(Excludes lymphoma and leukemia M9590 - M9993 and Kaposi sarcoma M9140)

#### Introduction

- *Note 1:* The group name "urinary sites" include: Renal pelvis C659; ureter C669; trigone of bladder C670; dome of bladder C671; lateral wall of bladder C672; anterior wall of bladder C673; posterior wall of bladder C674; bladder neck C675; ureteric orifice C676; urachus C677; overlapping lesion of bladder C678; bladder NOS C679; urethra C680; paraurethral gland C681; overlapping lesion of urinary organs C688; and urinary system NOS C689.
- **Note 2:** Tables and rules refer to **ICD-O** rather than **ICD-O-3**. The version is not specified to allow for updates. Use the currently approved version of ICD-O.
- *Note 3:* 2007 MPH Rules and 2018 Solid Tumor Rules are used based on date of diagnosis.
  - Tumors diagnosed 01/01/2007 through 12/31/2017: Use 2007 MPH Rules
  - Tumors diagnosed 01/01/2018 and later: Use 2018 Solid Tumor Rules
  - The original tumor diagnosed before 1/1/2018 and a subsequent tumor diagnosed 1/1/2018 or later in the same primary site: Use the 2018 Solid Tumor Rules.
- **Note 4:** For those sites/histologies which have recognized **biomarkers**, the biomarkers are most frequently used to target treatment. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries and/or histologic type. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

### In US, 90% of bladder tumors are urothelial carcinoma; less than 5% are pure squamous cell carcinoma or pure adenocarcinoma.

Urothelial carcinoma originates in urothelial/transitional cells which line the urethra, bladder, ureters, and renal pelvis and has two major subdivisions: papillary and non-papillary.

- Papillary carcinoma: (commonly in bladder, ureter, or renal pelvis): A warty growth which projects from the wall on a stalk
  - o Non-invasive papillary urothelial carcinoma (occasionally called in situ)
  - o Invasive papillary urothelial carcinoma
- Non-papillary urothelial: originates within the mucosa and does not project from the wall
  - o Non-invasive carcinoma in situ (CIS)
  - o Invasive urothelial carcinoma

*Note:* Both urothelial carcinoma and papillary urothelial carcinoma can be in situ /2 or invasive /3. Code the behavior specified in the pathology report.

### **Multifocal/Multicentric Tumors of Urinary Sites**

Multifocality of urothelial carcinoma is a common finding. The phenomenon of multiple tumors has been theorized as being a result of the field effect.

The field effect concept has two main theories:

- 1. Monoclonal: A single malignant cell spreads throughout the urothelium by:
  - a. Intraluminal spread with secondary implantation in different sites within the urinary tract **OR**
  - b. Intraepithelial migration
- 2. **Oligoclonal**: Multifocal/multicentric tumors develop secondary to a field effect precipitated by carcinogens. The carcinogens cause genetic alterations at different sites within the urinary tract.

Neither theory has been conclusively proven.

Flat/urothelial carcinoma in situ can have a widespread effect as a result of direct spread of neoplastic cells within the epithelium.

The rules for coding histology and defining the number of primaries are an attempt to reconcile these observations in order to provide **incidence** data that are consistent and reproducible.

### **Changes from 2007 MPH Rules**

- 1. 2007 Rules instruct "Code the histology from the most representative specimen." For all sites except breast and CNS, 2018 Rules instruct "Code the most specific histology from biopsy or resection. When there is a discrepancy between the biopsy and resection (two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor)."
- 2. There are no significant changes in histology terms or codes in the 2016 WHO edition.

### **Equivalent or Equal Terms**

These terms can be used interchangeably:

- And; with
  - *Note:* "And" and "with" are used as synonyms when **describing multiple histologies** within a **single tumor**. Urothelial carcinoma **and** small cell neuroendocrine carcinoma is **equivalent** to urothelial carcinoma **with** small cell neuroendocrine carcinoma.
- Flat transitional cell carcinoma; flat urothelial carcinoma; urothelial carcinoma in situ; noninvasive flat carcinoma; in situ transitional cell carcinoma
- Multifocal; multicentric
- Noninvasive may describe either in situ papillary carcinoma or flat urothelial cell carcinoma
- Papillary transitional cell carcinoma; papillary urothelial carcinoma
- Simultaneous; synchronous; existing at the same time; concurrent; prior to first course treatment
- Topography; site code
- Tumor; mass; tumor mass; lesion; neoplasm
  - o The terms tumor, mass, tumor mass, lesion, and neoplasm are **not** used in a standard manner in clinical diagnoses, scans, or consults. **Disregard** the terms **unless** there is a physician's statement that the term is malignant/cancer
  - o These terms are used **ONLY** to determine multiple primaries
  - o **Do not** use these terms for casefinding or for determining reportability
- Type; subtype; variant
- Urothelial carcinoma; transitional cell carcinoma
- Urothelium; epithelium; transitional epithelium

(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

### Terms that are Not Equivalent or Equal

These terms are **not equivalent**. There are no casefinding implications.

- Carcinoma, NOS (8010) and adenocarcinoma, NOS (8140) are not equivalent
- Phenotype is not equivalent to subtype/type/variant
- Noninvasive, papillary urothelial carcinoma, flat urothelial carcinoma are not equivalent

Note: Noninvasive is **not equivalent** to either **papillary** urothelial or **flat** urothelial carcinoma. **Both Ta** and **Tis** tumors are technically noninvasive. Code the histology specified by the pathologist.

Papillary growth pattern is not equivalent to papillary urothelial carcinoma

#### **Instructions for Coding Primary Site**

The following instructions are in priority order.

- 1. Code overlapping lesion of urinary bladder **C678** when:
  - A. A single tumor of any histology overlaps subsites of the bladder
  - B. A single tumor or non-contiguous tumors which are:
    - Urothelial carcinoma in situ 8120/2 AND
    - Involves only bladder and one or both ureters (no other urinary sites involved)

Note: Overlapping non-invasive tumors of the bladder and ureter almost always originate in the bladder. They extend/overlap into the ureter by spreading along the mucosa. It is important to code these primaries to bladder C678, NOT to overlapping lesion of urinary organs C688.

- 2. Code bladder NOS C679 when there are multiple non-contiguous tumors within the bladder AND the subsite/origin is unknown/not documented.
- 3. Code overlapping lesion of urinary organs C688 when a single tumor overlaps two urinary sites and the origin is unknown/not documented.

*Note*: See the following examples of contiguous urinary sites where overlapping tumor could occur:

- Renal pelvis and ureter
- Bladder and urethra

- Bladder and ureter (for all histologies other than in situ urothelial cell)
- 4. Code Urinary System NOS **C689** when there are **multiple non-contiguous tumors** in **multiple organs** within the urinary system. *Note:* The physician subject matter experts (SME) discussed the issue of coding primary site for **multifocal/multicentric** urinary tract carcinoma. Although the SMEs understood and acknowledged the importance of coding a specific primary site, there is **no literature** or **criteria** for **determining** the organ of **origin** for multiple tumors involving multiple urinary sites.

### **Table 1: ICD-O Primary Site Codes**

Use the following table to determine the correct site code.

**Column 1** contains the site term and ICD-O code.

Column 2 contains synonyms for the site code and term in column 1.

Site Term and code	Synonyms
Bladder, anterior wall C673	-
Bladder, dome C671	Roof
	Vault
	Vertex
Bladder, lateral wall C672	Lateral to ureteral orifice
	Left wall
	Right wall
	Sidewall
Bladder neck C675	Internal urethral orifice
	Vesical neck
Bladder NOS C679	Lateral posterior wall (no hyphen)
Bladder, overlapping lesion C678	Fundus
_	Lateral-posterior wall (hyphen)
Bladder, posterior wall C674	-

Site Term and code	Synonyms
Bladder, trigone C670	Base of bladder
	Below interureteric crest
	Below interureteric field
	Below interureteric ridge
	Floor of bladder
Bladder, urachus C677	Mid umbilical ligament
	Urachal remnant
Bladder, ureteric orifice C676	Just above ureteric orifice
Overlapping lesion of urinary organs C688	-
Paraurethral gland C681	-
Renal pelvis C659	Pelvis of kidney
	Pelviureteric junction
	Renal calyces
	Renal calyx
Ureter C669	-
Urethra C680	Cowper gland
	Prostatic utricle
	Urethral gland
Urinary system NOS C689	-

### Table 2: Specific Histologies, NOS, and Subtypes/Variants

Use Table 2 as directed by the **Histology Rules** to assign the more common histology codes for urinary tract neoplasms.

Column 1 contains specific and NOS histology terms.

- Specific histology terms do not have subtypes/variants
- NOS histology terms do have subtypes/variants.

Column 2 contains synonyms for the specific or NOS term. Synonyms have the same histology code as the specific or NOS term. Column 3 contains subtypes/variants of the NOS histology. Subtypes/variants do not have the same histology code as the NOS term.

Column 3 may contain NOS histologies which are part of a bigger histologic group. For example, sarcoma NOS 8800/3 (column 1) is a generic term which encompasses a number of soft tissue tumors, including rhabdomyosarcoma 8900/3 (column 3). Rhabdomyosarcoma is also a NOS because it has a subtype/variant 8910/3. The subtype/variant is indented under the NOS (rhabdomyosarcoma) in column 3. There is also a note in column 1 which calls attention to the fact that rhabdomyosarcoma has a subtype/variant.

When subtypes/variants are indented under a NOS in Column 3, use coding rules for a NOS and a single subtype/variant. For example, rhabdomyosarcoma 8900/3 and embryonal rhabdomyosarcoma 8910/3 are a NOS and a subtype/variant, NOT two different subtypes.

Table begins on next page

(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Adenocarcinoma NOS 8140	Mixed adenocarcinoma Urachal adenocarcinoma	Clear cell carcinoma 8310 Endometrioid carcinoma 8380
Note: Urachal carcinoma NOS is coded 8010/3. Urachal adenocarcinoma is coded 8140/3.		Enteric adenocarcinoma 8144 Mucinous adenocarcinoma 8480
Malignant melanoma 8720/3		
Malignant perivascular epithelioid cell tumor 8714/3	Malignant PEComa	
Sarcoma NOS 8800/3		Angiosarcoma 9120/3 Chondrosarcoma 9220/3
Note: Rhabdomyosarcoma 8900 is a NOS with a subtype/variant of embryonal rhabdomyosarcoma/sarcoma botryoides 8910/3.		Leiomyosarcoma 8890/3 Liposarcoma 8850/3 Malignant peripheral nerve sheath tumor (MPNST) 9540/3 Pleomorphic sarcoma 8802/3 Rhabdomyosarcoma 8900/3 Embryonal rhabdomyosarcoma/sarcoma botryoides 8910/3
Small cell neuroendocrine carcinoma 8041	Neuroendocrine carcinoma SmCC	Large cell neuroendocrine tumor 8013 Well-differentiated neuroendocrine tumor 8240
Squamous cell carcinoma 8070	Pure squamous cell carcinoma SCC	Verrucous carcinoma 8051

Table continues on next page

(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Note 1: Previously called transitional cell carcinoma, a term that is no longer recommended.  Note 2: Micropapillary 8131 is a subtype/variant of papillary urothelial carcinoma 8130. It is an invasive /3 neoplasm with aggressive behavior.  Note 3: The histology term is exactly Plasmacytoid/signet ring cell/diffuse variant. All three terms are used together to indicate a specific variant (coded 8082/3).	Clear cell (glycogen-rich) urothelial carcinoma 8120/3 Infiltrating urothelial carcinoma 8120/3 Infiltrating urothelial carcinoma with divergent differentiation 8120/3 Infiltrating urothelial carcinoma with endodermal sinus lines 8120/3 Infiltrating urothelial carcinoma with glandular differentiation 8120/3 Infiltrating urothelial carcinoma with squamous differentiation 8120/3 Infiltrating urothelial carcinoma with trophoblastic differentiation 8120/3 Lipid-rich urothelial carcinoma 8120/3 Microcystic urothelial carcinoma 8120/3 Nested urothelial carcinoma 8120/3 Plasmacytoid urothelial carcinoma 8120/3 Urothelial carcinoma in situ 8120/2	Giant cell urothelial carcinoma 8031/3 Lymphoepithelioma-like urothelial carcinoma 8082/3 Plasmacytoid/signet ring cell/diffuse variant (see Note 3) Papillary urothelial (transitional cell) carcinoma in situ 8130/2 invasive 8130/3 Micropapillary urothelial carcinoma 8131/3 Poorly differentiated carcinoma 8020/3 Sarcomatoid urothelial carcinoma 8122/3

(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

### **Table 3: Non-Reportable Urinary Tumors**

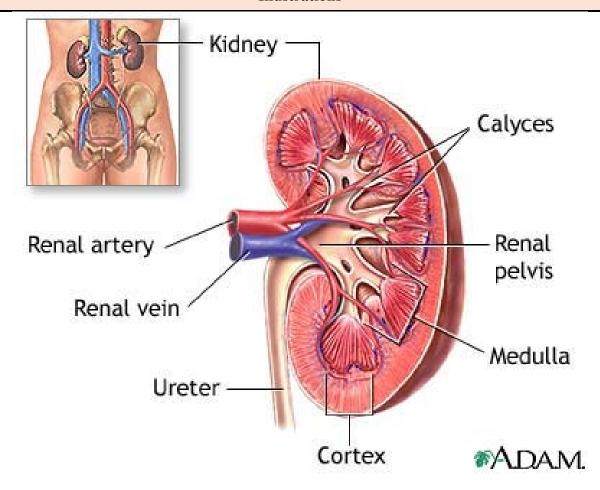
Column 1 contains the terms and codes (if applicable) for the non-reportable histology.

Column 2 contains synonyms of the histology term in column 1. Synonyms have the same code as the term in Column 1.

Histology Term and Code	Synonyms	
Benign perivascular epithelioid cell tumor 8714/0	Benign PEComa	
Granular cell tumor 9580/0		
Hemangioma 9120/0		
Inflammatory myofibroblastic tumor 8825/1		
Inverted urothelial papilloma 8121/0		
Leiomyoma <b>8890/0</b>		
Melanosis No code		
Neurofibroma 9540/0		
Nevus <b>8720/0</b>		
Papillary urothelial neoplasm of low-malignant potential 8130/1		
Paraganglioma <b>8693/1</b>	Extra-adrenal pheochromocytoma	
Solitary fibrous tumor <b>8815</b> /1		
Squamous cell papilloma 8052/0	Keratotic papilloma	
Urothelial dysplasia No code		
Urothelial papilloma 8120/0		
Villous adenoma 8261/0		

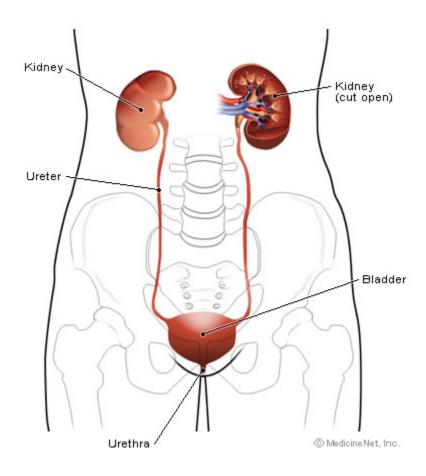
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

#### Illustrations



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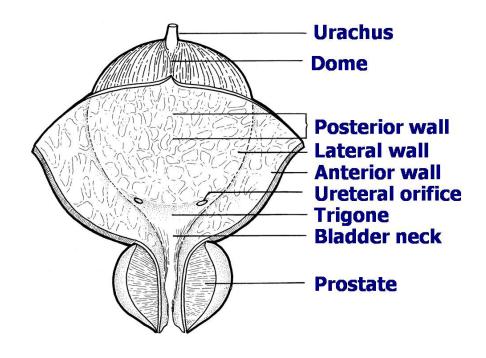
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)



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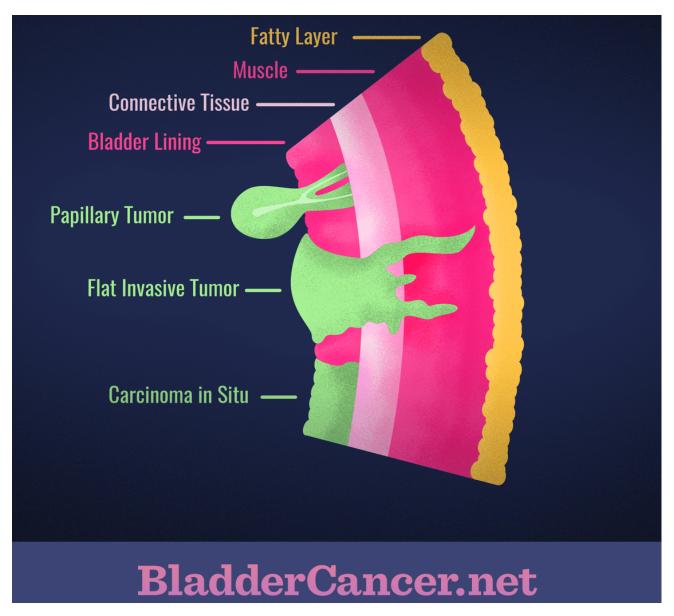
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

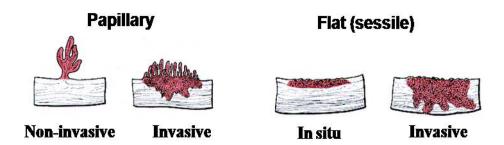


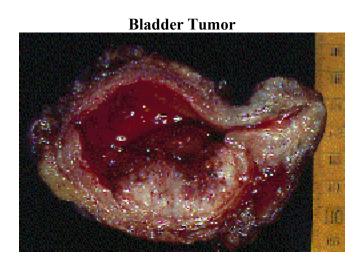
Source: TNM Atlas, 3rd edition, 2nd revision

(Excludes lymphoma and leukemia M9590 - M9993 and Kaposi sarcoma M9140)



(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)



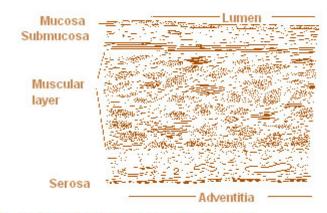


Courtesy Jean-Loup Huret reprinted from Atlas Genet Cytogenet Oncol Haematol. October 2003. van Tilborg A

A, van Rhijn BW. Bladder: Urothelial carcinomas. URL: <a href="http://atlasgeneticsoncology.org/Tumors/bladID5001.html">http://atlasgeneticsoncology.org/Tumors/bladID5001.html</a>, by permission of the Atlas.

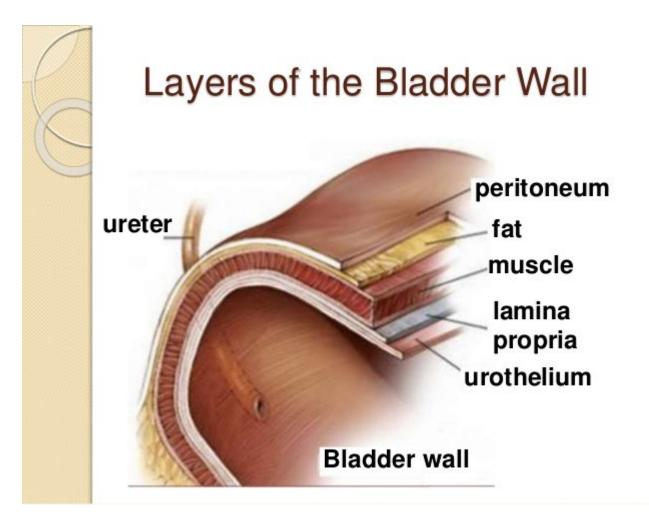
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

### Bladder Wall

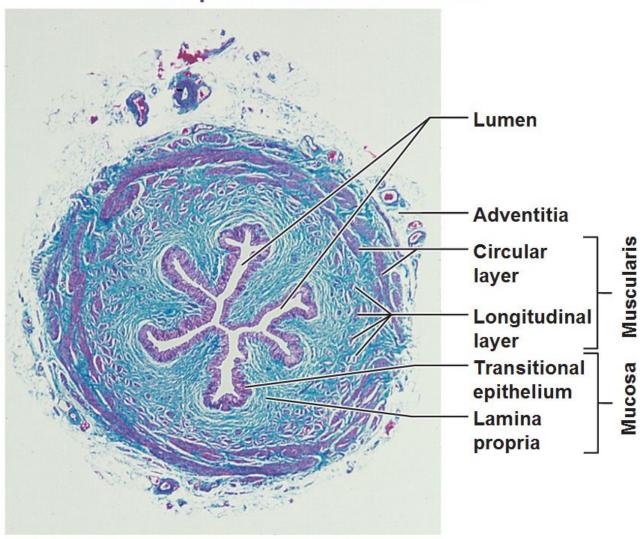


Source: Feneis, Pocket Atlas of Human Anatomy, 2nd ed.

(Excludes lymphoma and leukemia M9590 - M9993 and Kaposi sarcoma M9140)



### Microscopic Structure of the Ureter



- Note 1: These rules are NOT used for tumor(s) described as metastases. Metastatic tumors include but are not limited to:
  - Bones
  - Brain
  - Regional and distant lymph nodes for the primary site being abstracted as identified in Summary Staging Manual
  - Involvement of the pelvic or abdominal wall
  - Liver
  - Lung
- Note 2: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on date of diagnosis.
  - Tumors diagnosed 01/01/2007 through 12/31/2017: Use 2007 MPH Rules
  - Tumors diagnosed 01/01/2018 and later: Use 2018 Solid Tumor Rules
  - The original tumor diagnosed before 1/1/2018 and a subsequent tumor diagnosed 1/1/2018 or later in the same primary site: Use the 2018 Solid Tumor Rules.

### **Unknown if Single or Multiple Tumors**

- Rule M1 Abstract a single primary<sup>i</sup> when it is not possible to determine if there is a single tumor or multiple tumors.
  - Note 1: Use this rule only after all information sources have been exhausted.
  - *Note 2:* Examples of cases with minimal information include:
    - Death certificate only (DCO)
    - Cases for which information is limited to pathology report only
      - o Outpatient biopsy with no follow-up information available
      - o Multiple pathology reports which do not specify whether a single tumor or multiple tumors have been biopsied and/or resected

This is the end of instructions for Unknown if Single or Multiple Tumors.

i Prepare one abstract. Use the histology rules to assign the appropriate histology code.

(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

### **Single Tumor**

#### Rule M2 Abstract a single primary when there is a single tumor.

- *Note 1:* A single tumor is always a single primary.
- *Note 2:* The tumor may overlap onto or extend into adjacent/contiguous site or subsites.
- *Note 3:* The tumor may have in situ and invasive components.
- *Note 4:* The tumor may have two or more histologic components.

#### This is the end of instructions for Single Tumor.

<sup>i</sup> Prepare one abstract. Use the <u>histology rules</u> to assign the appropriate histology code.

### **Multiple Tumors**

- *Note 1:* Multiple tumors may be a single primary or multiple primaries.
- *Note 2:* Separate, non-contiguous tumors are always multiple primaries when:
  - In the urinary system (see <u>Table 1</u>) **AND** in a site other than the urinary system *Example*: Patient has urothelial carcinoma of the bladder and non-metastatic adenocarcinoma of the lung. The lung is not a urinary site. Abstract two primaries.
  - Non-synchronous tumors other than urothelial carcinoma and urothelial carcinoma subtypes in multiple urinary sites (see Rule M14)

### Rule M3 Abstract multiple primaries<sup>ii</sup> when there are:

- Separate/non-contiguous tumors in both the right AND left renal pelvis AND
- No other urinary sites are involved with separate/non-contiguous tumors
- Note 1: Only abstract a single primary when pathology confirms tumor(s) in the contralateral renal pelvis are metastatic.
- **Note 2:** This rule is used <u>only</u> when there is <u>no involvement</u> by separate/non-contiguous tumors in the ureter(s), bladder, or urethra.

(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

### Rule M4 Abstract multiple primaries<sup>ii</sup> when there are:

- Separate/non-contiguous tumors in the right AND left ureter AND
- No other urinary sites are involved with separate/non-contiguous tumors
- Note 1: Only abstract a single primary when pathology confirms tumor(s) in contralateral ureter are metastatic.
- **Note 2:** This rule is used <u>only</u> when there is <u>no involvement</u> by separate/non-contiguous tumors in the renal pelvis, bladder, and urethra.

### Rule M5 Abstract a single primary<sup>i</sup> when synchronous tumors are <u>noninvasive in situ /2</u> urothelial carcinoma (flat tumor) 8120/2 in the following sites:

- Bladder C67 AND
- One or both ureter(s) C669
- Note 1: No other urinary organs are involved.
- **Note 2:** Use this rule **ONLY** for noninvasive in situ urothelial carcinoma (may be called noninvasive urothelial carcinoma or noninvasive flat tumor). For other histologies, continue through the rules.
- *Note 3:* Urothelial carcinoma in situ spreads by intramucosal extension and may involve large areas of mucosal surface. The default for these cases is coding a bladder primary.

### Rule M6 Abstract multiple primaries<sup>ii</sup> when an invasive tumor occurs more than 60 days after an in situ tumor.

- *Note 1:* Abstract both the invasive and in situ tumors.
- Note 2: Abstract as multiple primaries even if physician states the invasive tumor is disease recurrence or progression.
- **Note 3:** This rule is based on long-term epidemiologic studies of recurrence intervals. The specialty medical experts (SMEs) reviewed and approved these rules. Many of the SMEs were also authors, co-authors, or editors of the AJCC Staging

(Excludes lymphoma and leukemia M9590 - M9993 and Kaposi sarcoma M9140)

- Rule M7 Abstract a single primary<sup>i</sup> when the patient has multiple occurrences of /2 urothelial carcinoma in the <u>bladder</u>. Tumors may be any combination of:
  - In situ urothelial carcinoma 8120/2 AND/OR
  - Papillary urothelial carcinoma noninvasive 8130/2 (does not include micropapillary subtype)
  - *Note 1:* Timing is irrelevant. Tumors may be synchronous or non-synchronous.
  - Note 2: Abstract only one /2 urothelial bladder primary per the patient's lifetime.
  - *Note 3:* There are no /2 subtypes for urothelial carcinoma with the exception of papillary urothelial carcinoma.
  - **Example:** On 1/3/2018, the patient had a TURB with a diagnosis of in situ urothelial carcinoma 8120/2. On 5/8/2019, pathology from TURB is papillary urothelial carcinoma non-invasive 8130/2. This is a single primary; the papillary urothelial carcinoma is recorded as a recurrence for those registrars who collect recurrence data.
- Rule M8 Abstract multiple primaries<sup>ii</sup> when the patient has micropapillary urothelial carcinoma 8131/3 of the <u>bladder</u> AND a urothelial carcinoma 8120/3 (including papillary 8130/3) of the <u>bladder</u>.
  - *Note 1:* This is a new rule for 2018.
  - **Note 2:** Micropapillary urothelial cell carcinoma is an extremely aggressive neoplasm. It is important to abstract a new primary to capture the incidence of micropapillary urothelial carcinoma. Micropapillary is excluded from the typical "NOS and subtype/variant" rule (same row in Table 2).
- Rule M9 Abstract a single primary when the patient has multiple <u>invasive</u> urothelial cell carcinomas in the <u>bladder</u>. All tumors are either:
  - Multiple occurrences of urothelial or urothelial subtypes (with exception of micropapillary) **OR**
  - Multiple occurrences of micropapillary
  - *Note 1:* Timing is irrelevant. Tumors may be synchronous or non-synchronous.
  - *Note 2:* Abstract only one /3 invasive urothelial bladder primary **AND** only one micropapillary urothelial 8131/3 bladder primary per the patient's lifetime.
    - An occurrence of micropapillary and an occurrence of urothelial carcinoma would be multiple primaries (see previous rules).

(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

- Rule M10 Abstract multiple primaries<sup>ii</sup> when the patient has a subsequent tumor after being clinically disease-free for greater than three years after the original diagnosis or last recurrence.
  - **Note 1:** This rule **does not apply** when both/all tumors are urothelial carcinoma of the bladder (all subtypes/variants of 8120 except for 8131).
  - *Note 2:* Clinically disease-free means that there was **no evidence** of recurrence on follow-up.
    - Scans are NED
    - Urine cytology is NED
    - Scopes are NED
  - **Note 3:** When there is a recurrence within three years of diagnosis, the "clock" starts over. The time interval is calculated from the date of last recurrence.
  - **Note 4:** When it is **unknown/not documented** whether the patient had a recurrence, default to **date of diagnosis** to compute the time interval.
  - **Note 5:** The physician may state this is a **recurrence**, meaning the patient had a previous urinary site tumor and now has another urinary site tumor. **Follow the rules**; do not attempt to interpret the physician's statement.
  - **Example:** Patient is diagnosed with multifocal/multicentric urothelial carcinomas in the ureter and renal pelvis in January 2018. Both the kidney and ureter are surgically removed. In June 2022 the patient presents with tumor in the contralateral ureter. The physician states this is a recurrence of the original urothelial carcinoma. Code a new primary for the 2022 ureter carcinoma.
- Rule M11 Abstract a single primary<sup>i</sup> when there are urothelial carcinomas in multiple urinary organs.
  - *Note 1:* This rule is **ONLY** for urothelial carcinoma **8120** and all subtypes/variants of urothelial carcinoma. This rule does not apply to any other carcinomas or sarcomas.
  - *Note 2:* Behavior is irrelevant.
  - *Note 3:* This rule applies to multifocal/multicentric carcinoma which involves two or more of the following urinary sites:
    - Renal pelvis
    - Ureter
    - Bladder
    - Urethra

Rule M12 Abstract multiple primaries<sup>ii</sup> when separate/non-contiguous tumors are two or more different subtypes/variants in Column 3 of <u>Table 2</u> in the Equivalent Terms and Definitions. Timing is irrelevant.

*Note:* The tumors may be subtypes/variants of the **same** or **different** NOS histologies.

- Same NOS: Leiomyosarcoma 8890/3 and liposarcoma 8850/3 are both subtypes of sarcoma NOS 8800/3 but are distinctly different histologies. Abstract multiple primaries.
- **Different NOS**: Verrucous carcinoma 8051 is a subtype of squamous cell carcinoma NOS 8070; giant cell urothelial carcinoma 8031 is a subtype of urothelial carcinoma 8120. They are distinctly different histologies. Abstract multiple primaries.
- Rule M13 Abstract multiple primaries<sup>ii</sup> when separate/non-contiguous tumors are on different rows in <u>Table 2</u> in the Equivalent Terms and Definitions. Timing is irrelevant.

*Note:* Each row in the table is a **distinctly different** histology.

**Example:** Small cell neuroendocrine carcinoma 8041 and urothelial carcinoma 8120 are on different rows of Table 2. Abstract two primaries, one for the small cell neuroendocrine carcinoma and a second for the urothelial carcinoma.

- Rule M14 Abstract multiple primaries when the ICD-O site code differs at the second ( $C\underline{X}xx$ ) and/or third ( $Cx\underline{X}x$ ) character.
- Rule M15 Abstract a single primary<sup>i</sup> when synchronous, separate/non-contiguous tumors are on the same row in <u>Table 2</u> in the Equivalent Terms and Definitions.

*Note:* The same row means the tumors are:

- The same histology (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3) **OR**
- A NOS histology in column 3 with an indented subtype/variant
- **Example**: TURBT shows invasive papillary urothelial carcinoma 8130/3 and CIS/in situ urothelial carcinoma 8120/2. Abstract a single primary. Papillary urothelial carcinoma and urothelial carcinoma are on the same row in Table 3.

(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

- Rule M16 Abstract a single primary<sup>i</sup> (the invasive) when an in situ tumor is diagnosed after an invasive tumor AND tumors occur in the same urinary site.
  - *Note 1:* The rules are hierarchical. Only use this rule when previous rules do not apply.
  - **Note 2:** The tumors may be a NOS and a subtype/variant of that NOS. See <u>Table 2</u> in the Equivalent Terms and Definitions for listings of NOS and subtype/variants.
  - **Note 3:** Once the patient has an invasive tumor, the subsequent **in situ** is recorded as a **recurrence** for those registrars who collect recurrence data.
- Rule M17 Abstract a single primary<sup>i</sup> (the invasive) when an invasive tumor is diagnosed less than or equal to 60 days after an in situ tumor AND tumors occur in the same urinary site.
  - *Note 1:* The rules are hierarchical. Only use this rule if none of the previous rules apply.
  - *Note 2:* The tumors may be an NOS and a subtype/variant of that NOS.
  - *Note 3:* When the case has been abstracted, change behavior code on original abstract from /2 to /3. Do not change date of diagnosis.
  - *Note 4:* If the case has already been submitted to the central registry, report all changes.
  - **Note 5:** The physician may stage both tumors because staging and determining multiple primaries are done for different reasons. Staging determines which treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).
  - *Note 6:* See the **COC** and **SEER** manuals for **instructions** on coding **other data items** such as Date of Diagnosis, Accession Year and Sequence Number.
- Rule M18 Abstract a single primary<sup>i</sup> when tumors do not meet any of the above criteria.

**Note:** Use this rule as a last resort. Please confirm that you have not overlooked an applicable rule.

#### This is the end of instructions for Multiple Tumors.

<sup>&</sup>lt;sup>i</sup>Prepare one abstract. Use the histology coding rules to assign the appropriate histology code. For registries collecting recurrence data: When a subsequent tumor is "single primary," record that subsequent tumor as a recurrence.

<sup>&</sup>quot;Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.

### Priority Order for Using Documentation to Identify Histology

#### **IMPORTANT NOTES**

- 1. Code the histology diagnosed prior to neoadjuvant treatment.
  - Note 1: Histology changes do occur following immunotherapy, chemotherapy, hormone, and radiation therapy.
  - Note 2: Neoadjuvant treatment is any tumor-related treatment given prior to surgical removal of the malignancy.

### **Exceptions:**

**Exception:** If the initial diagnosis is based on histology from **FNA**, **smears**, **cytology**, or from a regional or metastatic site, and neoadjuvant treatment is given and followed by resection of primary site which identifies a different or specific histology, code the histology from the primary site.

2. Code the histology using the following priority list and the Histology Rules. Do not change histology in order to make the case applicable for staging.

The priority list is used for single primaries (including multiple tumors abstracted as a single primary)

Code the **most specific** pathology/tissue from either **resection** or **biopsy**.

- *Note 1:* The term "most specific" usually refers to a subtype/variant.
- Note 2: The histology rules instruct to code the invasive histology when there are in situ and invasive components in a single tumor.
- **Note 3:** When there is a discrepancy between the biopsy and resection (two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor).

Use documentation in the following priority order to identify the histology type(s):

- 1. Tissue or pathology report from primary site (in priority order)
  - A. Addendum(s) and/or comment(s)
  - B. Final diagnosis / synoptic report as required by CAP
  - C. CAP protocol
  - **Note 1:** Addendums and comments on the pathology report are given a high priority because they often contain information about molecular testing, genetic testing, and/or special stains which give a more specific diagnosis.
  - *Note 2:* The pathologist's diagnosis from the pathology report is always reliable, so the final diagnosis is the second priority. The final diagnosis is often the synoptic CAP report.

*Note 3:* The CAP protocol is a checklist which:

- Provides guidelines for collecting the essential data elements for complete reporting of malignant tumors and optimal patient care
- Allows physicians to check multiple histologies
- 2. Cytology (usually urine)
- 3. Tissue/pathology from a metastatic site
  - *Note 1:* Code the behavior /3.
  - **Note 2:** The **tissue** from a **metastatic** site often shows **variations** from the primary tumor. When it is the only tissue available, it is **more** accurate than a **scan and only physician documentation**.
- 4. Code the histology **documented** by the physician when none of the above are available. Use the documentation in the following **priority order:** 
  - A. Treatment Plan
  - B. Documentation from Tumor Board
  - C. Documentation in the medical record that refers to original pathology, cytology, or scan(s)
  - D. Physician's reference to type of cancer (histology) in the medical record
  - *Note 1:* Code the specific histology when documented.
  - *Note 2:* Code the histology to 8000 (cancer/malignant neoplasm NOS) or as stated by the physician when nothing more specific is documented.
- 5. Scans: CT, MRI. There is no priority order because scans are not a very reliable method for identifying specific histology(ies) for these sites.

**Note**: Only code differentiation or features when there is a <u>specific code</u> for the NOS with differentiation or the NOS with features in <u>Table 2</u> or the ICD-O and all updates. This instruction applies to single and multiple histologies.

#### **Coding Histology**

- Note 1: The priority is to code the most specific histology. DO NOT USE BREAST HISTOLOGY CODING RULES FOR THIS SITE.
- *Note 2:* Only use this section for one or more histologies within a single tumor.
- *Note 3:* Do not use this section in place of the Histology Rules.
- 1. Code the most specific histology or subtype/variant, regardless of whether it is described as:
  - A. The majority or predominant part of tumor
  - B. The minority of tumor
  - C. A component
  - **Example 1:** Diagnosis for a single tumor is adenocarcinoma 8140 with the majority or predominant part of tumor being endometrioid carcinoma 8380. Code the subtype/variant: endometrioid carcinoma 8380.
  - **Example 2:** Diagnosis for a single tumor is small cell neuroendocrine carcinoma 8041 with minority of tumor being large cell neuroendocrine tumor 8013. Code the subtype/variant: large cell neuroendocrine tumor 8013.
  - *Example 3:* Diagnosis for a single tumor is sarcoma NOS 8800/3 with a component of leiomyosarcoma 8890/3. Code the subtype/variant: leiomyosarcoma 8890/3.
  - Note 1: The terms above (A, B, C) must describe a <u>carcinoma</u> or <u>sarcoma</u> in order to code a histology described by those terms. *Example:* When the diagnosis is adenocarcinoma with a clear cell <u>carcinoma</u> component, code clear cell carcinoma 8310.
    - **Negative Example:** When the diagnosis is simply adenocarcinoma with a clear cell component, code adenocarcinoma NOS 8140. Do not assume this is a clear cell carcinoma. This could be clear cell differentiation or features.
  - *Note 2:* When the most specific histology is described as differentiation or features, see #2.

2.	Code tl	he histology described as differ	rentiation or features/features of ONLY when there is a specific ICD-O code for the "NOS
	with	features" or "NOS with	differentiation".
	Note:	Do not code differentiation or	features when there is no specific ICD-O code.

- 3. Code the specific histology described by ambiguous terminology (list follows) ONLY when A or B is true:
  - A. The only diagnosis available is one histology term described by ambiguous terminology
    - CoC and SEER require reporting of cases diagnosed only by ambiguous terminology
    - Case is accessioned (added to your database) based on ambiguous terminology and no other histology information is available/documented
    - **Example:** Outpatient biopsy says probably papillary urothelial cell carcinoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology papillary urothelial cell carcinoma. The case meets the criteria in #3A.
  - B. There is a NOS histology and a more specific (subtype/variant) described by ambiguous terminology
    - Specific histology is clinically confirmed by a physician (attending, pathologist, oncologist, etc.) **OR**
    - Patient is receiving treatment based on the specific histology described by ambiguous term
    - **Example 1:** The pathology diagnosis is sarcoma NOS consistent with leiomyosarcoma. The oncology consult says the patient has leiomyosarcoma of the bladder. This is clinical confirmation of the diagnosis, code leiomyosarcoma. The case meets the criteria in **bullet 1**.
    - Example 2: The pathology diagnosis is adenocarcinoma consistent with mucinous adenocarcinoma. The treatment plan says the patient will receive the following treatment for mucinous adenocarcinoma. Treatment plan confirms mucinous adenocarcinoma; code mucinous adenocarcinoma. The case meets the criteria in **bullet 2**.

If the specific histology does not meet the criteria in #3B, then code the NOS histology.

### List of Ambiguous Terminology

Apparently
Appears
Presumed
Comparable with
Compatible with
Consistent with
Favor(s)

Most likely
Presumed
Suspect(ed)
Suspect(ed)
Suspicious (for)
Typical (of)

Malignant appearing

- 4. **<u>DO NOT CODE</u>** histology when described as:
  - Architecture
  - Foci; focus; focal
  - Growth pattern
  - Pattern

(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

### **Single Tumor**

### **Rule H1** Code the histology when only **one histology** is present.

- **Note 1:** Use <u>Table 2</u> to code histology. New codes, terms, and synonyms are included in **Table 2** and coding errors may occur if the table is not used.
- *Note 2:* When the histology is **not listed** in **Table 2**, use the **ICD-O** and all **updates**.
- *Note 3:* Submit a question to Ask a SEER Registrar when the histology code is not found in Table 2, ICD-O or all updates.
- Note 4: Only code squamous cell carcinoma (8070) when there are no other histologies present (pure squamous cell carcinoma).
- *Note 5:* Only code adenocarcinoma (8140) when there are no other histologies present (pure adenocarcinoma).

### Rule H2 Code the invasive histology when in situ and invasive histologies are present in the same tumor.

### Rule H3 Code the subtype/variant when there is a NOS and a single subtype/variant of that NOS such as the following:

- Adenocarcinoma 8140 and a subtype/variant of adenocarcinoma
- Papillary urothelial carcinoma 8130 and a subtype/variant of papillary urothelial carcinoma
- Rhabdomyosarcoma 8900 and a subtype/variant of rhabdomyosarcoma
- Sarcoma 8800 and a subtype/variant of sarcoma
- Small cell neuroendocrine carcinoma 8041 and a subtype/variant of small cell neuroendocrine carcinoma
- Squamous cell carcinoma 8070 and a subtype/variant of squamous cell carcinoma
- Urothelial carcinoma 8120 and a subtype/variant of urothelial carcinoma

*Note*: Use <u>Table 2</u> to identify NOS histologies and subtypes/variants.

### Rule H4 Code mixed small cell carcinoma 8045 when the final diagnosis is any of the following:

- Small cell neuroendocrine mixed with any other type of carcinoma (does not apply to sarcoma)
- Two or more subtypes/variants of small cell neuroendocrine carcinoma
- Subtype/variant of small cell neuroendocrine mixed with any other carcinoma (does not apply to sarcoma)

**Example:** Diagnosis from TURB is urothelial carcinoma <u>and</u> small cell neuroendocrine carcinoma. Code mixed small cell carcinoma 8045.

#### **Rule H5** Code mixed urothelial carcinoma as follows:

- Code **8120** when urothelial is mixed with:
  - o Adenocarcinoma or adenocarcinoma subtypes
  - o Squamous cell carcinoma or squamous cell carcinoma subtypes
- Code **8130** when papillary urothelial is mixed with:
  - o Adenocarcinoma or adenocarcinoma subtypes
  - o Squamous cell carcinoma or squamous cell carcinoma subtypes
- Code **8131/3** when micropapillary urothelial is mixed with:
  - o Adenocarcinoma or adenocarcinoma subtypes
  - o Squamous cell carcinoma or squamous cell carcinoma subtypes

*Note*: Adenocarcinoma and subtypes/variants as well as squamous cell carcinoma and subtypes/variants are coded ONLY when pure (not mixed with any other histology).

*Example*: Pathology says majority of tumor is squamous cell carcinoma 8070/3 with a minority composed of papillary urothelial cell carcinoma 8130/3. Code the papillary urothelial cell carcinoma 8130/3. The squamous cell carcinoma is not pure and cannot be coded.

This is the end of instructions for Single Tumor.

Code the histology using the rule that fits the case.

### **Multiple Tumors Abstracted as a Single Primary**

#### **Rule H6** Code the histology when only **one** histology is present in **all** tumors.

- Note 1: Use <u>Table 2</u> to code histology. New codes, terms, and synonyms are included in **Table 2** and coding errors may occur if the table is not used.
- *Note 2:* When the histology is **not listed** in **Table 2**, use the **ICD-O** and all **updates**.
- Note 3: Submit a question to Ask a SEER Registrar when the histology code is not found in Table 2, ICD-O or all updates.
- Note 4: Only code squamous cell carcinoma (8070) when there are no other histologies present (pure squamous cell carcinoma).
- *Note 5:* Only code adenocarcinoma (8140) when there are no other histologies present (pure adenocarcinoma).

#### Rule H7 Code the invasive histology when there are invasive and in situ histologies:

- Mixed in each of the tumors **OR**
- In separate tumors (one or more invasive and one or more in situ)

### Rule H8 Code the subtype/variant when all multifocal/multicentric tumors are a NOS and a <u>single</u> subtype/variant of that NOS such as the following:

- Adenocarcinoma 8140 and a subtype/variant of adenocarcinoma
- Papillary urothelial carcinoma 8130 and a subtype/variant of papillary urothelial carcinoma
- Rhabdomyosarcoma 8900 and a subtype/variant of rhabdomyosarcoma
- Sarcoma 8800 and a subtype/variant of sarcoma
- Small cell neuroendocrine carcinoma 8041 and a subtype/variant of small cell neuroendocrine carcinoma
- Squamous cell carcinoma 8070 and a subtype/variant of squamous cell carcinoma
- Urothelial carcinoma 8120 and a subtype/variant of urothelial carcinoma

*Note 1:* Use <u>Table 2</u> to identify NOS histologies and subtypes/variants.

*Note 2:* All tumors may be mixed histologies (NOS and a subtype/variant of that NOS) **OR** one tumor may be a NOS histology and the other tumor a subtype/variant of that NOS.

Rule H9 Code mixed small cell carcinoma 8045 when the final diagnosis for all tumors is any of the following:

- Small cell neuroendocrine mixed with any other type of carcinoma (does not apply to sarcoma)
- Two or more subtypes/variants of small cell neuroendocrine carcinoma
- Subtype/variant of small cell neuroendocrine mixed with any other carcinoma (does not apply to sarcoma)

*Example:* Diagnosis from TURB is urothelial carcinoma <u>and</u> small cell neuroendocrine carcinoma. Code mixed small cell carcinoma 8045.

This is the end of instructions for Multiple Tumors Abstracted as a Single Primary.

Code the histology using the rule that fits the case.