Multiple Primary Histology General Coding Rules
Use a process for determining two critical pieces of information about the cancer disease experienced by each of the patients entered into our cancer registries:

- the number of separate or primary cancers each person has, and
- the type of tissue which gave rise to each of those cancers.
In addition to documenting the impact of the disease on the lives of the patients in our registries, we collect data to support and document the need for research into the causes, diagnosis, treatment, and support of patients with cancer and the results of new technologies, methodologies, and programs applied in disease prevention, diagnosis, and treatment.

The gathering and assessment of cancer information is an integral component in the management of this disease on both a personal and a societal level, and the usefulness of the information, especially at the aggregated level, depends on the correctness or validity of data entered into the cancer registries.
Questions Asked...

Among the host of questions dependent on an accurate counting of cancers within and among individuals and description of histologies:

- Are prevention programs successful?
- Are there correlates between cancer numbers and genetic factors, personal behaviors, socioeconomic conditions, other disease exposures, diagnostic methodologies, environmental conditions?
- Where may research resources be utilized?
- Where and what kind of treatment programs are needed?
These questions are posed at the national as well as regional, state, local, and facility levels, thus creating the need for information or coding rules that are consistent or standardized across all these levels.

Cancer registrars apply standardized rules to describe and record each individual human’s experience of cancer, and the MP/H rules themselves represent a technologic advance in systematizing a process for identifying and coding information to achieve consistency among all registry practitioners.
How do the rules help to achieve this consistency in determining the number of primary cancers and assigning histology codes?

Look first at the structure of the rules.
The principle source of information for a cancer diagnosis is the pathology report, the pathologist’s description of cells or tissue taken from the cancerous lesion and viewed under the microscope.

The MP/H rules start with a few general rules about how to use the rules themselves and where to look on the pathology report for information.

Then the body of the rules follow the traditional presentation of cancer data by primary site and/or histology, with separate units or modules for head and neck, colon, lung, melanoma of skin, breast, kidney, urinary sites, malignant and benign tumors of the brain and central nervous system, and all other sites.
As the rules are still in the process of development, we will see additional modules for hematopoietic primary cancers (leukemias, lymphomas, and plasma cell neoplasms).
The information in each primary site unit includes:

- terms which are considered synonymous for that site
- descriptions of histologic types which may commonly occur in that site or are very specific for the site
- histology and anatomy diagrams
- special information used in applying the rules
After the definitions, diagrams, tables, and charts in each unit, we come to the rules themselves, which actually lead us through a process for determining the correct number of primary cancers and applying the correct histology code.

The rules are organized into separate multiple primary and histology modules, with the multiple primary decision made first, and then the histology decision following.

The multiple primary rules are organized into modules based on the number of tumors identified for the case, and the histology rules are organized into modules based on the number of tumors abstracted as a single primary cancer.
A major innovation in these rules is that they are presented in three separate formats intended to correspond to different learning or working styles, so that each user has a better opportunity to find a match between how he or she processes information and how that information is laid out, and thus to reach a final coding decision most efficiently and correctly.
Another innovation built into these rules, from a cancer perspective, is the codification of the relationship between tumors and cancers.

Multiple tumors occurring at the same time or at different times may be considered one primary cancer or multiple primary cancers.

Both the multiple primary and the histology rules start from the identification of individual tumors and tumor histologies, and end with the identification of cancers and cancer histologies.
A basic rule for the system is to apply the first rule that fits the case circumstances and then stop, but registrars raise many questions comparing the codes reached with the application of later versus earlier rules in each rule module.

The rules were also designed to address the issue of recurrent versus new primary cancers; however probably the second area where registrars raise the most questions is in the identification of possible recurrent versus metastatic tumors.
The Surveillance, Epidemiology, and End Results program (SEER) at the National Cancer Institute (NCI) has taken the lead in developing the MP/H rules, though the list of participants in the project includes the major North American organizations which establish standards for cancer registration. As noted on the SEER MP/H website:

- American College of Surgeons (ACoS)
- Commission on Cancer (CoC)
- American Joint Committee on Cancer (AJCC)
- Centers for Disease Control and Prevention (CDC)
- National Program of Cancer Registries (NPCR)
- National Cancer Registrars Association (NCRA)
- North American Association of Central Cancer Registries (NAACCR)
- 15 central registry representatives
- Statistics Canada

The MP/H Task Force was a diverse group with membership from all but two SEER regions:
Physician guidance by specialty pathologists and clinicians was integral to the review and revision process.

Regular consultation with the editors of ICD-O-3 clarified ICD-O-3 codes and ensured that the new rules accurately reflect the ICD-O-3 intent and purpose.
The SEER website, at www.seer.cancer.gov/tools/mphrules/, is the primary gateway to information on the MP/H rules, including their development, the manual, revisions, clarification and interpretation of coding issues.

SEER conducted a reliability study in fall of 2008, and a major revision of the rules is planned for release in 2010.

The SEER website includes numerous training materials for the rules, including the initial cases which were used for training, transcripts of webinar type training sessions, and units in the online training modules which present cases for abstracting and coding.
Abstracting and coding training opportunities offered by NAACCR, NPCR, and NCRA all include application of the MP/H rules in their presentations.

As noted in the quote from the SEER website, the rules are designed to reflect the intent and purpose of the ICD-O-3 (International Classification of Diseases for Oncology, Third Edition), published by the WHO (World Health Organization).

The ICD-O contains topographic codes for anatomic sites of disease involvement and morphologic or histologic codes for types of tissue in which neoplasms arise.

The ICD-O classification system is used by the named standard setters to enumerate, identify, and collect data on cancer incidence, and the MP/H Rules have been adopted as the process for applying the ICD-O system beginning with cancers diagnosed in 2007.
Also of note to registrars, the WHO “Blue Books” provide the scientific description for the histologies listed in the ICD-O-3.

Titles in the series include:

- pathology and genetics of tumors of the nervous system
- the digestive system
- hematopoietic and lymphoid tissues
- breast and female genital organs
- soft tissues and bone
- skin tumors
- urinary system and male genital organs
- endocrine organs
- head and neck tumors
- tumors of the lung, pleura, thymus, and heart
These general principles apply across all sites.

As do each of the primary site modules, the “General Instructions” starts with a list of “Equivalent or Equal Terms,” and then a list of “Definitions.”
The equivalent terms are considered to be synonymous in their use and interpretation when encountered in cancer diagnostic statements.

The equivalent terms listed in the “General Instructions”, and thus applicable to all primary sites, are:

• “adenocarcinoma” and “glandular carcinoma”
• “multicentric” and “multifocal”
• “tumor”, “mass”, “lesion”, and “neoplasm”
Definitions

The definitions describe in common language some terms used in cancer medical documentation or some histologies specific to primary sites.

“Multiple primaries” are defined as “More than one reportable case.”

“Single primary” is defined as “One reportable case.”
Reportability of a cancer or benign CNS tumor is governed by other rules; the MP/H rules are only applied to cases which have already been determined to be reportable, so the use of “tumor, mass, lesion, neoplasm” for these rules does not translate into meaning that “neoplasm” in a medical chart identifies a reportable disease.
“Contiguous” is defined as a single tumor that bridges multiple sites based on their ICD-O-3 topography codes.

“Focal” is defined as “limited to one specific area,” which may be either macroscopic or microscopic.

- In practice “focal” has caused problems in the application of the rules; interpretation of this term will probably be developed more fully in the 2010 revision, but for now we can note that answers to registrars’ questions indicate that histologic terms modified by “focal” are not considered in coding.
“Recurrence” is given two meanings:

- A second lesion has developed from cancer cells that were not destroyed by the initial therapy,
- A more general sense that cancer has occurred again in the patient.

We will see the inclusion of a general principle and specific timing rules for primary sites that are intended to redefine “recurrence” to a uniform coding standard.
1. Use these rules to determine number of reportable primaries. The rules do not relate to determining reportability, coding stage, or grade. Reportability, stage, and grade have their own rules which are located elsewhere in the compendia of cancer abstracting principles, and which are to be followed as stated elsewhere.
2. The 2007 rules replace are previous multiple primary and histology coding rules. Only these rules are to be used for cases to which they apply, even if they result in coding decisions which conflict with decision that may have been reached under other rules.
3. The rules are effective for cases diagnosed January 1, 2007 and later. Any case with an earlier diagnosis date is to be abstracted using rules in effect at the time of diagnosis. As an extension of this principle, these rules apply to all tumors identified January 1, 2007 and later. For example, a patient may have been diagnosed with a breast cancer in 2005, and returns with a tumor in the same breast in 2009. The MP/H rules would be used to determine if the 2009 tumor is a new primary cancer, rather than the rules in effect in 2005.
4. Read the general instructions and site-specific equivalent terms and definitions before using the rules. The general instructions apply to all cases, the site-specific terms and definitions are just that and may provide information that is not readily recalled by the registrar.
5. Use the format that is easiest to follow. Skipping around among formats is not recommended. The rules are intended to be identical among the three formats.
6. Notes and examples are added for clarity. The notes and examples are not rules. Notes and examples might indicate that you have arrived at the correct rule, but you should not match your cases to the notes and examples to determine the correct coding.
7. Do not use a physician’s statement to decide whether the patient has a recurrence of a previous cancer or a new primary. Use the multiple primary rules as written unless a pathologist compares the present tumor to the original tumor and states that this tumor is a recurrence of cancer from the previous primary. Add to this rule that it applies only to tumors which are not described as metastases occurring in regional or distant sites. Again, registrars have posed many questions about coding new primary cancers when pathologists have not compared histologies from primary and metastatic tumors.
Some additional important points to note in the general rules:

Metastatic tumors are not covered by the rules, except in the case of lung cancer, where certain presentations of multiple tumors indicate:

- Metastatic disease
- Cases of unknown primary
- Cases where the only histology specimens are from metastatic sites
A metastasis from a known primary is not counted as another tumor for that primary cancer, and the diagnosis of metastasis does not require comparison of specimens from the primary and the metastatic sites.
In the absence of a pathology or cytology report from the primary site, the rules allow coding from a physician’s statement about the diagnosis.

In the absence of a specimen being taken from the primary site, but a specimen taken from a metastatic site, the rules allow the coding of the histology or cytology from the metastatic site.
For pathology reports, code from the most representative tumor specimen described on any of the pathology reports for a single tumor, and code from the final diagnosis including comments and addenda (unless instructed to do otherwise).

• For example, in the case of an incisional biopsy followed by resection, the specimen from the resection would be taken from the most representative.

However, if the initial biopsy removed all or the greatest part of the tumor, compared to the resection, then the specimen from the biopsy would be the most representative.
Stop when you have reached a rule that fits your case situation. Sometimes you need to make a second pass through the rules to determine a correct histology code, but this situation is usually self-evident.

For example, you may reach a rule that tells you to code the most invasive histology, and you may need to return through the rules to determine which of two equally invasive histologies to code.

A decision point like this generally indicates a return to the start of the histology module, rather than a progression to a later rule.
Ambiguous terms are defined for the MP/H rules. Additional ambiguous terms which may appear in other lists, reportability for example, do not migrate into the MP/H list. Ambiguous terms are treated as positive statements for histology coding; for example a diagnosis stated as “non-small cell favor adenocarcinoma” is coded the same as a diagnosis stated as “adenocarcinoma.”
Before looking at the process of using the rules, look at the three formats to see how they differ in appearance.

The text rules are presented as statements of conclusion.

The matrix rules are presented as rows and columns in a table.

The flowchart rules are presented as decision points based on yes and no answers to questions.
Look at the process for applying the rules to gain a good understanding of how to approach the determination of single or multiple primaries for any site and the assignment of a histology code.

Refer to the flowchart diagram methodology from that format for the rules to describe the procedure.
First determine the general site and histology of the cancer we are dealing with, site so that we can select the correct unit within the MP/H rules and histology to rule out lymphoma, leukemia, and Kaposi sarcoma (lymphoma and leukemia are excluded from the current rules, and Kaposi sarcoma of any site is included in the other sites group).

Next we determine the number of tumors involved in the case, tumor being described as a mass, lesion, or neoplasm-the group of cancerous cells occurring together as a growth that can be described under the microscope, seen on a radiologic exam, or palpated in accessible organs.
We move first to the multiple primary modules for each primary site. For all sites there are three modules:

- “Unknown if Single or Multiple Tumors”
- “Single Tumor”
- “Multiple Tumors”
Two rules are constant across all the units:

- A single tumor is always a single primary.
- If it is unknown if there is a single or multiple tumors, the case is abstracted as a single primary.
Circumstances in which it is unknown if there is a single or multiple tumors may include a case where only metastatic lung disease has been identified, through a liver biopsy for example, and no information is available about the primary lung cancer, or where a large head and neck cancer is identified in the tongue for example, but the biopsy site is identified as floor of mouth.
Given the single tumor/single primary rule, a single breast tumor for example, with no preceding history of breast cancer, would be a single primary cancer.

“Single tumor” within the structure of the rules means single tumor per lifetime in any primary site.
Where multiple tumors are identified, we move to the “Multiple Tumors” module and step through the rules in order until the first rule is reached that applies to the case.

As mentioned previously, many rules are similar across the primary sites but they do not appear in the same order for each site; also as each rule is reached, it may indicate a single primary cancer or multiple primary cancers.
The rules are not ordered so that all cases with multiple tumors coded as a single primary are grouped together and all cases with multiple tumors coded as multiple primaries are grouped together; the order of rules leading to single and multiple primary cancers is different for each primary site, and the sequence of rules must be stepped through for each site.
General Guidelines

- 1 organ
- 1 tumor
- M module – single tumor
  - 1 primary cancer
- H module – single tumor, single primary
General Guidelines

- 1 organ
- 2 tumors
- M module – multiple tumors
  - 2 primary cancers
- H module – single tumor, single primary
- H module – single tumor, single primary
General Guidelines

- 1 organ
- 2 tumors
- M module – multiple tumors
  - 1 primary cancer
- H module – multiple tumors, single primary
General Guidelines

- 1 organ
- 3 tumors
- M module – multiple tumors
  - 2 primary cancers
- H module – multiple tumors, single primary
- H module – single tumor, single primary
General Guidelines

- 1 organ
- 4 tumors
- M module – multiple tumors
  - 2 primary cancers
- H module – multiple tumors, single primary
- H module – multiple tumors, single primary
General Guidelines

- 2 organs
- 2 tumors
- M module – multiple tumors
  - 2 primary cancers
- H module – single tumor, single primary
- H module – single tumor, single primary
General Guidelines

- 2 organs
- 2 tumors
- M module – multiple tumors
  - 2 primary cancers
- H module – multiple tumors, single primary
- H module – single tumor, single primary
General Guidelines

- 2 organs
- 4 tumors
- M module – multiple tumors
  - 2 primary cancers
- H module – multiple tumors, single primary
- H module – multiple tumors, single primary
Urinary Example

- 2 urinary organs
- 3 tumors, same histo
- M module – multiple tumors
  - 1 primary cancer
- H module – multiple tumors, single primary
We have counted the tumors, and determined the number of primary cancers based on the decision we reached by applying the correct multiple primary rule.
At this point we may have:

- one tumor and one cancer
- two tumors and one cancer
- two tumors and two cancers
- more than two tumors and one cancer
- more than two tumors and two or more cancers
For each cancer that we have, we select a histology coding module for that site based on single or multiple tumors for that cancer.

If we started with three tumors and determined that we had one cancer with one tumor and one cancer with two tumors, we would go to the histology module “Single Tumor Abstracted as a Single Primary” to determine the histology for the first cancer, and “Multiple Tumors Abstracted as a Single Primary” for the second cancer.

We determine the histology for each cancer based on the number of tumors for that particular cancer, not on the total number of tumors with which we started the case.
For most sites the Histology rule modules include “Single Tumor Abstracted as Single Primary” and “Multiple Tumors Abstracted as Single Primary.”

The Cutaneous Melanoma rules have a single histology module, “Single Melanoma or Multiple Melanomas Abstracted as a Single Primary.”

The Breast Histology rules include “Single Tumor: In Situ Carcinoma Only,” “Single Tumor: Invasive and In Situ Carcinoma,” “Single Tumor Invasive Carcinoma Only,” and “Multiple Tumors Abstracted as a Single Primary.”

The Other Sites Histology rule modules also include “Single Tumor: In Situ Only,” “Single Tumor: Invasive and In Situ,” “Single Tumor: Invasive Only,” and “Multiple Tumors Abstracted as a Single Primary.”
As for the Multiple Primary Rules, we step through the histology rules, in the right module, until we find a rule that applies to our case situation, and then stop.

As noted previously, we may have to step through the histology rules more than once to determine the correct code, but once we reach a stop and return point, we generally stop and return rather than proceed to a later rule.
Traditionally correspondence in the first three digits of the ICD-O primary site code has been accepted as defining the same primary site (for the same or related histologies) in the United States; thus separate tumors with site codes of C019 and C049 would be separate primaries, but tumors with site codes of C040 and C041 may be the same primary.

Colon and skin traditionally have been exceptions to this rule, where all four digits are used to define separate primary sites: tumors in sites C187, sigmoid colon, and C184, hepatic flexure, are generally separate primaries.

Bilateral involvement of paired sites, except for some sites and histologies, has indicated separate primaries.
The MP/H rules maintain distinctions to the fourth digit for colon, skin, bone, peripheral nerves, and soft tissues, and add this distinction for certain head and neck and CNS sites and anus codes.

The MP/H rules continue to define bilateral Wilm’s tumors for kidney and retinoblastoma for eye as single primaries, but only code epithelial bilateral tumors of the ovary as a single primary.

Bilateral inflammatory carcinoma of the breast is a single primary, and the number of primary cancers with bilateral renal pelvis and ureteral tumors depends on involvement of other urinary sites.
The number of primary cancers with bilateral involvement of the lungs depend on the number of tumors in each lung and the histologic workup that is carried out.

Midline is considered a separate laterality for cutaneous melanomas and benign/borderline tumors of the central nervous system.

MP/H rules based on site codes and laterality are in many cases but not always the first rules that are applied when multiple tumors are involved—the overarching rule is always to use the first rule that applies and then stop.

- For colon cancers for example, adenocarcinoma in adenomatous polyposis coli (familial polyposis) is a single primary, whether tumors present in multiple segments or only a single segment of the colon, rectosigmoid, and rectum.
An invasive tumor following an in situ tumor more than 60 days after diagnosis of the in situ tumor is considered a second primary cancer.

This rule has been included in SEER rules, though not Commission on Cancer (COC) rules, since 1995, and the purpose is to ensure that all invasive cancers are included in published incidence statistics.

For example, if a patient was diagnosed with a noninvasive papillary urothelial carcinoma of the bladder in March, and then on bladder tumor recheck in June a carcinoma invasive into the lamina propria was found, the June tumor would be abstracted as a second primary.

• (If the June tumor were noninvasive, it would not be considered a new primary.)
The only exception to invasive after in situ rule would be if for some reason initial workup of the cancer diagnosis extended out beyond the 60-day time period. However, the record would have to be carefully inspected to distinguish between cases with an extended workup period and cases showing actual progression of disease.
Traditionally cancers have been abstracted as second primaries if they recurred more than 60 days after the original diagnosis unless a physician stated they were recurrent from the first cancer.
As we have noted, the MP/H rules discuss the diagnosis of “recurrence” as having a twofold meaning in clinical practice:

- A return of disease from the same clonal population of cancer cells not completely eradicated in the first course of treatment.
- A new occurrence of cancer in the same organ.
The MP/H rules establish timing guidelines for the identification of new primary cancers within the same primary site as an earlier cancer, which preempt the clinical diagnosis of “recurrence” unless a pathologist determines by histologic examination that the later lesion is a clonal recurrence of the earlier cancer.
The timing rules vary by primary site:

- 5 years for head and neck and breast cancers
- 1 year for most colon cancers
- 3 years for lung, kidney, and urinary cancers except for bladder
- 60 days for cutaneous melanomas
A right breast ductal carcinoma “recurring” in 2007 after an initial R breast ductal carcinoma diagnosed in 2000 would be abstracted as a new primary.

Some sites have no timing rules, such as malignant and benign CNS tumors and prostate adenocarcinomas.

As with all rules, the applications of timing rules for recurrent versus new primary are specific to their location within the set of rules- they are only used if prior rules have not supplied the answer to the single versus multiple primary question.
In addition to having timing rules based on disease recurrence patterns, many primary site groups have other special rules related to cancers that tend to occur in these sites.
Thus the colon module contains rules for determining number of primaries when cancer is diagnosed in polyps.

The lung module contains rules for determining number of primary cancers when multiple tumors are present at diagnosis, not all of which may be biopsied.

The breast module contains special rules for determining number of primaries when combinations of ductal, lobular, and Paget disease are involved.

The urinary group contains a unique set of rules for determining number of primaries when cancer arises in the same tissue type in multiple involved organs.

A number of sites grouped in the other sites module have histology-specific coding rules.
A new concept within the MP/H rules is the diagnosis of multiple histologies in a single tumor or multiple tumors that may belong to a related line or family of histologies that are abstracted as a single primary cancer.
This concept is expressed in three forms:

1. For some sites a rule states that a tumor with a general histologic diagnosis and a tumor with a more specific related diagnosis are a single primary cancer. The general and more specific histology rule appears in the head and neck, colon, lung, kidney, and other sites multiple primary modules. Different histology groups are listed for different primary sites, but the longest list pairs cancer/malignant neoplasm NOS and a specific histology, carcinoma NOS and a specific carcinoma, adenocarcinoma NOS and a specific adenocarcinoma, squamous cell carcinoma NOS and a specific squamous cell carcinoma, melanoma NOS and a specific melanoma, and sarcoma NOS and a specific sarcoma. 8070/3, squamous cell carcinoma, would be a more specific code than 8010/3, carcinoma, and in turn keratinizing squamous cell carcinoma, 8071/3, would be a more specific code than 8070/3.
2. For some sites a rule states that multiple tumors with histologies on a single branch of a histology tree are single primary cancers, and conversely that multiple tumors with histologies on different branches of the tree are multiple primaries. The histology tree rules appear in the malignant and benign CNS modules. Histology trees are also included for head and neck and lung, but they are used in the histology rather than the multiple primary rules for these sites.
3. Tables of related cell types for intraductal and ductal carcinomas for breast, renal cell carcinomas for kidney, and urothelial carcinomas for urinary sites display histology relationships that may not be recognized through inspection of the ICD-O morphology codes.
Chart 1 shows the neuroepithelial tree.

The most generic histology type, “neuroepithelial,” is located at the top of the tree.
All histologies lower on the tree represent more specific types of neuroepithelial tumors.

These histologies belong to eight families or groups:

- embryonal tumors
- ependymal tumor
- pineal tumors
- choroid plex tumors
- neuronal and mixed neuronal-glial tumors
- neuroblast tumors
- glial tumors
- oligodendroglial tumors
Three of these families are further divided with every more specific histologies or subtypes as we descend on the tree.

Thus a gliosarcoma is a more specific type of glioblastoma, which is a more specific type than pleomorphic xanthoastrocytoma, in the group of astrocytic tumors, which fall within the more general category yet of gliomas.
Chart 2 shows related malignant non-neuroepithelial tumors which can occur in the central nervous system.

All the coded histologies are located at the last branching level of the tree.

Rather than showing non-specific and more specific histologies, this tree actually shows groups of related histologies, categorized as peripheral nerve tumors, germ cell tumors, and malignant meningiomas.

We can surmise that coding rules will state that tumors in different branches of this tree will represent separate primaries, but that a rule based on the numeric value of the codes may be used to if two or more of these histologies are combined in a single primary cancer.
Benign/Borderline Tumors

There are two histology trees for the benign/borderline tumors of the central nervous system.

These trees bear some resemblance to the malignant CNS charts.
Glial tumors are subdivided into ependymomas and neuronal and neuronal-glial neoplasms, with the coded histologies falling under these two headings.

Ependymal and neuronal/neuronal-glial tumors are also family names on the malignant CNS chart.

The use of “Glial tumors” as a heading on the benign/borderline chart rather than “Neuroepithelial” is perhaps confusing when comparing charts.

The nerve sheath tumor tree for the benign/borderline CNS tumors compares to the peripheral nerve group in the malignant CNS non-neuroepithelial tree.
Breast

Tables 1 and 2 for use in the breast coding modules list the histologies and codes for subtypes of intraductal and invasive duct carcinomas.

The note on Table 1 indicates that these histologies may also appear as subtypes of invasive duct carcinomas.

Likewise, the note on Table 2 indicates that these histologies may also occur as in situ carcinomas.

In contrast to a similar table of renal cell types, the notes for these tables state that they are not intended to be complete listing of all possible intraductal and duct cell histologies for breast cancers.
Kidney

Table 1 for kidney displays specific renal cell types. The table note indicates that this is a complete listing of specific renal cell types.
Urinary

Table 1 displays a list of urothelial cell types which may appear in urinary organs.

In contrast to the previous tables reviewed for breast and kidney, the note for this table does not indicate either that this is a complete listing of all urothelial cell types or that there may be other urothelial cell types.
The traditional registry rule for coding tumors with multiple histologies as single primaries has relied on the numeric morphology codes themselves: histologies with the same first three digits have been coded as a single primary cancer.

This rule, or a variation stating that histology codes that differ in the first three digits are multiple primaries, has been retained in many of the multiple primary modules as one of the last possible rules to be considered.

Thus, squamous cell carcinoma, 80703, and adenocarcinoma, 81403, represent distinct histologic entities.
The effectiveness of this rule as a coding principle has been weakened by the insertion of new codes into the ICD-O classification schema where room was found within the numbering system, rather than strict adherence to describing histology relationships through number relationships among the codes.

In other words, for example, if a 10-number sequence is used to describe 10 related histologies, and two additional histologies are later identified that logically fit between numbers 3 and 4 in the first series, those codes are no longer available; they are given new codes that cannot by themselves show how those two histologies are related to the original 10.
Perhaps the most obvious example of using an available space in a sequence of codes that does not explicate a relationship among the coded entities is the addition of 80463 in ICD-O-3 for non-small cell carcinomas; codes 8041 through 8045 all represent types of small cell carcinomas.
The final multiple primary rule in all the modules is that multiple tumors that have not met any of the previous criteria are abstracted as a single primary cancer.

This rule essentially states for each site: all circumstances in which multiple tumors are known to represent multiple primary cancers have already been accounted for; all remaining circumstances in which multiple tumors can occur are either known to represent a single primary cancer, or are probably so rare that they can safely be assumed to represent a single primary cancer.

Multiple melanomas with the same histology on the same limb within the same timeframe would fall under this last rule, as would multiple tumors in both lungs with either the same or unknown histologies and within the same timeframe.
As noted, for most sites there are two histology coding modules:

- for single tumors
- for multiple tumors abstracted as a single primary
The first two rules in both single tumor and multiple tumor modules are the same across all sites:

- Code the histology documented by the physician when there is no pathology or cytology specimen or the pathology/cytology report is not available.
- Code the histology from a metastatic site when there is no pathology or cytology specimen from the primary site.
A modified version of the first of these rules is also used in the single in situ and single in situ and invasive tumor modules for breast and other sites, but not the second rule, as in situ tumors by definition must be diagnosed pathologically and they do not have metastatic disease.

The effect of these rules is to allow the assignment of a specific histologic code to reflect the best clinical knowledge about a cancer when direct information from the primary site is not available to the coder.
An example for using the first rule would be the case of a patient seen at a facility for chemotherapy only, no pathology report is available, but the treating physician documents Stage IIB lobular carcinoma of the breast.

A case calling on the second rule would be metastatic malignant melanoma on lung biopsy with no primary skin lesion found on clinical examination.
The next group of rules that appear in many histology modules, for both single and multiple tumors, mirror multiple primary rules that showed up for many sites.

As a single tumor is always abstracted as a single primary cancer, a single histologic type is always coded when identified; as an example, adenocarcinoma of the gallbladder.
Corresponding to the multiple primary rules that tumors with a nonspecific and a specific histology are abstracted as a single primary cancer, and tumors on the same branches of a histology tree are also abstracted as a single primary cancer, we have rules that the more specific histology is coded, comparing a general to a specific histology or comparing histologies on the same branch of a tree; as examples, intestinal type adenocarcinoma of the stomach, neuroendocrine small cell carcinoma of the lung.
The rules contain a strictly applied list of terms that can be used to identify types of histologies: “type, subtype, predominantly, with features of, major, or with differentiation.”

The identification of subtypes for in situ histologies includes these terms, and in addition “architecture and pattern.”
The latter two terms are not to be used to identify subtypes for invasive histologies; and as interpreted by responses on the COC I and R and the SEER SINQ, terms modified by adjectives such as “focal” are also not to be considered.

When multiple specific histologic types are involved in a single tumor or single primary, the rules usually point to a combination code.
And finally, as the MP rules maintain a traditional coding principle based on the numeric structure of the morphology codes, so also the histology rules preserve the traditional rule of coding the histology with the highest numeric code; as an example, balloon cell melanoma, 8722, coded in preference to nodular melanoma, 8721.

We want to emphasize here that the same approach exists in using the histology coding rules—use the first rule that applies and then stop.

The highest numeric code rule is usually the last rule that occurs in each histology module, and in practice will not often be used; when the rule is used, the higher code is usually distinguished only at the fourth character, as in the example.
The histology rules introduce a new concept to registrars:

- To code the invasive histology when a single tumor has both invasive and in situ components or when both invasive and in situ tumors are abstracted as a single primary,
- To code the most invasive histology when multiple invasive tumors are abstracted as a single primary.
The coding of urothelial versus papillary urothelial histologies presents an exception to this concept for urinary cancers.

But where this rule appears, it thus incorporates a component of the staging system into the selection of histology code, based on the idea that the most invasive tumor represents the greatest disease threat to the patient and will guide clinical decision-making.
A tumor with invasive duct carcinoma and lobular carcinoma in situ will be coded as duct carcinoma.

A single primary cancer with two tumors on base of tongue with keratinizing squamous cell carcinoma invading the submucosa and squamous cell carcinoma invading the muscle of the tongue will be coded as squamous cell carcinoma.
Head and Neck Histology Rules

H3, H9 – Single Histology

H4, H10 – Most Invasive Histology

H5, H11 – Most Specific Histology

H6, H12 – Higher ICD-O-3 Code
Three innovations were introduced in the MP/H format to assist in the assignment of correct histology codes to primary cancers:

- Histology trees
- Tables of related cell types
- Tables of combination codes
We have already looked at histology trees used to display family relationships among histology groups.

The trees for malignant and benign CNS tumors are used in both multiple primary and histology determinations.

The trees for head and neck and lung tumors are referred to by the histology rules only.
For the lung, Chart 2 is a simplified version of Chart 1, displaying the relationships among the most common histologies in lung cancer.

Note the same branching structure in both of these charts, with more specific histologies located on the lower branches.

Also note that all histology statements on these two charts are coded, in contrast to the CNS charts where non-coded categories were identified.
Note that if you see a diagnostic statement with multiple terms in the same box on the head and neck chart, the histology rules will lead us to the numerically higher code.

If you see a diagnostic statement with multiple terms in the same box on the lung chart, the histology rules will lead us either to the numerically higher code or to a combination code.
The lung chart shows that carcinoid and small cell carcinoma are more specific diagnoses than neuroendocrine carcinoma.

This relationship is not explicitly stated in the rules for other primary sites, but SEER answers to registrars’ questions about these histologies support this interpretation; a diagnosis of small cell carcinoma with neuroendocrine differentiation would be coded as 80413, a diagnosis of carcinoid, neuroendocrine carcinoma of unknown primary site would be coded as 82403.
We have looked at the tables listing specific types of ductal, intraductal, renal cell, and urothelial cancers, not identifiable by ICD-O coding structure; these tables are also referred to in both multiple primary determination and histology coding.
The third innovation is the introduction of charts showing the mapping between multiple histologic types combined into complex morphology codes for single primary cancers.

The ICD-O-3 morphology list was expanded with the addition of complex or mixed histology codes, creating a coding challenge and a need for the development of rules directing their use.

These mixed histology tables are included for lung, breast, and other sites.

The kidney module does not refer to a combination code table, but does include a rule for coding mixed renal cell types.

Likewise the malignant CNS histology module does not refer to a combination code table, but includes a rule for coding mixed glioma.
Most of the lung combination codes are for squamous cell and adenocarcinoma histologies.

Note that Table 1 for lung is used to code combination histologies in single tumors only, and does not direct the use of a combination code when the histologies occur in separate tumors.

This table lists required terms in the first column, additional required terms in the second column, and then the ICD-O-3 term and code for the combined histologies.

The table itself contains additional coding instructions: the small cell combination code is available for “small cell” only and not a subtype of small cell, and the adenocarcinoma and squamous cell carcinoma combination code is available for “adenocarcinoma” only and not a subtype of adenocarcinoma.

The table does not indicate that subtypes of squamous cell carcinoma must not be considered.
Breast

Table 3 in the breast rules presents the combination codes for this site.

This table is referenced by the three histology modules for single in situ tumors, single invasive tumors, and multiple tumors.

Similarly to the lung table, the columns show a required histology, the combining histologies, the ICD-O-3 combination term, and the ICD-O-3 code for the combination term.

To be elaborated on in the site by site review of rules, the combination shown in the fifth row of the table creates a conflict with histology rules specifying that the more specific histologic type should be coded when the diagnostic statement identifies “duct” and one specific type of duct histology.

The rules should take precedence over the table statement.
Other Sites

The combination code table for other sites is similar in structure to the tables for lung and breast.

This table also reference by the three histology coding modules for single in situ, single invasive, and multiple tumors.

The histology combinations here reflect the variety of sites that are included in the other sites rules, with combinations for small cell and squamous cell carcinomas, hepatocellular carcinoma, adenocarcinoma, and thyroid, gynecologic, and germ cell malignancies.
Looking across the histology coding modules for the primary sites, we can identify a number of rules which are site-specific, leading to correct histology coding decisions that do not fit into the general categories we have already examined.

These rules usually occur early in the lineup, so that special conditions are identified and handled correctly before the coder begins to consider more generic rule statements.

- For example, inspecting the rules for coding histology for a single colon tumor, after the first two rules relating to clinical statement and metastatic site coding, we encounter eight rules that are specific to colon histologies: Rule H3 corrects misuse of a code that does not describe a colon histology, Rule H4 specifies correct coding when colon polyps are diagnosed, Rules H5, H6, and H7 all address correct coding of mucinous and signet ring histologies in the colon, and Rules H8, H9, and H10 all address correct coding of carcinoid tumors.

Reviewing the rules for coding histology when multiple colon tumors are abstracted as a single primary, we again see five rules, H17 through H21, that all address correct coding when colon polyps are diagnosed.
Similarly, we can identify histology coding issues for other sites by looking at the special rules that have been included.

Reviewing the histology rules for cutaneous melanoma, we see two rules directing coding for diagnoses that include a description of regressing melanoma, and two rules directing coding for diagnoses including lentigo maligna.

The breast module contains a special rule for coding inflammatory carcinoma as a pathologic versus clinical diagnosis.

For the urinary sites, special rules are included to guide coding of transitional cell/urothelial carcinoma histologies, in both the single tumor and multiple tumor modules.

The benign CNS histology module includes a special rule for coding multiple meningiomas of uncertain behavior.
A final point to emphasize again, for both Multiple Primary and Histology rules, is the general principle that the first rule that applies represents the decision point, later rules are not considered for further guidance in abstracting a case.
The rules are site-specific in how they are presented.

In some cases the earlier rules cover the exceptional situations that are rarely encountered and later rules are generally applied; in other cases the earlier rules cover usual situations and the later rules are rarely used.
There is also a related interplay between the Multiple Primary and the Histology coding rules.

- For example, if the correct Multiple Primary rule for a case situation states that tumors with histology codes differing among the first three characters represent multiple primaries, and one of the rules for coding multiple tumors abstracted as a single primary states that the numerically highest ICD-O-3 code is the correct code, then by definition the first three characters of the ICD-O-3 histology code must be the same for this primary cancer, and the histology code selection for the cancer will be based on the fourth ICD-O-3 character.
In summary, we use the MP/H rules to determine number of primary cancers and to assign histology codes to those cancers for all cases diagnosed from January 1, 2007 onward. Using the rules, we first determine the location and number of tumors from documentation in the medical record, eliminating tumors from consideration which are known metastases from previously diagnosed cancers.

Referring to the multiple primary rules for the appropriate site, and selecting the correct multiple primary module for the number of tumors, we determine the number of primary cancers based on matching the rules to the case circumstances; the first matching rule supplies the answer.

Then for each cancer that is identified, we determine the correct histology module to use based on single versus multiple tumors for that cancer; again, the first rule matching the case circumstances supplies the answer.

We encounter many similar multiple primary and histology rules across the modules for the different primary sites, but the ordering of the rules may be different across the sites.
We also note that some of the rules included in the first published version have undergone revisions which have been posted on the SEER website.

The rules have also been subject to interpretation and clarification through the online inquiry systems hosted by the Commission on Cancer, the Inquiry and Response system or IandR, and hosted by SEER, the SEER Inquiry System or SINQ.

SEER has also conducted a reliability study of the rules in the fall of 2008, and we can anticipate further revisions to the published rules addressing identified issues and clarifying their use by registrars.
Thus we can anticipate that the MP/H rules will continue to be a dynamic coding system, based on requests for assistance in their interpretation, on modifications to their structure resulting from both coding quality and cancer trending studies, and on identification of and discrimination among new histologic types embodied in future WHO publications on histology.