Examples	Code
SINGLE TUMOR					
H1	No specimen or report available			<i>1:</i> Priority for using documents to code the histology <ul style="list-style-type: none"> • 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 <i>2:</i> Code the specific histology when documented <i>3:</i> 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			<i>1:</i> Priority for using documents to code the histology <ul style="list-style-type: none"> • 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 <i>2:</i> Code the specific histology when documented <i>3:</i> Code the histology to 8000 (neoplasm, NOS) as stated by the physician when nothing more specific is documented	Histology documented by the physician

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

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

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.*

Note: Use this rule only after all information sources have been exhausted.

*** 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.*

Note: *The tumor may overlap onto or extend into adjacent/contiguous site or subsite.*

*** 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 (Cxxx) and/or third characters (Cxxx), or fourth (Cxxx) are multiple primaries. **

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

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

- 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.**

Benign and Borderline Intracranial and CNS Tumors
Histology Coding 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.

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.

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.

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

- 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.
