Multiple Primary and Histology Site Specific Coding Rules
Benign and Borderline Intracranial and CNS Tumors
Prerequisites

Completion of Multiple Primary and Histology General Coding Rules
There are many ways to view the Multiple Primary/Histology rules, or rather ways in which they are diagramed to aid in understanding how they are put together.

The rules themselves are provided in three formats to support different styles of learning and interaction with instructions:

- text
- matrix
- flowchart

Any abstraction from the rules does not replace the rules, but may provide insight into their underlying structure.
Borrowing from the three formats for the rules themselves, structure can be diagramed in a *text or outline form*, a *matrix or table form*, and a *flowchart form*.

You have previously reviewed the table format when you looked at the two color coded spreadsheets for the multiple primary and the histology rules.

The table form shows most clearly the alternating patterns of single versus multiple primary decisions across the primary sites, the commonality of rules across the primary sites, and the clustering of site-specific rules in different primary sites.
Links to illustrations and/or diagrams will be provided for each site to diagram the process of multiple-primary decision making in a sequential fashion, comparing existing and new records in a registry database. The charts included here assume the tumors have already been assigned to the appropriate anatomic site.
Multiple Primary and Histology Coding Rules

BENIGN AND BORDERLINE INTRACRANIAL AND CNS TUMORS
Multiple primary and histology coding rules for benign and borderline tumors of the central nervous system are presented in separate modules from those for malignant tumors.

The rules and definitions section notes that the rules apply to tumors within the cranial vault and the spinal canal, they do not apply to tumors of peripheral nerves.

Tumors with behavior codes of “/0”, benign, and “/1”, borderline, are included, but the code of “/0” is not upgraded to “/1” if the behavior of the tumor progresses over time.
As with malignant tumors of the central nervous system, there is no timing rule for these tumors other than the identification of certain histologies which are specifically treated as recurrences.

Unlike malignant tumors, laterality is considered in determining multiple primaries for benign/borderline tumors; a table of paired sites is included for reference.

Relationship trees are presented for two groups of benign/borderline histologies, glial tumors and nerve sheath tumors; as with other such trees, the more specific histologies are located on the lower branches of the trees.
Reviewing the Multiple Primary rules, the first module is Unknown if Single or Multiple Tumors with the standard

- M1 rule, when it is not possible to determine if there is a single or multiple tumors, consider the case as a single tumor and abstract as a single primary.

The Single Tumor module contains the standard

- M2 rule, a single tumor is always a single primary.

Going to the rules for assessing Multiple Tumors, the first rule,

- M3, repeats the rule seen in the Malignant CNS Tumor modules, an invasive CNS tumor and either a benign or borderline CNS tumor are always multiple primaries; if a borderline tumor were to progress to a malignant tumor, the malignant tumor would be a new primary.
The next rule,

- M4, is the rule for ICD-O-3 histology codes, but it includes differences at the fourth character, so that tumors involving different lobes of the brain or different cranial nerves with specific ICD-O-3 codes would be considered separate primaries.

The next rule,

- M5, separates primary tumors by laterality for benign CNS conditions; as with melanomas, midline is considered a separate laterality from right and left.
The remaining rules relate to histology.

- Rule M6 states explicitly that an atypical choroid plexus papilloma following a choroid plexus papilloma is a single primary.
- Rule M7 similarly states that neurofibromatosis following neurofibroma is a single primary.

Rules M8, M9, and M10 refer to the chart or tree of histology groups:

- M8, tumors with two or more histologic types on the same branch are a single primary.
- M9, tumors with multiple histologic types on different branches are multiple primaries.
- M10, tumors with two or more histologic types, at least one of which is not listed in one of the trees, are multiple primaries.
Rule M11 is the standard ICD-O-3 histology rule, histologies with codes differing among the first three characters are multiple primaries.

M12, is the default rule, tumors that have not met any of the preceding criteria are single primaries.

Examples in this latter group include multiple tumors in the same site with same laterality and histology as an earlier primary, and multiple tumors in the same site with same histology but unknown if laterality is the same as an earlier primary.
There are fewer histology coding rules for the Benign/Borderline CNS Tumors than for other sites.

Most of the rules are the same in both the Single Tumor and Tumors Abstracted as a Single Primary module.

- H1 and H5 are the standard rule about using the histology documented by the physician when there is no pathology/cytology specimen or the report is unavailable.

After this rule, the Multiple Tumors module includes a site-specific rule, that multiple meningiomas with borderline behavior are coded to “95301.”
The next rule in both modules, • H2 and H7, is the standard, code the histology when only one histologic type is identified.

Another rule is added in the Multiple Tumors module, to code the histology from the original diagnosis when a later tumor shows progression in behavior.

• Rules H3 and H8 indicate that the more specific histology is coded when the diagnosed histologies are contained on the charts.
• Rules H4 and H10 are the standard, code the histology with the higher ICD-O-3 code.
Case

- The patient has a history of a meningioma involving the meninges over the right frontal lobe, resected in 2003.
- In 2008 the patient presents with recurrent meningiomas on MRI, involving the right frontal lobe and the left olfactory groove.
In determining number of primary lesions, Rule M1 does not apply, the record indicates the patient has had three separate lesions.

Rule M2 does not apply, there is more than a single tumor.

Rule M3 does not apply, there is no indication of a malignant histology.
Rule M4 does not apply, the topography codes are the same for all lesions, C700.

Rule M5 does apply, there are tumors on both left and right sides, this is a case of multiple primary tumors.

At this point you can distinguish between the left and right-sided meningiomas, but we still have two meningiomas on the right side, and you need to go back through the rules again to determine if these two tumors represent a single or multiple primaries.
Again, for the same reasons as before, Rules M1 through M4 do not apply.

We know Rule M5 does not apply, because both these tumors are on the same side.

Rules M6 and M7 do not apply, the histology is a meningioma, and neither choroid plexus papilloma nor neurofibroma are involved.
Rules M8, M9, and M10 do not apply, meningioma is not included on either of the histology trees on the chart for benign CNS tumors.

Rule M11 does not apply, there is only one histology involved.

The final rule M12 states that this is a single primary, and indeed our case does fit the circumstances in Example 1, tumors in the same site with the same histology and the same laterality as the original tumor are a single primary.
The original meningioma diagnosed in 2003 was reportable by agreement to some central registries; the second meningioma on the right will not be reportable, because by the rules it is recurrent.

Therefore, it was already reported to those registries that did collect benign CNS tumors in 2003, or it was diagnosed before a reportable year for registries that did not collect benign CNS tumors in 2003.

Using the Single Tumor module to code the histology of the left-sided tumor diagnosed in 2008, Rule H1 applies, the histology is stated by the physician as meningioma and there is no pathology or cytology specimen taken.

Because there is a single histology term, “meningioma”, there is no need to go back through the rules for assistance in coding a complex diagnostic statement.