Multiple Primary and Histology
Site Specific Coding Rules
MALIGNANT CENTRAL NERVOUS SYSTEM 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.

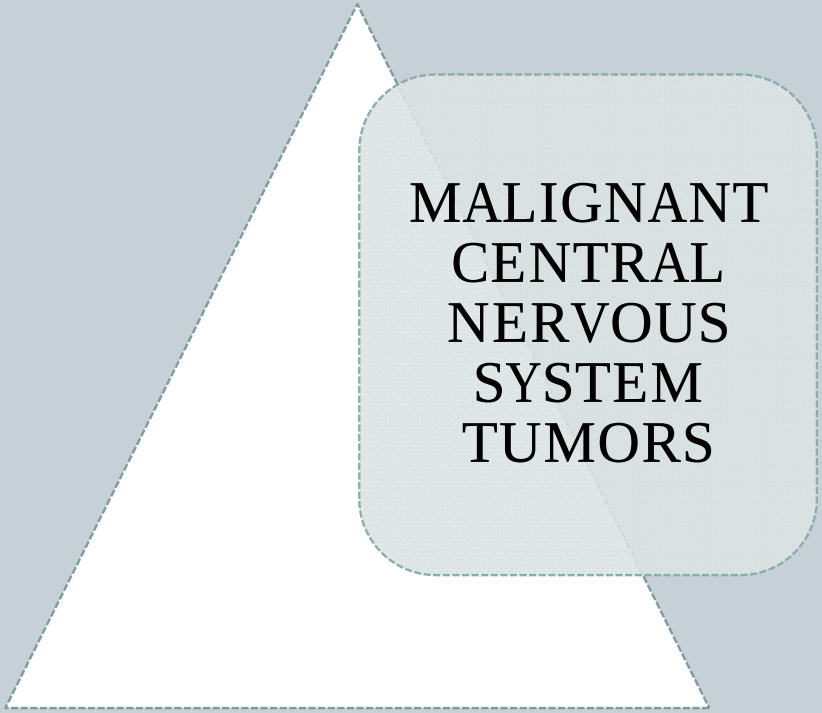
Borrowing from the three formats for the rules themselves, structure can be diagramed in a *text or outline form*, a *matrix or table form*, and a *flowchart form*.

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



Malignant and benign/borderline tumors of the central nervous system have separate rule modules.

However, there are similarities in the presentations for both sets of rules, and in the histology relationships traced through the trees, as described in the introductory unit.

The terms and definitions section for both malignant and benign tumors of the central nervous system adds “variant” to the list of terms that designate a codable subtype of histology.

The two types of cells in the nervous system are described as neurons, or the cells that carry nerve messages, and neuroglia, or the cells that support the nerve cells.

Tumors arising from these two cell types are identified, as well as the lobes of the brain.

A distinction is made between the codes for central primitive neuroectodermal tumors which occur in the central nervous system and peripheral primitive neuroectodermal tumors which occur in soft tissues outside the central nervous system.

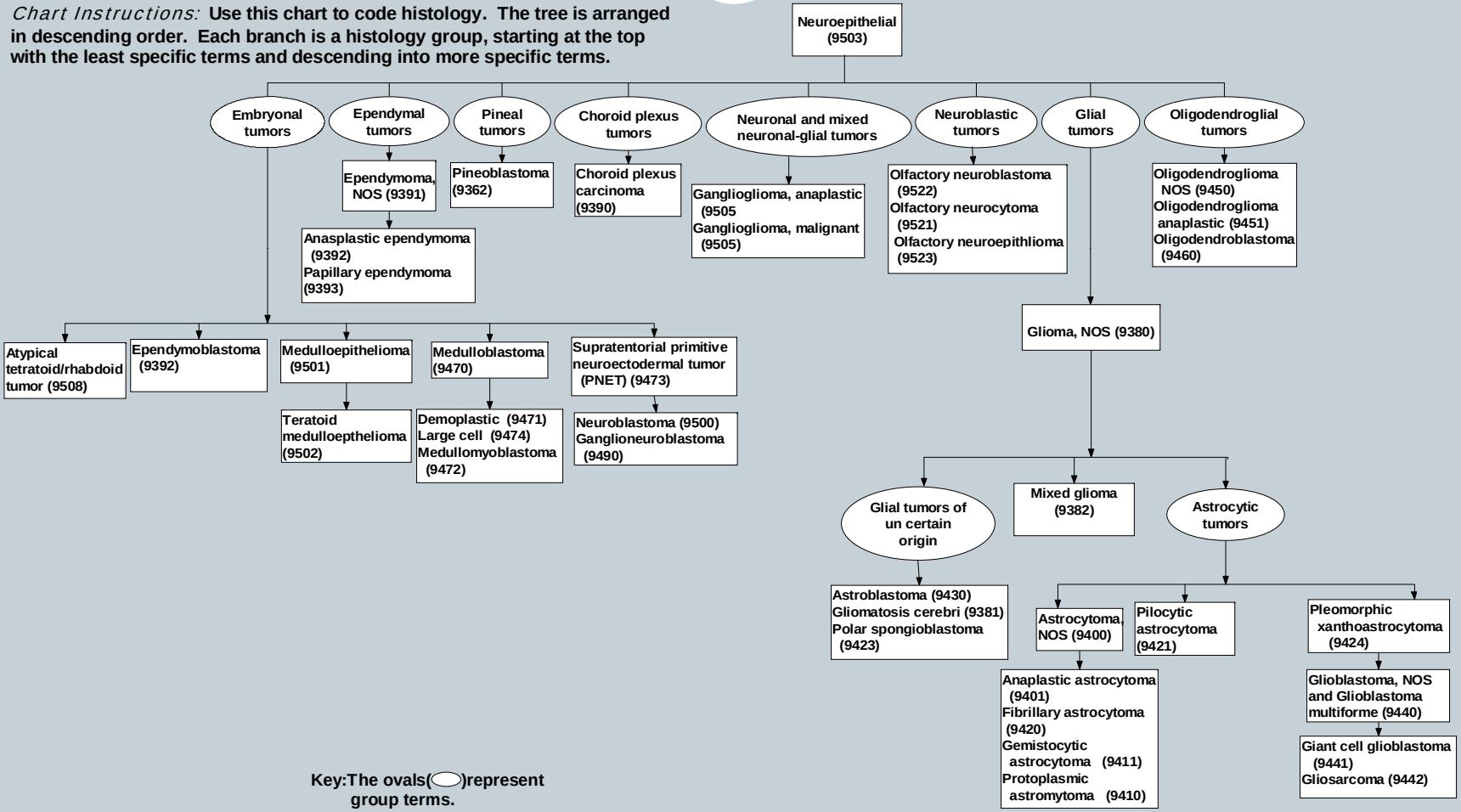
Two charts or family trees of malignant histologies are included: Chart 1 for neuroepithelial malignancies and Chart 2 for non-neuroepithelial malignancies.

Main branches within the neuroepithelial tumors include:

- embryonal
- ependymal
- pineal
- choroid plexus
- neuronal and mixed neuronal-glial
- neuroblastic
- glial
- oligodendroglial tumors

CHART 1

Chart Instructions: **Use this chart to code histology. The tree is arranged in descending order. Each branch is a histology group, starting at the top with the least specific terms and descending into more specific terms.**

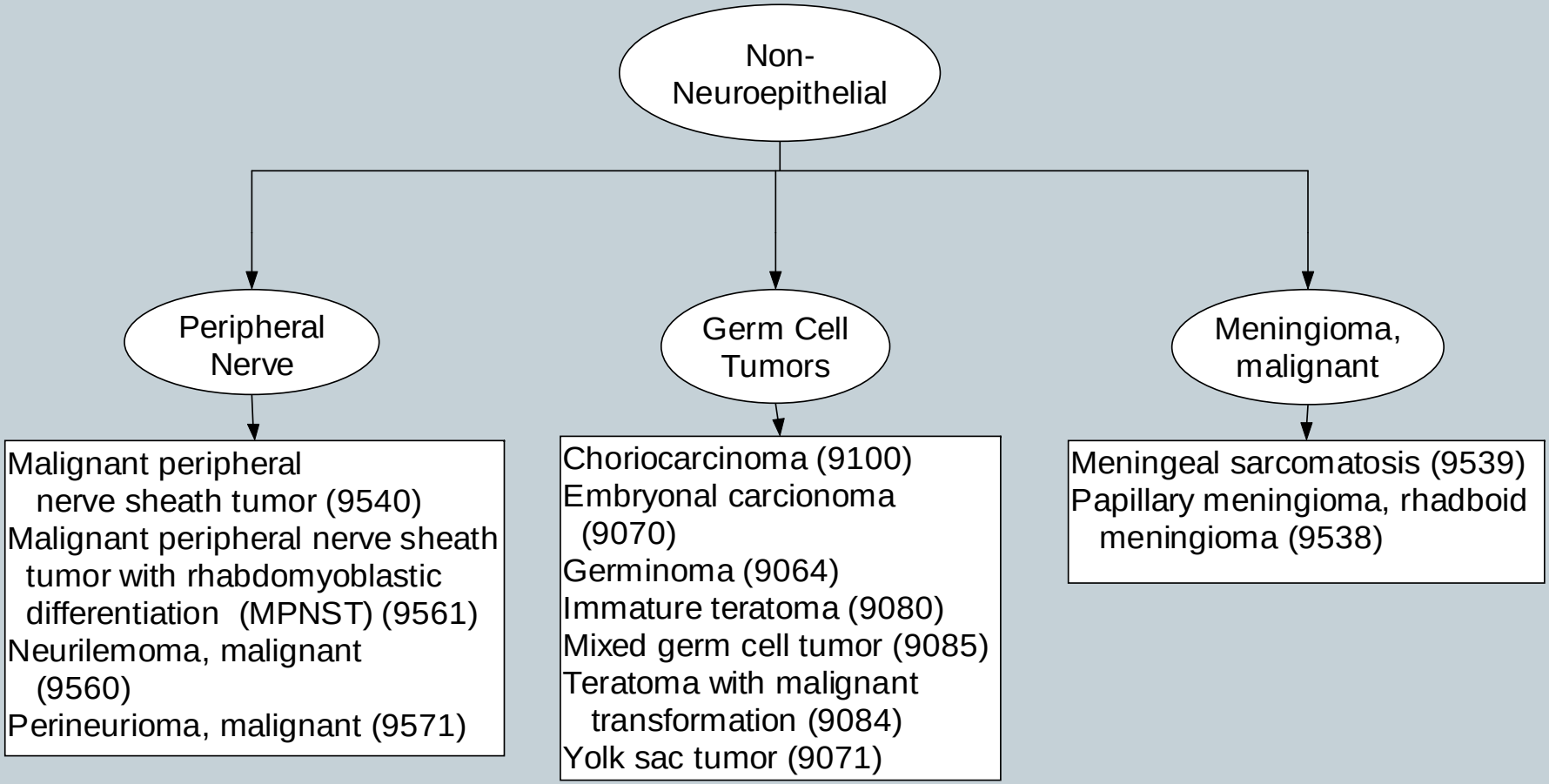


Key:The ovals(⬭)represent group terms.

Main branches within the non- neuroepithelial tumors include:

- peripheral nerve
- germ cell tumors
- malignant meningioma

CHART 2


Non-Neuroepithelial
Peripheral Nerve
Germ Cell Tumors
Meningioma, malignant
Malignant peripheral nerve sheath tumor (9540)
Malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation (MPNST) (9561)
Neurilemoma, malignant (9560)
Perineurioma, malignant (9571)
Choriocarcinoma (9100)
Embryonal carcionoma (9070)
Germinoma (9064)
Immature teratoma (9080)
Mixed germ cell tumor (9085)
Teratoma with malignant transformation (9084)
Yolk sac tumor (9071)
Meningeal sarcomatosis (9539)
Papillary meningioma, rhadboid meningioma (9538)

The first rule within the Multiple Primary Unknown if Single or Multiple Tumors module	• M1, is unique to the Malignant CNS Tumors, and states that an invasive brain tumor and either a benign brain tumor or an uncertain/borderline brain tumor are always multiple primaries.
The second rule,	• M2, is standard, when it is not possible to determine if there is a single or multiple tumors, the case is abstracted as a single primary cancer.
The rule for a single tumor is standard,	• M3, a single tumor is always a single primary.

Within the Multiple Tumors module, the first rule here,

- M4, repeats the statement in M1, so that multiple benign and malignant tumors, whether number of tumors known or not, will always be treated as separate primaries.

The next rule,

- M5, is the standard rule about ICD-O-3 topography codes differing among the first three characters indicating separate primary cancers.

It is worth noting here that the rules for benign CNS tumors consider topography codes differing at the fourth character to be separate primaries; thus malignant tumors coded to C711 and C712 could be the same primary depending on histology, but benign tumors coded to C711 and C712 would be separate primaries.

The next four rules for malignant CNS tumors address histology.

- Rule M6 is site-specific, a glioblastoma or glioblastoma multiforme following a glial tumor is a single primary.
- Rule M7 is site-specific, tumors with ICD-O-3 histology codes located on the same branch in either Chart 1 or Chart 2 are a single primary.
- Rule M8 is also site-specific, tumors with ICD-O-3 histology codes located on different branches in Chart 1 or Chart 2 are multiple primaries.

The next rule is standard,

- M9, tumors with ICD-O-3 histology codes differing among the first three characters are multiple primaries.

And the last rule,

- M10, is the standard default rule, tumors that have not met any criteria of previous rules are abstracted as a single primary cancer.
- Examples given for this rule are tumors of the lobes of the brain.

Laterality and timing rules are not included for malignant CNS tumors.

Timing is specifically negated for certain histologies in both malignant and benign CNS tumors, as seen here with Rule M6 for glioblastoma following a glial tumor.

The Single Tumor and Multiple Tumors Abstracted as a Single Primary histology modules for Malignant CNS Tumors include the same rules in the same pattern, with one additional rule for mixed gliomas for single tumors.

- Rules H1 and H7 are standard, code histology documented by the physician when the cytology/pathology report is not available or no specimen was taken.
- Rules H2 and H8 are standard, code histology from a metastatic site when there is no specimen from the primary site.

The next rule is specific to the Single Tumor histology module,

- H3, code 93823, mixed glioma, when at least two of the following cell types are included in the tumor: astrocytic, oligodendroglial, and ependymal.

The following three rules are the same again for both single and multiple tumor modules:

- H4 and H9, code the histology when only one histology type.
- H5 and H10, code the specific histologic type when the diagnosis includes a non-specific and a specific term on the same branch in Chart 1 or Chart 2.
- H6 and H11, code the histology with the numerically higher ICD-O-3 code.

As with all charts or trees of related histologies, the more specific histologic terms are contained on the lower branches of the tree.

For case review

- The patient was diagnosed and treated for an ependymoma of the cerebellum at age five in 1990.
- The patient presented with symptoms of headache, confusion, and speech disturbances at age 25, and an MRI of the brain revealed a new enhancing 3 x 4 cm mass in the right frontal lobe.
- Diagnosis on stereotactic biopsy was anaplastic astrocytoma.

Referring to the multiple primary rules, the number of tumors is known, so the Unknown if Single or Multiple Tumors module does not apply.

The patient has two tumors, history of prior tumor and the current tumor, so the Single Tumor module does not apply.

Going to the Multiple Tumors module, both tumors are malignant, so Rule M4 does not apply.

The topography codes for both tumors have the same first three characters, “C71”, so Rule M5 does not apply.

This is a case of ependymoma followed by astrocytoma, so Rule M6, glial tumor followed by glioblastoma multiforme, does not apply.

Rule M7 does not apply, ependymoma and astrocytoma are on different branches in Chart 1 for neuroepithelial tumors.

Rule M8 does apply because these histologies are on different branches, so these are multiple primaries.

The first tumor, the ependymoma, was diagnosed before 2007, and would have been abstracted and assigned a histology code according to the rules in use in 1990.

For the second tumor diagnosed when the MP/H rules are in effect, we have a single tumor, an anaplastic astrocytoma.

Referring to the Single Tumor module for histology, Rules H1 and H2 do not apply, there is histology from a primary site specimen.

Rule H3 does not apply, this is not a mixed glioma.

Rule H4 does apply if you consider “anaplastic astrocytoma” as a single histologic term.

If you think of this as a more specific type of astrocytoma, then the next rule, H5, would lead you to the same histologic code, 94013 for anaplastic astrocytoma.