2007 MULTIPLE PRIMARY RULES

Bladder is one of the sites to which the urinary tract site-specific rules apply. Prior to the 2007 rules, the primary sites of the urinary tract cancers were grouped based on their ICD-O first edition codes. In the new rules, the sites are grouped on a functional and histologic basis. The kidneys, which are glandular organs, are physiologically different from the renal pelvis and ureters, which are conduits for the urine created in the kidney. In addition, kidney histologies are very different from those of the lower urinary tract organs. Kidney cancers now have a separate set of multiple primary and histology coding rules.

In contrast, the renal pelvis, ureters, bladder, and urethra, including the prostatic urethra, all have an internal epithelium lined with the same type of cells—transitional cells, now preferably called **urothelium**. The functions of these organs and the types of cancers are similar, and therefore these organs will be considered as a group for the multiple primary and histology coding rules. Although this chapter discusses only bladder cancers, the rules that apply to bladder cancer will be referred to as the urinary tract rules.

The lining of the urinary tract—the urothelium—is exposed to carcinogens from the renal pelvis to the urethra. As a result, the urothelium may start to break down and develop multicentric or multifocal tumors in the different organs. Because of this, special rules had to be developed for counting multiple primaries in the organs of the urinary tract. Two possible causes for this urothelial breakdown have been proposed:

- ***Field effect*** — this theory suggests that carcinogenic changes have altered the epithelium, making it more susceptible than "healthy" tissue to developing malignant tumors. Field effect is also sometimes called 'field defect' or 'regional diathesis.'
- ***Implantation*** — this theory proposes that multiple tumors are the result of tumor cells that have been shed from an original tumor higher in the urinary tract. The tumor cells then attach to the urothelium in a different site and begin to grow there. Implantation is also sometimes called 'seeding.'

Unique Features of Urinary Tract Rules

- These rules apply to tumors of the renal pelvis (C65.9), ureter (C66.9), bladder (C67._), and other urinary sites (C68._) including urethra and prostatic urethra.
- These rules do not apply to kidney, which has separate site-specific rules.
- 11 multiple primary rules
- THREE years between diagnoses
- Histology coding rules in two sections (15 rules total)
  - 8 rules (H1 – H8) in Single Tumor
  - 7 rules (H9 – H15) in Multiple Tumors Abstracted as a Single Primary
- Includes a reference table to show the codes for variants in urothelial cancer terminology

In the multiple primary rules, the renal pelvis, ureter, bladder and urethra are defined as contiguous sites because of their shared urothelial lining. One of the specific rules is that urothelial tumors in multiple contiguous sites are a single primary. However, the tumors do not have to be in adjacent organs or overlapping two organs to be counted as a single primary. For example, if a urothelial tumor is present in the renal pelvis and separate urothelial tumor is present in the bladder, this is considered a single primary by either the field effect or implantation theory. Therefore, only one abstract is prepared. The primary site would be coded to encompass all of the involved organs—urinary system, NOS, C68.9.