Multiple Primary and Histology Site Specific Coding Rules
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

HEAD AND NECK
The primary coding question with head and neck sites is the determination of primary site.

The MP/H rules provide guidelines for coding the primary site, based on a hierarchy of sources including physician information, pathologic diagnostic information, and diagnostic test information.

The terms and definitions section also defines levels of invasion for head and neck sites, provides a list of paired sites, and a family tree of histology groups and types.

Multiple anatomic diagrams help in locating structures in the head and neck and seeing their relationships to each other.
The Head and Neck sites share the first two multiple primary rules common across most sites:

- M1, if it is not possible to determine if there is a single tumor or multiple tumors, the case is abstracted as a single tumor.
- M2, a single tumor is always a single primary.

Examples given for the first rule include cases where there may be a discrepancy between clinical documentation of tumor location and site of biopsy, which can frequently occur in head and neck cancers because of the complexity of structures and the location of many structures in close proximity to one another.

A physician may have biopsied only one area of a larger tumor, or a satellite tumor from a larger tumor, and complete documentation may not be available to the abstracter.
Ten rules are available for determining multiple primary cancers where multiple tumors are known to exist.

The first four rules address cases where the first three digits of the ICD-O-3 code are the same:

- M3, tumors on the right and left side of paired sites are multiple primaries.
- M4, tumors on upper and lower lip are multiple primaries.
- M5, tumors on upper and lower gum are multiple primaries.
- M6, tumors in the nasal cavity and middle are multiple primaries.

Then the next rule,

- M7 states that tumors with ICD-O-3 topography codes that do differ within the second or third characters are multiple primaries.
The two timing rules are next:

- M8, an invasive tumor diagnosed more than 60 days after an in situ tumor is a new primary.
- M9, tumors diagnosed more than five years apart are multiple primaries.

Two rules based on histology codes follow:

- M10, tumors with a non-specific histology and a more specific histology are the same primary.
- M11, tumors with ICD-O-3 histology codes that are different within the first three characters are multiple primaries.
The non-specific/specific rules pairs

- cancer/malignant neoplasm and a specific histology,
- carcinoma and a specific carcinoma,
- adenocarcinoma and a specific adenocarcinoma,
- squamous cell carcinoma and a specific squamous cell carcinoma,
- melanoma and a specific melanoma, and
- sarcoma and a specific sarcoma.

Note here the placement of a rule, based on histology relationships, that will probably cover many more cases than the more general rule based on the numeric value of the codes.
The final rule

- M12, states that any case involving multiple tumors that has not been decided by the preceding rules is abstracted as a single primary.

Examples of types of cases are given:

- multifocal tumors in a single site
- in situ and invasive tumors diagnosed within 60 days of each other
- in situ tumor diagnosed more than 60 days after an invasive tumor

These are examples only and not meant to be referred to as coding rules.
The histology rules are divided into two modules,

- coding of a primary cancer with a single tumor
- coding of a primary cancer with multiple tumors

The rules and their sequence are the same for both modules.

Again the first two rules are common across all sites:

- H1 and H7, code the histology documented by the physician when there is no pathology or cytology specimen or the report is not available.
- H2 and H8, code the histology from a metastatic site when there is no pathology or cytology specimen from the primary site.
The next rules probably cover most coding situations:

• H3 and H9, code the histology when there is only one histology type.
• H4 and H10, code invasive over in situ component for a single tumor, or the most invasive tumor for multiple tumors.

The next rules,

• H5 and H11, reflect back to the multiple primary rule for combining tumors with non-specific and specific histology statements, requiring that the more specific histologic term is coded over a non-specific term.

This rule also refers to the tree of head and neck histology types included for this group of sites.
As discussed previously, for all histology trees included in the MP/H rules, histologies on lower branches of the tree are considered more specific than histologies located higher on the tree, with histologies at the same level or grouped together in boxes not taking precedence over each other.

The final rules,

- H6 and H12, are applied only if a previous rule has not led to a decision: the histology with the numerically higher ICD-O-3 code is selected.
Applying the Head and Neck Rules

Simple set of case facts

- A patient presents with a sore on the underside of the tongue.
- Biopsies performed of the floor of mouth and tongue are read as squamous cell carcinoma in situ.
- Patient undergoes wide excision of the lesion, and on the operative report the surgeon notes 2 cm lesion of the ventral tongue with extension onto the floor of mouth.
- The pathology report reads “verrucous carcinoma of tongue with squamous cell carcinoma in situ extending close to but not involving margin.”
For coding, a single tumor is identified, and the site is determined to be ventral surface of anterior tongue, CO22, based on the operative report.

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

Reviewing the Single Tumor histology module, Rule H1 does not apply, there is a pathology report.

Rule H2 does not apply, there is a pathology report for the primary site.

Rule H3 does not apply, more than one histologic type is identified.
Multiple Primary and Histology Coding Rules
The Colon section provides rules for the colon sites only, rectosigmoid and rectum are included in the Other Sites group.

The Colon rules focus on three general guidelines:

- the importance of identifying and coding involvement of adenocarcinoma in adenomas or polyps
- the conditions under which mucinous or signet ring cell types of adenocarcinoma can be coded
- the correct use of carcinoid codes

The terms and conditions section does note that “exophytic” and “polypoid” are not to be read as synonymous with “polyp.”
The levels of invasion into and through the colon wall are identified, along with terms that may appear in a pathology report to indicate level of invasion, such as:

- intramucosal
- transmural

Some types of colon histologies are described in this section, which also contains a diagram of standard colon measurements that may be of assistance in locating primary site of tumors as described on colonoscopy.
The multiple primary rules for Colon are divided into the standard modules of

| The multiple primary rules for Colon are divided into the standard modules of | • Unknown if Single or Multiple Tumors,  
| • Single Tumor, and  
| • Multiple Tumors. |
|---|---|
| The first rule, | • M1, is a standard rule, if it is not possible to determine if there is a single or multiple tumors, consider the case as having a single tumor and abstract as a single primary. |
| The second rule, | • M2, is a standard rule, a single tumor is always a single primary. |
Looking at the Multiple Tumors module, the first rule here, M3, is very specific to colon cancers.

Adenocarcinoma in adenomatous polyposis coli or familial polyposis with one or more malignant polyps is a single primary.

This rule has been interpreted as applying when adenocarcinoma and the condition of familial polyposis occur together, even if the cancer is not specifically identified in the pathology report as arising from one of the polyps.
The next three rules are similar to rules in other sites.

- M4 states that tumors in sites with ICD-O-3 topography codes differing among all four characters are separate primaries.
- For some sites the fourth character of the ICD-O-3 topography code does not indicate a separate primary site, but this rule maintains the traditional separation of colon primary sites by subsite code (except for familial polyposis); thus a tumor in the hepatic flexure, C183, and a tumor in the transverse colon, C184, represent separate primary cancers.
- Rule M5 contains the timing rule for colon primaries, tumors diagnosed more than one year apart are multiple primaries.
- The next rule, M6, is standard across most sites, an invasive tumor following an in situ tumor more than 60 days after diagnosis is another primary cancer.
Next,

- Rule M7 is again a site-specific rule for colon cancers, a frank malignant or in situ adenocarcinoma and an in situ or malignant tumor in a polyp are a single primary.
- This rule is being applied after M3, so that the polyp and the invasive adenocarcinoma by definition would be occurring in the same subsite of the colon.
- Rule M8 is the standard rule about abstracting as a single primary a non-specific and a specific histology; the pairings for colon are cancer/malignant neoplasm and a specific histology, carcinoma NOS and a specific carcinoma; adenocarcinoma NOS and a specific adenocarcinoma; and sarcoma NOS and a specific sarcoma.
The next rule,

- M9, is again specific to colon, multiple in situ and/or malignant polyps are a single primary.

The final first three characters are multiple primaries.

- M11, tumors that have not met any of the previous criteria are abstracted as a single primary.
- Again, by the time Rule M11 is applied, all tumors are in the same subsite of the colon.
• H1 and H15 state that when no pathology/cytology specimen is taken or the report is not available, code histology from the medical information provided by the physician.

• Rules H2 and H16 state that if no pathology or cytology is available from the primary site but is available from a metastatic site, the diagnosis from the metastatic site is coded.
The next eight rules in the “Single Tumor” module address the histology issues noted.

- Rule H3 states that if a report diagnosis intestinal type adenocarcinoma of the colon, adenocarcinoma is coded; “intestinal type” applies to gastric cancers, it is not a specific type of histology for colon cancers.
- Rule H4 states that a polyp code should be used if there is any description of the cancer arising in a polyp.

The three polyp codes are:
- 8210, adenocarcinoma in adenomatous polyp
- 8261, adenocarcinoma in villous adenoma
- 8263, adenocarcinoma in tubulovillous adenoma
Colon is unique in that this is the only site where the rules specifically state that, in two instances, information can be drawn from other parts of the pathology report or the medical record when there is a final pathologic diagnosis.

In the first instance, information about “polyp” can be taken from the microscopic part of the pathology report or from other documentation that the tumor arose in a polyp, such as an operative report describing a polypectomy.

Except for the case of familial polyposis though, the documentation must show that the carcinoma arose in the polyp.
The next rule addresses the second instance where information can be taken from the microscopic section of the pathology report to aid in histology coding.

As stated in the next rule:

- H5, mucinous/colloid adenocarcinoma 8480/3, or signet ring cell carcinoma, 8490/3, is coded when the final diagnosis states mucinous/colloid or signet ring cell; or the final diagnosis states adenocarcinoma NOS and the microscopic documents that 50% or more of the tumor is mucinous/colloid, or 50% or more of the tumor is signet ring cell.
The next rule,

• H6, explicitly presents the instruction for coding when the final diagnosis is adenocarcinoma NOS and the microscopic states that less than 50% of the tumor is mucinous/colloid or signet ring cell, or the percentage of mucinous/colloid or signet ring cell involvement is not stated.

The next rule,

• H7 is the final of the three rules addressing mucinous/colloid and signet ring cell types of adenocarcinoma; the code for adenocarcinoma of mixed types, 8255/3, is used when there is a combination of the two histologies. Mucinous/colloid and signet ring cell histologies are both associated with mucin; in the mucinous/colloid type, the mucin is extra-cellular, and in the signet ring cell type, the mucin is intra-cellular.
The next trio of rules address problems in coding carcinoid histologies in the colon.

- Rule H8 states that carcinoid NOS, 8240/3, is coded when the diagnosis is neuroendocrine carcinoma and carcinoid tumor.
- Rule H9 states that composite carcinoid, 8244/3, is coded when the diagnosis is adenocarcinoma and carcinoid tumor.
- Rule H10 states that adenocarcinoid, 8245/3, is coded when the diagnosis is exactly adenocarcinoid.
The final four rules in the “Single Tumor” module for colon are standard rules:

- **H11**, code the histology when a single histologic type is identified.
- **H12**, code the invasive histology when both in situ and invasive components are identified.
- **H13**, code the most specific histologic term.
- **H14**, code the histology with the numerically highest ICD-O-3 code.
The non-specific/specific histology pairs for colon are:

- cancer/malignant neoplasm NOS and a more specific histology
- carcinoma NOS and a more specific carcinoma
- adenocarcinoma NOS and a more specific adenocarcinoma
- sarcoma NOS and a more specific sarcoma
The Multiple Tumors Abstracted as a Single Primary module contains the same standard rules as the Single Tumor module, though in a slightly different order, and rules relating to coding multiple polyps.

We have already reviewed H15 and H16.
The first polyp rule,

- H17, adenocarcinoma in adenomatous polyposis coli, 8220/3, is coded when there is a clinical diagnosis of familial polyposis and a pathologic diagnosis of adenocarcinoma in adenomatous polyps, there are more than 100 polyps in the resected specimen, or the number of polyps is not specific but the diagnosis is stated as familial polyposis.
• H18, states that the highest polyp code, 826/3, is coded when there are multiple in situ or malignant polyps present, at least one of which is tubulovillous; this rule however does not apply to the combination of a frank adenocarcinoma and adenocarcinoma in tubulovillous adenoma.
The third polyp rule,

- H19, can be compared with rule H17; 8221, adenocarcinoma in multiple adenomatous polyps, is coded when there are between 1 and 100 polyps with adenocarcinoma in the resected specimen, or there are multiple polyps with adenocarcinoma, the number is not given, and familial polyposis is not mentioned.
- Relating this rule to the multiple primary rules, the multiple polyps would have to be found in the same segment of the colon to be considered as a single primary coded with this rule.
The next rule is standard across many sites,

- H20, code the histology of the most invasive tumor.
- This rule would apply in the case of a frank adenocarcinoma and adenocarcinoma in a polyp, with the histology of the more invasive tumor being coded.
- The rule specifically states that if the tumors being considered are equally invasive, this is not the final rule for the case.

Rule H21 is again a polyp rule, similar to H4 for single tumors: adenocarcinoma in a polyp is coded if there is any documentation about a tumor arising in a polyp, elsewhere in the pathology report or in the record.
The last three rules in the module are again standard rules:

- **H22**, code the histology when only one histology type is identified.
- **H23**, code the more specific histology.
- **H24**, code the histology with the numerically higher ICD-O-3 code.
Taking a “polyp” case for practice

- Patient presented for screening colonoscopy, with multiple polyps found and biopsied.
- Biopsy was positive for adenocarcinoma in polyp in the sigmoid colon, and a large adenoma with adenocarcinoma in the cecum.
- The margins were clear on the sigmoid polypectomy, and the patient was taken to surgery for right hemicolecotomy.
- Final pathologic diagnosis at surgery indicated a 4cm x 5cm adenocarcinoma arising in a tubulovillous adenoma of the ascending colon invading into the subserosal fat, with a second smaller adenocarcinoma 2cm distal to the cecum also invading into the subserosal fat, with 3 of 12 pericolonic lymph nodes positive for metastatic adenocarcinoma.
- Three tumors are identified: one in the sigmoid colon and two in the ascending colon, assuming that the adenocarcinoma in the cecum at colonoscopy is one of the two tumors of the ascending colon at hemicolecotomy.
Going to the “Multiple Tumors” module to determine the number of primary cancers, you note that M3 does not apply, you do not have a diagnosis of familial polyposis.

Rule M4 does apply, there are tumors in the sigmoid colon and the ascending colon, so at least these are separate primary cancers.

There are still two tumors in the ascending colon to decide about.
Rule M5 does not apply, these tumors were diagnosed at the same time, and for the same reason Rule M6 does not apply.

Rule M7 does apply, we have a frank adenocarcinoma and a tumor in a polypl in the ascending colon, so these two tumors are a single primary.

We stop, and go to the histology coding modules.
Taking the single tumor in the sigmoid colon first and going to the “Single Tumor” module, Rules H1 and H2 do not apply, there is a pathology report for the primary site tumor.

Rule H3 does not apply, this is not a diagnosis of “intestinal adenocarcinoma.”

Rule H4 does apply, the diagnosis is adenocarcinoma in a polyp, and the code for this histology is 82103.
There are two tumors abstracted as a single primary in the ascending colon, so use the “Multiple Tumors Abstracted as a Single Primary” to assign the histology code for this cancer.
Again, Rules H15 and H16 do not apply, there is a pathology report from the primary site.

Rule H17 does not apply, this is not a case of familial polyposis.

Rule H18 does not apply, we have a frank adenocarcinoma and adenocarcinoma in a polyp for this primary, rather than multiple malignant polyps.
Rule H19 again does not apply, this is not a case of multiple malignant polyps.

Rule H20 does not apply, both tumors are equally invasive into the subserosal fat.

Rule H21 does not apply, we have a frank adenocarcinoma and an adenocarcinoma in tubulovillous adenoma, and H21 only refers to the polyp codes.
Rule H22 does not apply, there is more than one histologic type.

Rule H23 does apply, there is an adenocarcinoma and a more specific type of adenocarcinoma, adenocarcinoma in tubulovillous adenoma.

The histology code for this second cancer in the ascending colon is 8263/3.
Multiple Primary and Histology Coding Rules

LUNG
The primary question with lung cancers is the determination of multiple primary cancers when both lungs are involved with one or more tumors, and the correct selection of histology code when multiple histologic diagnoses may be rendered.

The rules and definitions section provides information about histologic terms, such as large cell neuroendocrine carcinoma, pleomorphic carcinoma, and sarcomatoid carcinoma, and a description of the anatomic designation of Pancoast tumor.

A family tree is provided for lung histology groups, similar to the tree for head and neck histologies, with again the more specific histologies located further down on the chart.

The Lung section also contains the first combination histology chart, a table which shows the appropriate combination histology code to use when specified histologies occur together in a single tumor.
The multiple primary rules for Lung contain the common M1 and M2 rules, unknown number of tumors abstracted as a single primary and a single tumor always a single primary.

The example given for one situation where the number of tumors may be unknown is the case of two or more tumors in one lung and one or more tumors in the second lung and only one tumor is biopsied.

The case of one tumor in each lung with only one tumor biopsied is covered by another rule in the Multiple Tumors module, and is not considered to fall within the jurisdiction of the M1 rule.
There are ten rules that apply in multiple tumor cases.

The first rule,

- M3, relates to topography, tumors differing within the first three characters of the ICD-O-3 code are separate primaries; the change from previous rules for lung is to separate tracheal from lung primaries.

The next two rules relate to histology:

- M4, non-small cell and small cell carcinomas are separate primaries.
- M5, adenocarcinoma with mixed subtypes and bronchioloalveolar carcinomas are separate primaries.
- These two rules are included to identify as separate primaries histology codes which happen to have the same first three characters, another break from one of the more general histology rules.
The next two rules address two situations in which both lungs are involved:

- M6, a single tumor in each lung represents multiple primaries, unless one of the tumors is stated or proven to be metastatic from the other.
- M7, multiple tumors in both lungs with ICD-O-3 histology codes differing among the first three characters are multiple primaries.

The timing rules for lung follow:

- M8, tumors diagnosed more than three years apart being multiple primaries.
- M9, the standard rule for invasive tumors more than 60 days after in situ tumors being multiple primaries.
Two more histology rules follow;

- M10, non-small and more specific non-small cell carcinomas are a single primary, with reference to the lung histology tree.
- M11, tumors with ICD-O-3 codes differing among the first three characters are multiple primaries.

Here again, note the interplay of rules, with M10 taking care of the exceptions to the more general histology code rule embodied in M11.
The final rule, M12, treats all case situations which have not been decided by previous rules as single primaries.

Within the group of examples for this rule fall other situations with bilateral lung involvement: solitary tumor in one lung with multiple tumors in the contralateral lung, diffuse bilateral nodules, and multiple tumors in both lungs.

Presumably all the tumors in the last example would have a histologic diagnosis and thus not be included within the example for the M1 rule.
Multiple tumors in a single lung is also given as an example for this last default rule.

Similarly to the Head and Neck histology rules, the Lung histology rules are the same for both the Single Tumor module and the Multiple Tumors Abstracted as a Single Primary module, with the exception of the addition of the combination histology rule for single lung tumors.
The histology rules in order:

- H1 and H8, code the histology documented by the physician when no pathology or cytology report is available or obtained.
- H2 and H9, code the histology or cytology from a metastatic site when no specimen is taken from the primary site.
- H3 and H10, code the histology of a single histologic type.
- H4 and H11, code the invasive histology when a single tumor has both invasive and in situ components or the most invasive histology when two or more invasive tumors.
- H5 and H12, code the most specific histologic term when non-specific and more specific histologies are stated.
The non-specific histologies for lung are listed as:

- cancer/malignant neoplasm and a more specific histology
- carcinoma and a more specific carcinoma
- adenocarcinoma and a more specific adenocarcinoma
- squamous cell carcinoma and a more specific squamous cell carcinoma
- sarcoma and a more specific sarcoma

The next rule,

- H13, in the Single Tumor module refers to selection of a combination code from the table when two or more specific histologies are identified within a single tumor.
Note the absence of this rule in the Multiple Tumors Abstracted as a Single Primary module.

Multiple tumors will follow their own path through the rules, which should not lead to an end point of combining separate adenocarcinomas and squamous cell carcinomas into adenosquamous carcinomas.

The final rule for both single and multiple tumor coding,

- H7 and H13, is to code the histology with the numerically higher ICD-O-3 code.
Investigating further the interplay between the histology tree and the combination code table, as referenced by rules H5 and H6, note that the combination codes are used for histologies which reside on different branches of the tree (small cell and adenocarcinoma), and for histologies grouped together at the same level on the same branch (acinar and papillary adenocarcinoma).

Also note the overlap between Rule H5 and Rule H6 in coding squamous cell carcinoma and squamous cell nonkeratinizing carcinoma as either a non-specific histology with a more specific histology, or as a combined histology.

You conclude that if you were presented with a single tumor with multiple histologies that were not included in the combination chart, your coding decision would be directed by Rule H7 and you would use the numerically highest ICD-O-3 code.
Applying the Lung rules to a set of case facts

- A patient presents with long smoking history and a cough.
- A mass is noted in the right upper lobe of the lung on chest x-ray, and CT-scan shows the 5cmx2cm mass in the right upper lobe and also a second smaller mass in the right lower lobe.
- The right upper lobe mass is biopsied and diagnosed as a squamous cell carcinoma.
- The patient undergoes mediastinoscopy, with negative biopsy of three mediastinal nodes.
- The patient then undergoes right upper lobectomy and wedge resection of the right lower lobe, with diagnosis of squamous cell carcinoma of the right upper lobe and peribronchial lymph nodes, and small cell carcinoma of the right lower lobe.
- Two tumors are identified.
Consulting the Multiple Tumors module to determine the number of primary cancers, Rule M3 does not apply, as the first three characters for the site codes for both tumors are the same, C34.

Rule M4 does not apply, as neither of the tumors is diagnosed as non-small cell carcinoma, though one is diagnosed as small cell carcinoma.

Rule M5 does not apply, neither of the tumors are diagnosed as adenocarcinoma with mixed subtypes or bronchioloalveolar carcinoma.
Rules M6 and M7 do not apply, only one lung is involved.

Rules M8 and M9 do not apply, both tumors are diagnosed at the same time.

Rule M10 does not apply, again neither tumor is diagnosed as non-small cell carcinoma, though one is diagnosed as a more specific type of non-small cell carcinoma.
Rule M11 does apply, the histology codes for the two tumors are different within the first three characters, “80703” and “80413.”

These are multiple primary cancers.
Histology is determined for each cancer using the same Single Tumor module in the histology rules.

Rules H1 and H2 do not apply, there is a pathology report with a diagnosis for each primary site.

Rule H3 does apply to each case, only one histologic type is identified for the right upper lobe tumor, squamous cell carcinoma, 8070/3, and only one histologic type is identified for the right lower lobe tumor, small cell carcinoma, 8041/3.
Multiple Primary and Histology Coding Rules

CUTANEOUS MELANOMA
The rules in the Melanoma modules apply to cutaneous melanomas only.

Melanomas arising elsewhere are abstracted according to the appropriate modules for those sites.

The terms and definitions section for melanoma identifies the majority of melanoma histology types as:

- acral melanoma
- desmoplastic melanoma
- lentigo maligna
- superficial spreading melanoma
- nodular melanoma
Synonyms for in situ are listed, including:
- intraepidermal
- intraepithelial
- Clark level I

Two terms critical to correct coding of melanomas are identified:
- in transit or satellite metastases
- laterality

Midline is noted as a separate laterality for melanoma.
The significance of regressing melanoma is also discussed.

The layers of the skin are identified and illustrated with anatomic diagrams.

The multiple primary rules include the standard M1 rule for unknown number of tumors and M2 rule for single tumors.
The Multiple Tumors, or Multiple Melanomas, module includes six rules.

The first rule is a variation on the standard rule for ICD-O-3 topography codes: sites with topography codes differing at the level of the fourth character are also considered separate primaries, so that a melanoma of the leg, C447, and a melanoma of the trunk, C445, would be separate primaries by Rule M3.

The next rule, M4, specifies that melanomas with different literalities are different primaries, and again midline is considered a laterality, so that melanomas of the right back and mid back would be separate primaries.

The next rule, M5, is the standard rule for ICD-O-3 histology codes differing among the first three characters.
The next two rules state timing for melanomas:

- Rule M6, an invasive melanoma diagnosed 60 days after in situ melanoma considered a second primary, is actually a redundant statement of Rule M7, any melanoma diagnosed more than 60 days from a previous melanoma considered a second primary.

The final rule,

- M8, is the default rule, any case situation not falling within one of the previous rules represents a single primary.
Examples given for this rule include melanomas located on the front and back of the same lateral site, such as left chest and left back, and melanomas located on different parts of the same limb, such as right wrist and right elbow.

Again, these are examples and not statements of rules.

The most counter-intuitive result is probably coding anterior and posterior melanomas with the same laterality as a single primary.
The Melanoma section is unique in that there is only one set of histology rules for both Single Melanoma and Multiple Melanomas Abstracted as a Single Primary.

The first four rules are standard across many sites:

- **H1**, code histology from a physician’s statement when pathology or cytology report not available or specimen not taken.
- **H2**, code cytology or histology from a metastatic site when no specimen from the primary site.
- **H3**, code histology when a single histologic type is identified.
- **H4**, code the invasive histology when both in situ and invasive components are diagnosed.
The next four rules are specific to melanoma:

- **H5**, code the specific histologic type when both regressing melanoma and another specific histologic type are identified.
- **H6**, code regressing melanoma when that is the only specified type.
- **H7**, code the specific histologic type when both lentigo maligna melanoma and another specific histologic type are identified.
- **H8**, code lentigo maligna melanoma when that is the only specific histologic type.
The next two rules are again standard across many sites:

- **H9**, code the most specific type when the diagnosis is melanoma and a single specific type.
- **H10**, code the histology with the numerically higher ICD-O-3 code.

Rule H9 does vary from the formula for the standard “specific type” rule, in that it includes the word “single”, though this word is implicit in other statements of this rule.

A single primary with a diagnosis of melanoma and two specific types would be explicitly coded according to the final histologic rule for melanomas, with the higher histology code.
Let us apply the rules for a melanoma case.

- A patient is diagnosed on punch biopsy with a superficial spreading melanoma of the right anterior thigh, at least Clark level II, Breslow depth 0.28 mm, and acral lentiginous melanoma of the right foot, Breslow depth 3.40 mm.
- At wide excision of both lesions, the diagnosis was residual melanoma in situ right anterior thigh, residual melanoma of right foot.
This patient has multiple melanomas, so the Multiple Melanomas module is consulted to determine number of primary cancers.

Rule M3 does not apply, because the lesions are both located on the right leg, C447.

Rule M4 does not apply, again because the lesions are both on the right leg.
Rule M5 does not apply because the melanoma codes do not differ within the first three characters, “8743/3” and “8744/3.”

The lesions are diagnosed at the same time, so rules M6 and M7 for melanomas diagnosed more than 60 days apart do not apply.

Rule M8 does apply, these melanomas do not meet any prior criteria, so they are a single primary.
Reviewing the pathology reports, for both melanomas the punch biopsy procedure seems to represent the most definitive tumor resection, so the selection of codes is between the specific histologies, superficial spreading melanoma and acral lentiginous melanoma.

Applying the histology rules, Rules H1 and H2 do not apply because a pathology report is available for the primary site tumors.

Rule H3 does not apply, there is more than one histology diagnosed.
Rule H4 might apply, specifying that invasive histology should be coded over in situ histology, but you have eliminated from your consideration the diagnoses from the wide excision report.

Rules H5, H6, H7, and H8 do not apply, because there is no statement of regressing melanoma or lentigo maligna melanoma.
Multiple Primary and Histology Coding Rules
The Breast rules focus on correct coding for ductal carcinoma, the most common type of breast histology, but also a histology with many subtypes that may appear in combination with other subtypes; ductal carcinoma may also appear in combination with lobular carcinoma, either in a single lesion or each histology in a separate lesion.

Many breast carcinomas may also be diagnosed as in situ lesions, and two additional histology rule modules are provided for coding in situ lesions and in situ and invasive lesions diagnosed together.

The terms and definitions sections notes that “and” and “with” are synonymous in diagnoses stated as “ductal and lobular” and “ductal with lobular.”

“NST”, no special type, is also noted as synonymous with “NOS,” not otherwise specified.
The terms and definitions section describes certain terms that may be encountered in abstracting breast cancers, such as

- intracystic carcinoma
- Paget disease
- Phyllodes tumor
- scirrhus carcinoma

Two tables list types of intraductal and ductal carcinomas to aid in identifying these diagnoses; these tables are also referred to by the multiple primary and the histology rules.

A third table identifies combination codes which may be used for mixed histologies, which are frequently diagnosed in breast cancers.
The table lists required histologies in the first column, combining histologies in the second column, and the combination histologic terms and ICD-O-3 codes for these terms in the third and fourth columns.

Again, this table is referred to by both multiple primary and histology rules.

A diagram of breast anatomy shows the ducts and lobules wherein carcinomas arise, and the surrounding muscular and skeletal structures which may be directly invaded.
The multiple primary rules are presented in the three standard modules.

- The Unknown if Single or Multiple Tumors module has the standard M1 rule, when it is not possible to determine if a single or multiple tumors are presented, the case is abstracted as a single primary.

- The Single Tumor module contains a somewhat anomalous rule, M2, inflammatory carcinoma in one or both breasts is a single primary. Presumably this rule addresses the situation where inflammatory carcinoma in the first breast is perceived as directly extending into the second breast, and this rule is placed before M3 to emphasize the concept of this condition as a single primary.

- The third rule, M3, is the standard, a single tumor is always a single primary.
Going to the Multiple Tumors module, the first rule,

- **M4**, is a standard rule, tumors in sites with histology codes differing among the first three characters are multiple primaries.

Breast subsites are differentiated at the fourth character, so the impact of this rule is to distinguish breast cancers from other sites within the body, but not to distinguish among multiple tumors within different subsites or quadrants of the breast.

The next rule,

- **M5**, is the timing rule for breast: tumors diagnosed more than five years apart are multiple primaries.
The next rule,

- M6 restates M3, inflammatory carcinoma in one or both breasts is a single primary; this rule comes after the timing rule, so inflammatory carcinoma in the second breast appearing more than five years after inflammatory carcinoma in the first breast would be a new primary.

The following rule,

- M7, states that tumors in both right and left breasts are multiple primaries; the case of bilateral inflammatory carcinoma, appearing before M7 and thus already dealt with, would be the single exception to this rule.

The next rule,

- M8, is a standard rule for most sites, an invasive tumor diagnosed more than 60 days after an in situ tumor is a new primary cancer.
The following three rules are related to specific conditions dealing with duct carcinoma, in combination with Paget disease, lobular carcinoma, and specific duct histologies.

- Rule M9 states that tumors with intraductal or duct histology and Paget disease are single primaries.
- Rule M10 states that tumors with intraductal or duct histology and lobular histology are single primaries.
- The third rule, M11, states that multiple intraductal and/or duct carcinomas are single primaries.

These rules refer to the tables listing intraductal and duct carcinoma histology types.
The final two rules are standard across most sites.

- Rule M12 states that tumors with ICD-O-3 histology codes differing among the first three characters are multiple primaries.
- The last rule, M13, provides the default criterion, any case situation not decided by previous rules is abstracted as a single primary cancer.

The pertinent example here is multiple lobular carcinomas existing in the same breast.
The first histology module is for Single Tumor: In Situ Carcinoma Only.

Many of the rules in this module are similar to rules in the Single Tumor: Invasive Carcinoma Only and Multiple Tumors Abstracted as a Single Primary modules, but the rules are not exactly the same and the ordering of the rules varies across these three modules.

The first rule in the in situ histology module,

- H1, is modified from the standard H1 rule, code the histology documented by the physician when the pathology/cytology report is not available.
- An in situ carcinoma can only be diagnosed on examination of a histologic specimen, so this rule does not contain the wording about a specimen not being taken.
- For a similar reason, the usual rule about coding histology from a metastatic specimen is not included, as the presence of a metastatic lesion by definition would remove the cancer from the in situ category.
Rules H2 and H3 are standard rules,

- H2 stating to code the histology when only one histologic type is identified.
- H3 stating that the more specific histology is coded when a non-specific and a specific term are used.
- The histology pairings for this rule are carcinoma in situ NOS and a more specific carcinoma in situ, adenocarcinoma in situ NOS and a more specific adenocarcinoma in situ, and intraductal carcinoma NOS and a more specific intraductal.
As shown below, the rule for intraductal carcinoma NOS and a more specific intraductal carcinoma applies in cases where there is only one more specific intraductal carcinoma diagnosed.

The list of terms that identify specific types for this rule do include “architecture” and “pattern”; the general rule for application across all sites is that “architecture” and “pattern” are only valid for identifying subtypes of in situ carcinomas.
• Rule H5 states that any combination of in situ lobular and in situ ductal carcinomas is coded as 8522/2; the specific ductal types that are to be considered in applying this rule are listed in Table 1.

• Rule H6 states that a combination of intraductal histology with two or more specific intraductal histologies, or any combination of two or more specific intraductal histologies, is coded as 8523/2.

• Rule H3 has taken care of the case where there is a combination of intraductal carcinoma and one specific type of intraductal carcinoma, which is coded to the one specific type rather than the combination code.

• Rule H7 states that any combination of lobular histology with other histologic types is coded as 8524/2.
The final rule in this module:

- H8, provides for coding combinations of in situ carcinomas that do not include either intraductal or lobular histologies, using the code for adenocarcinomas of mixed histologies, 8255/2.

The situation that is not covered by this set of rules is how to code a combination of intraductal with another in situ histology that is neither a type of intraductal or lobular carcinoma.

If this case situation should arise, perhaps the best route would be to apply the default rule of coding to the numerically highest ICD-O-3 code, while submitting an inquiry to SEER for guidance.
The Single Tumor: Invasive and In Situ Carcinoma module has a single rule,

- H9, code the invasive histology when both invasive and in situ components are present.

This rule is adequate if there is only a single histologic type contained in the diagnosis statement.

If there is more than one type, then rules from the next module, Single Tumor: Invasive Carcinoma Only, must be stepped through to determine the correct coding for the invasive component of the lesion.
Reviewing the rules in the Single Tumor: Invasive Carcinoma Only module, we see the standard first two rules:

- H10, code the histology documented by the physician when there is no pathology/cytology specimen or the report is not available.
- H11, code the histology from the pathology/cytology report for a metastasis when there is no specimen from the primary site.
• H12 is the standard rule about coding specific over non-specific histologies.
• The pairings for this module include carcinoma NOS and a more specific carcinoma, adenocarcinoma NOS and a more specific adenocarcinoma, duct carcinoma NOS and a more specific duct carcinoma, and sarcoma NOS and a more specific sarcoma.
• This rule differs from the similar rule in the in situ module, in that sarcoma is included here (sarcoma by definition not including in situ carcinomas), and the words “architecture” and “pattern” are missing from the list of diagnostic terms which identify histology subtypes.
The next rule is new to this module,

- **H13**, code inflammatory carcinoma when the final diagnosis of the pathology report specifically states inflammatory carcinoma.

Again, an inflammatory carcinoma by definition will not be an in situ carcinoma; “inflammatory” is also a clinical-pathologic diagnosis, and this presentation is coded in the Collaborative Stage extension field, the histology code being used to reflect the actual pathologic diagnosis rendered.

The next rule,

- **H14**, is a standard rule, code the histology when a single histologic type is specified.
The next rule,

• H15 states that the combination of two or more specific duct carcinomas is to be coded to the histology with the numerically higher ICD-O-3 code.
• This rule thus differs from the in situ rule for the same situation, which indicates that the duct combination code, 8523/2, would be used to code two or more specific types of intraductal histologies.

Comparing these two rules emphasizes the importance of determining the correct coding module and strictly following the order of the rules and paying close attention to their wording.

Next,

• Rule H16 is similar to H5 for in situ, use the combination code for duct and lobular, 8522/3, when both these histologies are diagnosed.
• Table 3 provides the list of duct histologies which can be considered.
The next rule,

- H17 again takes a departure from the in situ rules, in that the duct combination code, 8523/3, is used when there is a combination of duct carcinoma and any other carcinoma excluding duct and lobular carcinomas.

As noted in reviewing the in situ rules, this situation was not covered in that module.
The next two rules, H18 and H19 are similar to the in situ rules for these combinations:

- Rule 18, use the combination code for lobular combined with histologies other than duct, 8524/3.
- Rule 19, use the mixed adenocarcinoma combination code for combinations other than duct and lobular, 8255/3.

Compared to the in situ tumor module, this single tumor module contains an ambiguity for the case situation where the diagnostic statement identifies a non-specific duct carcinoma and two or more specific duct carcinomas.

Rule H6 handles this case in the in situ module: use the combination code, with reference to Table 3.
Table 3 indicates a possible answer in the row headed “Infiltrating duct and one or more of the histologies in Column 2”, but the pertinent invasive rules, H12 and H15, do not refer to Table 3, and coding ductal plus one specific ductal histology with the combination code contravenes Rule H12.

If this case situation should arise, perhaps the best route would be to proceed to Rule H15 and code the numerically highest ICD-O-3 code, while submitting an inquiry to SEER for guidance.
The Multiple Tumors Abstracted as a Single Primary module again contains rules both similar to and different from the previous modules.

The first two rules are very familiar,

• H20 and H21, code the histology documented by the physician when no pathology/cytology specimen or report not available, and code the histology from the pathology/cytology specimen from a metastatic site.

The next rule,

• H22, is like H13 in the single tumor module, code inflammatory carcinoma, 85303, only when specifically stated as the final pathologic diagnosis.

Next,

• Rule H23 is similar to Rules H2 and H14, code the histology when only one histologic type is identified.
The next three rules are specific to the Multiple Tumor module, considering Paget disease as a separate tumor from an underlying tumor.

- Rule H24 provides that the code 85432 is used when the pathology report specifically states that Paget disease is in situ and the underlying tumor is intraductal carcinoma.
- It is interesting to note here that a similar rule is not stated in the in situ module for Paget disease diagnosed without an underlying tumor, that the behavior code can be changed from /3 to /2 if the pathologist specifically diagnoses Paget as in situ, though the wording for this rule carries that implication.
- Rule H25 states that the code 85433 should be used with a diagnosis of Paget disease and an underlying intraductal tumor when the pathologist does not state that the Paget disease is in situ.
- Rule H26 states that the code 85413 should be used when the diagnosis is Paget disease and an underlying invasive duct carcinoma.
Rules H24 and H25 refer to Table 1 to identify the types of intraductal histologies that can be considered, and H26 refers to Table 2 for the types of ductal histologies.

Next,

- Rule H27 is like Rule H9, the invasive histology is coded when both invasive and in situ tumors are present.

Again, if there is a single invasive histology diagnosed, this rule is sufficient for coding purposes.
If multiple types of invasive histology are abstracted in the single primary, then the rules in either the Single Tumor or Multiple Tumor module must be revisited at this point, depending on the case circumstances, to determine the final correct histology code.

The next rule, H28 is like the in situ Rule H5, and the single tumor Rule H16, use the combination code for duct and lobular, 8522, when both these histologies are involved.

This could be a case of either multiple in situ or multiple invasive tumors, but it would not be a case of mixed ductal in situ and invasive lobular, or lobular in situ and invasive ductal, as this latter situation would be eliminated by the preceding Rule H27.
The final rule in this module is the standard, • H29, code the histology with the numerically higher ICD-O-3 code.

Note that this standard rule appears in all modules (excluding the single-rule in situ/invasive modules for breast and other sites) with the exception of the single in situ and single invasive histology modules for breast.

Also note that this module does not contain the rules for using combination codes for multiple types of ductal or intraductal carcinomas that we saw in the in situ and single tumor modules; Rule H29 will tell us to use the higher ICD-O-3 code in all instances.
Case Example

- On annual mammogram, the patient is found to have two lesions in her right breast, 1.2 cm at 12:00 and 1.0 cm in the central breast.
- At biopsy, the 12:00 lesion is an infiltrating ductal carcinoma and the central lesion is an infiltrating lobular carcinoma.
- The patient undergoes mastectomy, with diagnosis of a 1.4 cm infiltrating duct carcinoma with cribriform ductal carcinoma in situ, a 3.0 cm area of infiltrating lobular carcinoma in the central breast, and a small 0.5 cm tubular carcinoma in the upper outer quadrant adjacent to the 12:00 lesion.
Reviewing the record, there are three tumors in the right breast.

Refer directly to the Multiple Tumors module to make a decision on number of primary cancers.

Rule M4 does not apply, the first three characters of the site code for all three tumors are “C50.”
Rule M5 does not apply, the tumors are diagnosed synchronously.

Rule M6 does not apply, this is not a diagnosis of inflammatory carcinoma.

Rule M7 does not apply, all the tumors are in the right breast.
Rule M8 does not apply, the in situ and invasive tumors were diagnosed at the same time.

Rule M9 does not apply, this is not a case of Paget disease.

Rule M10 does apply to two of the tumors, the ductal and lobular tumors, so they are a single primary.
Rule M11 does not apply, there are no multiple intraductal or duct carcinomas.

Rule M12 does apply, there are tumors with histology codes that are different among the first three characters. The initial “unofficial” coding of these tumors is 8522/3 and 8211/3.

- The rule directs you to use Table 1 and Table 2 to identify intraductal and duct carcinomas, and tubular carcinoma does not appear in either of these tables.
There are two primary cancers, one with two tumors and one with one tumor.

Refer now to the histology rules to first code the primary with the two tumors.

Since one of these tumors has different histologic types for its invasive, ductal, and in situ, cribriform ductal, components, you want to know how to code this tumor specifically.
Refer to the Single Tumor: “Invasive and In Situ Carcinoma”, Rule H9 tells us to code the invasive histology; the code for this tumor will be 8500/3.

Now referring to the Multiple Tumors Abstracted as a Single Primary module, to code the cancer with ductal and lobular tumors, you know that Rules H20 and H21 do not apply because there is a histology report from the primary site.

Rule H22 does not apply, this is not a case of inflammatory carcinoma.
Rule H23 does not apply, there are at least two histologic types involved.

Rules H24, H25, and H26 do not apply, this is not a case of Paget disease.

Rule H27 does not apply, you have already taken care of the in situ component of one of the tumors, and you are now dealing only with two invasive histologies.

The next rule, H28, does apply, and you code this cancer as 8522/3 because there is a combination of lobular and duct carcinoma.
Finally, turn to the Single Tumor: Invasive Carcinoma Only module to code the remaining cancer.

Again Rules H10 and H11 do not apply, there is pathology from the primary site.

Rule H12 does not apply, there is no non-specific and specific histology statement.
Rule H13 does not apply, this is not a case of inflammatory carcinoma.

Rule H14 does apply, only one histologic type is identified.

This second cancer histology is coded as 8211/3.
Multiple Primary and Histology Coding Rules
The Kidney unit provides rules for working with renal cell or glandular carcinomas of the kidney parenchyma.

The terms and definitions section notes that transitional cell carcinomas usually arise in the renal pelvis and would be coded to renal parenchyma only in the rare instances when pathologically confirmed to have arisen there and not in the pelvis.

Types of renal cell carcinoma are described, including carcinoma of collecting ducts, chromophobe, chromophhilic, and medullary carcinoma of the kidney.
Levels of invasion are identified, and Wilm’s tumor grouped with the kidney cancers.

A table lists specific renal cell types by ICD-O-3 codes, for assistance in applying both the multiple primary and histology rules.

Diagrams of the internal and external structure of the kidney are provided.
Reviewing the multiple primary rules, note the standard:

- M1 and M2 rules, an unknown number of tumors are abstracted as a single primary, and a single tumor is always a single primary.

Referring to the rules for multiple tumors, the first rule is unique to kidney:

- M3, Wilms tumors are always considered a single primary, so bilateral involvement, whether synchronous or metachronous, does not indicate a second primary cancer.
• M4, that tumors in sites with ICD-O-3 topography codes differing among the first three characters are different primaries; the effect of this rule is to distinguish kidney from other primary sites, rather than to distinguish among kidney primaries, as there is only a single primary site code for the kidney parenchyma.

• M5, states that bilateral kidney tumors are multiple primaries; the exceptional case of Wilms tumors has already been dealt with by a prior rule.
The next two rules state the timing for kidney primaries:

- M6, tumors diagnosed more than three years apart are multiple primaries.
- The standard rule, M7, an invasive tumor more than 60 days after an in situ tumor is a new primary.

The following three rules are related to histology:

- M8, tumors with specific renal cell types are multiple primaries.
- M9, tumors with a non-specific diagnosis and a more specific diagnosis are single primaries.
- M10, tumors with ICD-O-3 histology codes that differ among the first three characters are multiple primaries.
Rules M8 and M9, both referring to the table of renal cell types, differ in that M8 addresses the situation where specific renal cell types are identified, and M9 the situation where renal cell NOS and another specific renal cell type are identified.

The last multiple primary rule,

- M11, is the standard rule, if rules to this point have not provided a decision, the case is abstracted as a single primary.

A case example covered by this rule would be the involvement of multiple tumors in one kidney, all with the same histology.
The structure of the histology rules for Kidney parallels that for Lung.

The rules are the same for both single tumors abstracted as a single primary and multiple tumors abstracted as a single primary, with the exception of the addition of a combination histology rule for single kidney tumors.
• H1 and H8, code the histology documented by the physician when no pathology or cytology report available or obtained.
• H2 and H9, code the histology or cytology from a metastatic site when no specimen from the primary site.
• H3 and H10, code the histology of a single histologic type.
• H4 and H11, code the invasive histology when a single tumor has both invasive and in situ components or the most invasive histology when two or more invasive tumors.
• H5 and H12, code the most specific histologic term when non-specific and more specific histologies are stated.
The non-specific histologies for kidney are listed as:
- cancer/malignant neoplasm and a more specific histology
- carcinoma and a more specific carcinoma
- adenocarcinoma and a more specific adenocarcinoma
- renal cell carcinoma and a more specific renal cell carcinoma
- sarcoma

The next rule within the Single Tumor module,
- H6, refers to using the combination code for mixed adenocarcinoma, 8255/3, when two or more specific renal cell histologies are identified within a single tumor.

The final rule for both single and multiple tumor coding,
- H7 and H13, is to code the histology with the numerically higher ICD-O-3 code.
The patient presents with left flank pain.
On workup, two masses are noted in the left kidney parenchyma, a large 7 cm x 8 cm mass in the upper pole of the kidney and a small 2 cm x 3 cm mass in the lower pole.
Renal cell carcinoma is suspected.
The patient is taken to surgery, and the final diagnosis is renal cell carcinoma clear cell type, 7.5 x 8 cm, and a second renal cell carcinoma with sarcomatoid features, 2 x 3 cm.
Two tumors are identified.
Referring to the Multiple Tumors module for kidney, Rule M3 does not apply, this is not a Wilms tumor.

Rule M4 does not apply, the site for both tumors is C649, kidney.

Rule M5 does not apply, the tumors involve the same kidney.
Rules M6 and M7 do not apply, the tumors are diagnosed at the same time and neither one is an in situ lesion.

Rule M8 does apply, there are two tumors, each with a diagnosis of a specific renal cell type, according to the table.

These are multiple primaries.
Each primary has one tumor. Refer to the Single Tumor module for each primary cancer to determine the correct histology code.

Rules H1 and H2 do not apply to either cancer since there is pathology from the primary site.

Rule H3 does not apply for either cancer since more than one histologic type is identified in each case.

Rule H4 does not apply since there is no indication of an in situ component in the pathologic diagnosis.
Rule H5 does apply to each case, since each diagnosis is stated as renal cell carcinoma with a specific type of renal cell carcinoma.

According to Note 2 for this rule, specific types for invasive cancers can be identified with the words “type” and “with features of.”

So the histology for the larger tumor will be coded as 8310/3, clear cell carcinoma, and the histology for the smaller tumor will be coded as 8318/3, renal cell carcinoma, sarcomatoid.
Tumors of the renal pelvis, ureter, bladder, and urethra/prostatic urethra are included in the Urinary rules.

These organs are lined by a transitional cell epithelium or urothelium where most cancers arise, often in a multifocal pattern.

The terms and definitions section discusses this phenomenon, which may be related to a field effect involving the entire urothelium or an implantation effect where tumor cells are carried in the urinary stream from one site to another.
This multifocality has been interpreted in a unique multiple primary rule for these sites, which considers multiple noncontiguous tumors arising in more than one of these sites as a single primary cancer.

Transitional cell and papillary transitional histologies are described, and the levels of invasion are listed for the bladder and other urinary sites.

The notes indicate that adenocarcinoma of the prostatic urethra is usually an extension from the prostate rather than primary in the urethra.
A table lists the types of urothelial/transitional cell carcinomas, referred to by both the multiple primary and histology coding rules, and anatomic diagrams of the urinary structures include a graphic illustration of levels of tumor invasion through the bladder wall.

The standard M1 and M2 rules are included for Urinary sites, an unknown number of tumors are abstracted as a single primary, and a single tumor is always a single primary.

Most of the multiple primary rules for multiple tumors are unique to the Urinary sites.
The first rule,

- M3, states that if no other urinary sites are involved, tumors in the right renal pelvis and left renal pelvis are multiple primaries.

The next rule,

- M4, is similar, if no other urinary sites are involved, tumors in the right ureter and left ureter are multiple primaries.

The next rule,

- M5, is standard, an invasive tumor more than 60 days after an in situ tumor is a new primary.
• M6, bladder tumors with any combination of papillary, transitional cell, or papillary transitional cell carcinoma are a single primary. This is followed by a unique rule for bladder tumors:

Rule M5 directs the abstracting of an invasive bladder tumor after an in situ tumor as a new primary cancer; however, once there is an invasive bladder cancer with a urothelial or transitional cell histology, all future bladder tumors with a urothelial or transitional cell histology are considered as recurrences of the invasive cancer and are not abstracted as new primaries.

Note that this rule does not apply if another histology is diagnosed within the bladder, such as small cell carcinoma or adenocarcinoma.
• M7, tumors diagnosed more than three years apart are multiple primaries.
• This timing rule therefore does not apply to the invasive bladder cancers, as the general rule requires that you stop when you reach the first rule that applies to the case situation.

• M8, implements the coding for the multifocality of urothelium involvement, stating that urothelial tumors in two or more of the urinary sites are a single primary.
• Again this rule follows the timing rule, so that multifocal involvement of these sites must occur within the three-year time period to be considered as a single primary.
The final three rules for the urinary sites are standard rules:

- M9, tumors with ICD-O-3 histology codes differing within the first three characters are multiple primary cancers.
- M10, tumors in sites with topography codes differing within the first three characters are multiple primary cancers.
- M11, tumors not meeting any prior criteria are single primary cancers.

Rule M8 links the urothelial sites, so rule M10 serves to distinguish these sites from other sites outside the urothelial organs.
The histology modules for the Urinary sites also contain site-specific rules to guide coding for the transitional cell histologies.

The first two rules are the standard

- H1 and H2 rules, code the histology documented by the physician when a pathology report is not available or specimen not taken, and code the cytology/histology from a metastatic site when there is no specimen from the primary site.
The next rule in both single tumor and multiple tumor modules,

- H3 and H11, is site-specific, code 8120 for transitional cell when there is pure transitional cell or transitional cell with a type of differentiation as listed in the rule and in the table.
- This rule thus varies from rules for other sites, in that the histology code for transitional cell is used in preference to certain delineated subtypes.

The following rule again is site-specific,

- H4 and H12, code 8130 when papillary transitional cell is stated in the diagnosis, including the combination of transitional cell and papillary transitional cell.
The following rules are standard:

- H5 and H13, code the histology when only one histologic type is identified.
- H6 and H14, code the invasive histology when a single tumor contains both in situ and invasive components or the most invasive histology when two tumors are abstracted as a single primary.
- H7, code the most specific histologic term (for single tumors only).
- H8 and H15, code the histology with the numerically higher ICD-O-3 code.

The specific histology list for the urinary sites includes cancer/malignant neoplasm and a more specific histology, carcinoma and a more specific carcinoma, and sarcoma and a more specific sarcoma.
Again, the more specific histology rule is not contained within the multiple tumors module for the urinary sites, so for any case circumstances that might possibly be involved, the decision would go to the higher ICD-O-3 code, the last rule in the module.
For our case circumstances

- Patient presents with gross hematuria and a remote history of transitional cell carcinoma of the bladder.
- At cystoscopy the urologist notes a papillary appearing tumor and another malignant-appearing lesion on the bladder wall.
- The patient undergoes transurethral resection of the bladder, and the final diagnosis is non-invasive papillary urothelial carcinoma and a second squamous cell carcinoma invading into the muscularis of the bladder wall.
- The patient undergoes cystoprostatectomy with diagnosis of squamous cell carcinoma extending into the perivesical tissue and adenocarcinoma of the prostate.
- Applying the rules, there are three tumors in the bladder, with the non-invasive papillary urothelial carcinoma removed at the TURB procedure.
Referring to the Multiple Tumors module to determine number of primary cancers, Rules M3 and M4 do not apply because renal pelvis and ureter are not involved.

Rule M5 does not apply, this is not a case of invasive tumor following an in situ tumor within 60 days.

Rule M6 does apply; there is a papillary transitional cell carcinoma of the bladder with a prior history of transitional cell carcinoma of the bladder, so this histology does not represent a new primary cancer but rather a recurrence of a previous cancer.
Proceeding on to a decision on the remaining two tumors, Rule M7 does not apply, there is no indication of a prior diagnosis of either the squamous cell carcinoma of the bladder or the adenocarcinoma of the prostate.

Rule M8 does not apply, the bladder is the only urothelial organ involved in this case.

Rule M9 does apply, there are separate histologies with codes differing among the first three ICD-O-3 characters, 80703 and 81403; there are multiple primary cancers, you can stop with this rule.
Turning to the histology rules, there are two tumors and two cancers. However, the site for one of these cancers belongs in another section of the rules, so only review the bladder cancer using the urinary rules. At this point there is one cancer and one tumor to review, therefore go to the Single Tumor module for histology.
Rules H1 and H2 do not apply, there is pathology from the primary site.

Rule H3 does not apply, the tumor does not contain transitional cell carcinoma.

Rule H4 does not apply, the tumor does not contain papillary transitional cell carcinoma.
Rule H5 does apply, code the histology when only one histologic type is identified.

The histology code for this tumor is 8070/3, squamous cell carcinoma.

To complete the case, we will visit the rules for other sites to determine number of primary cancers and histology for the prostate adenocarcinoma.
Multiple Primary and Histology Coding Rules

MALIGNANT CENTRAL NERVOUS SYSTEM (CNS) TUMORS
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
- oligodendrogial tumors
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

Malignant peripheral nerve sheath tumor (9540)
Malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation (MPNST) (9561)
Neurilemoma, malignant (9560)
Perineurioma, malignant (9571)

Germ Cell Tumors

Choriocarcinoma (9100)
Embryonal carcinoma (9070)
Germinoma (9064)
Immature teratoma (9060)
Mixed germ cell tumor (9085)
Teratoma with malignant transformation (9084)
Yolk sac tumor (9071)

Meningioma, malignant

Meningeal sarcomatosis (9539)
Papillary meningioma, rhabdoid 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, though laterality is included as a rule for benign 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 Abducted 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.

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

• 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.

• H3, code 9382/3, 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.
Case example

- 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.
Multiple Primary and Histology Coding Rules

BENIGN - BORDERLINE 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.
• 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.

• 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.

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

- 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.

- 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 “9530/1.”
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 example

- 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.
Multiple Primary and Histology Coding Rules

OTHER SITES
As stated in the first sentence of the terms and definitions for the Other Sites rules, these rules cover rectosigmoid, rectum, and all sites not included in the site-specific rules.

The gynecologic malignancies represent a large group in this section, and site-specific rules for this group may be forthcoming at some time.

Two histology notes are presented, relating to acinar adenocarcinoma of prostate, which will be coded as adenocarcinoma, and adenoacanthoma, which is identified as adenocarcinoma with squamous metaplasia.
The uterine adnexa are identified as the appendages of the uterus, including ovaries, fallopian tubes, and ligaments holding the uterus in place.

There are many paired organs included in this group, and they are listed in Table 1; these are sites for which laterality must be considered when applying the rules.

Table 2 presents combination codes for many histologies which may be encountered in coding these sites, including combinations for squamous cell, adenocarcinoma, thyroid malignancies, gynecologic malignancies, and germ cell tumors.
The Multiple Primary rules contain the three standard modules, Unknown if Single or Multiple Tumors, Single Tumor, and Multiple Tumors.

The first two modules contain the standard rules:

- M1, if it is not possible to determine single or multiple tumors, the case is abstracted as a single primary.
- M2, a single tumor is always a single primary.
As expected, the Multiple Tumors module contains a number of site-specific rules relating to the individual sites that have been grouped together in this unit, including the first five rules in the module.

- Rule M3, the first rule, states that adenocarcinoma of the prostate is always a single primary; Note 3 for this rule explicitly links the diagnosis of “adenocarcinoma” to any previous diagnosis of “acinar adenocarcinoma” which may have been coded, “acinar” no longer being coded for this diagnosis using the histology rules.
- Rule M4 states that retinoblastoma is always a single primary, so a retinoblastoma appearing at any time in the contralateral eye after the initial diagnosis will be coded as another tumor for the same primary rather than as a new primary.
- Rule M5 states that Kaposi sarcoma is always a single primary; again any tumor appearing after the first diagnosis will be treated as a recurrence rather than a new primary.
- Rules M6 and M7 add a timing element to their statements.
  - With Rule M6, follicular and papillary tumors in the thyroid diagnosed within 60 days are a single primary.
  - Rule M7 states that bilateral epithelial tumors of the ovary diagnosed within 60 days are a single primary.
The follicular and papillary tumors of the thyroid could be diagnosed together in one lesion, or separately in multiple lesions. The range of codes for epithelial tumors of the ovary is specified as 8000-8799, so that bilateral dysgerminomas of the ovary for example, with a code outside this range, would not fall under this rule and would be considered separate primaries.

- Rule M8 refers to the table of paired organs, bilateral tumors in the listed sites being considered as multiple primaries.
- Rule M9 is another site-specific rule relating to familial polyposis, which is always a single primary when there are one or more malignant polyps; the rule does extend to this diagnosis in the colon as well, so that malignant polyps in the rectum or rectosigmoid would not be a separate primary from malignant polyps found with this disease in other segments of the large bowel.
- Rule M10 is a standard rule, tumors diagnosed more than one year apart are multiple primaries.
Inspection of this rule in regard to Rules M6 and M7 reveals an ambiguity in the current formulations of these rules: How are follicular and papillary tumors in the thyroid, and bilateral epithelial tumors of the ovary, diagnosed between 60 days and one year of each other, to be abstracted?

As the rules currently stand, Rule M18 would direct that such cases, in the thyroid or in the ovaries, should be abstracted as a single primary.

The current rules imply but do not state that they would be separate primaries; this ambiguity may be clarified in a rules revision.
The next rule is standard:

- M11, tumors with ICD-O-3 topography codes differing among the first three characters are separate primaries.

The following rule:

- M12, extends this distinction to the fourth character of the ICD-O-3 topography code for certain sites.

Thus sites with different subsite or fourth characters are separate primaries for anus and anal canal, C21, for bone, joints, and cartilage, C40-C41, for peripheral nerves and autonomic nervous system, C47, for connective subcutaneous and other soft tissues, C49, and for skin, C44.
The next two rules are again related to polyps:

- M13, a frank in situ or malignant adenocarcinoma and an in situ or malignant tumor in a polyp are a single primary.
- M14, multiple in situ and/or malignant polyps are a single primary.

Note that these tumors would be occurring in the same site by ICD-O-3 code, so that a case presenting with a malignant tumor in an endometrial polyp and an invasive carcinoma of the ovary would not be a single primary.
• Rule M15 states that an invasive tumor following an in situ more than 60 days after diagnosis represents a second primary cancer.

• Rule M16 directs us to abstract a non-specific histology and a more specific histology as a single primary; for the Other Sites, the pairings are cancer/malignant neoplasm NOS and a specific histology, carcinoma NOS and a specific carcinoma, squamous cell carcinoma NOS and a specific squamous cell carcinoma, adenocarcinoma NOS and a specific adenocarcinoma, melanoma NOS and a specific melanoma; and sarcoma NOS and a specific sarcoma.

• Rule M17 specifies that tumors with ICD-O-3 histology codes differing among the first three characters are separate primaries.

• Rule M18 is the final default rule, multiple tumors that have not met any previous criteria are abstracted as a single primary cancer.
Turning to the Histology rules for Other Sites, note the modules organized like those for Breast cancers: Single Tumor: In Situ Only, Single Tumor: Invasive and In Situ, Single Tumor: Invasive Only, and Multiple Tumors Abstracted as a Single Primary.

For the sites included in Other Sites, you would probably expect most in situ tumors to be found in the rectum, rectosigmoid, and gynecologic sites, and indeed the site-specific rule in the first module addresses polyps.
As with the Breast in situ module, the first rule:

- is a modified version of the usual H1 rule, code the histology documented by the physician when the pathology or cytology report is not available.

The second standard rule does not appear, but:

- to code the histology from a metastatic site.

An in situ diagnosis must be made from a pathologic specimen, and in situ cancers by definition do not have metastases.
H3, relates to polyp coding, and is similar to colon rules: abstract the case using a polyp histology code when there is any reference to polyp involvement, including information in a section of the pathology report other than the final diagnosis, or in another section of the medical record.

This rule does not state that it is limited to tumors in the rectosigmoid or rectum, so would also apply to polyps found elsewhere, such as in another digestive organ or in the uterus.
The next rule,

- H4, is a standard rule, code the most specific histologic term when the diagnosis contains a non-specific and a single specific term; the pairings here, all in situ, are carcinoma NOS and a specific carcinoma, squamous cell carcinoma NOS and a specific squamous cell, adenocarcinoma NOS and a specific adenocarcinoma, and melanoma NOS and a specific melanoma.

The next rule,

- H5 refers to the table of combination codes, which should be used if there are multiple specific histologies, or in contrast to Rule H4 a non-specific code with multiple specific histologies.
The list of terms to identify subtypes includes “pattern and architecture”, as we are dealing here with in situ carcinomas only.

Next,

- Rule H6 is the final rule in this module, code the histology with the numerically higher ICD-O-3 code.
The Single Tumor: Invasive and In Situ module contains only one rule,

- H7, code the single invasive histology.
- This rule embodies the general concept in the MP/H system that invasive histologies are coded over in situ histologies.

If there is a single histologic statement for the invasive component, Rule H7 would suffice for correct coding.
If there is more than one histologic statement for the invasive part of the single tumor, the Single Tumor: Invasive Only module must be consulted to determine the correct coding for the invasive histology.

The structures of the rules for the Single Tumor: Invasive Only and Multiple Tumors Abstracted as a Single Primary modules are similar, though the multiple tumors module contains additional rules for coding intraepithelial neoplasms and Paget disease, and the thyroid rules appear in different places in the two modules.
The first three rules for the two modules are the same:

- H8 and H18, the standard rules for coding the histology documented by the physician when the pathology/cytology report is not available or a specimen was not taken.
- H9 and H19, code the histology from a metastatic site when there is pathology/cytology from a metastatic specimen but no specimen from the primary site.
- H10 and H20, code adenocarcinoma, 8140, for prostate when the diagnosis is acinar adenocarcinoma of the prostate.

At this point the multiple tumors module inserts two rules for coding intraepithelial neoplasias.
• Rule H21 specifies that 80772 is the correct code for squamous intraepithelial neoplasia in sites such as vulva, vagina, and anus.
• Rule H22 specifies that 81482 is the correct code for glandular intraepithelial neoplasia in sites such as the pancreas, or the prostate if PIN III is picked up as reportable by agreement.
• Rule H22 is a coding rule, rather than a reportability rule.
• Presumably these codes would also be used if these lesions were considered as single tumors, though these rules do not appear in the in situ module for Other Sites.
The next rule in the Single Tumor and Multiple Tumors modules is again the same,

- H11 and H23, the standard rule to code the histology when a single histologic type is identified.

Next,

- H24 is the rule that appears only in the multiple tumor module, to code the histology of the underlying tumor when the diagnosis is extramammary Paget disease with an underlying tumor of the anus, perianal region, or vulva.
- This differs from the rules for this situation in the Breast module, where there are combination codes in ICD-O-3 for Paget disease and underlying intraductal or ductal carcinoma; such codes have not been developed for extramammary Paget disease.
The next rule is again the same for both modules,

- H12 and H25, and the same as the in situ rule H3, directing the use of one of the polyp codes, 8210, 8261, and 8263, when there is documentation of adenocarcinoma in a polyp, in the final diagnosis, in other sections of the pathology report, or in other sections of the medical record.

At this point the order of rules in the single tumor and multiple tumor modules varies.
In the single tumor module,

- Rule H13, to code the more specific histology, appears before the two rules addressing coding of thyroid carcinomas, H14 and H15.

Again the non-specific and specific histologies are the same as listed in the in situ module, cancer/malignant neoplasm NOS and a more specific histology, carcinoma NOS and a more specific carcinoma, squamous cell carcinoma NOS and a more specific squamous cell, adenocarcinoma NOS and a more specific adenocarcinoma, melanoma NOS and a more specific melanoma, and sarcoma NOS and a more specific sarcoma.
• H14 that papillary carcinoma of the thyroid is to be coded as 8260/3, to correct miscoding of this diagnosis to 8050/3.
• H15 that follicular and papillary carcinoma of the thyroid should be coded as papillary carcinoma follicular variant, 8340/3.

The multiple tumors module contains these same rules, but the two thyroid rules appear first,

• H26 and H27.
Another rule is inserted here

- H28, to code the histology of the invasive tumor when there are combinations of invasive and in situ (similar to H7).

Then the non-specific/specific histology rule appears,

- H29.
The difference in ordering of the thyroid histology rules in the Multiple Tumor module may prevent a possible error that could occur using the Single Tumor module rules, if an abstracter were to read the diagnosis of “papillary carcinoma” as indicating a non-specific diagnosis, “carcinoma”, with a more specific type of carcinoma, “papillary”, and stop at Rule H13 without proceeding on to Rule H14.

- H14 does provide the correct coding direction, and may be better placed in the Single Tumor module as well if it were to appear before Rule H13.
The final two rules in the Single Tumor and Multiple Tumor modules are the same.

- H16 and H30 refer to Table 2 with combination codes, stating that the combination code should be used when there are multiple specific histologies or a non-specific histology with multiple specific histologies.
- Note the difference between this rule and that in H13 and H29, which direct coding for a non-specific histology and a single specific histology statement.

The next rules,

- H17 and H31 are the final default rules, like H6 in the in situ module, to code the numerically higher ICD-O-3 code.
- Again this is the final rule, and will only be applied if none of the previous rules have provided the coding answer for the case circumstances.
Case

- The patient notes a testicular mass and is referred to a urologist, who on ultrasound identifies a 3 cm right testicular tumor.
- Right radical orchiectomy is performed, and the final diagnosis is mixed germ cell tumor composed of 40% seminoma, 30% embryonal carcinoma, 15% yolk sac tumor, and 15% choriocarcinoma.
- The patient has a testicular cancer, so the Other Sites rules are used to abstract the case.
- There is a single tumor involved, so the Single Tumor module is consulted to determine the number of primary cancers.
Given Rule M2, a single tumor is always a single primary, this is a single primary cancer.

This is not an in situ tumor, so the Single Tumor: Invasive Only module is consulted to determine the histology.

There is a histology report for the primary site, so neither H8 nor H9 apply.
This is not a prostate cancer, Rule H10 does not apply.

Rule H11 does not apply, there are four histologies noted in the single lesion to be coded.

Rule H12 does not apply, this is not a polyp case.
Rule H13 does not apply, the diagnosis does not identify one of the non-specific histologic types noted for this rule.

Rules H14 and H15 do not apply, this is not a thyroid cancer.

Rule H16 may apply; this directs you to code the appropriate combination/mixed code when there are multiple specific histologies.
Going to the table, note the instruction at the top to “Use the combination codes listed in this table only when the histologies in the tumor match the histologies listed below.”

Comparing the histologies to the table, we see that there is not a match to any rows of the table.

Teratoma and embryonal carcinoma have a combination code; teratoma, seminoma, and yolk sac tumor have a combination code; choriocarcinoma, teratoma, seminoma, and embryonal carcinoma have a combination code.
There is not a combination code for the mix of seminoma, embryonal tumor, yolk sac tumor, and choriocarcinoma.

The next rule, H17, tells you to use the numerically highest ICD-O-3 code.

The possible codes in this case are 9061/3, 9070/3, 9071/3, and 9100/3, so applying this rule, you would code this case as 9100/3, choriocarcinoma.
However, this code would seem to misrepresent the case, and at this point you might want to do some research to see if there have been any questions posed on the IandR or SINQ that might aid in applying the rules to this case.

Note one question and answer on the IandR that shows a more inclusive interpretation of the combination codes, though not a question that contains precisely these germ cell elements.

The best approach is probably to use the best-fitting combination code, 9101/3, which also has the virtue of being the numerically highest ICD-O-3 combination code for germ cell elements, flag the case, and submit a question to IandR or SEER to confirm our coding.