Multiple Primary and Histology Site Specific Coding Rules
OTHER SITES
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
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.

The remaining four rules are standard, seen across most of the site modules.
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,

- 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.
The third in situ rule

• 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.
• 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.

• 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.
• 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.
The thyroid rules specify in

- H14 that papillary carcinoma of the thyroid is to be coded as 82603, to correct miscoding of this diagnosis to 80503.
- H15 that follicular and papillary carcinoma of the thyroid should be coded as papillary carcinoma follicular variant, 83403.

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.
• 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.

• 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.
• 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 90613, 90703, 90713, and 91003, so applying this rule, you would code this case as 91003, 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, 91013, 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.