There is a new WHO Classification of Tumours of the Urinary System and Male Genital Organs (4th edition), and with a new edition, it’s a great time to review the recent updates in Genitourinary surgical pathology.

Most of the changes are cosmetic with numerous entities renamed. The changes likely to be most critical to your practice are indicated below in orange.

### Group 1: ≤ 6
- Variants of prostatic adenocarcinoma come and go. The oncocytic and lymphoepithelioma-like variants are out, and pleomorphic giant cell and microcystic variants have been added.

### Group 2: 3+4=7
- Intraductal carcinoma is a new diagnostic entity comprising ducts filled with obviously malignant solid or cribriform tumor that has surrounding basal cells. Do not confuse this entity with HGPIN.

### Group 3: 4+3=7
- Carcinoid tumor has been renamed well differentiated neuroendocrine tumor. Small cell carcinoma has been renamed small cell neuroendocrine carcinoma. Large cell neuroendocrine carcinoma is a new entity.

### Group 4: 4+4=8, 3+5=8, 5+3=8
- Carcinoma of the collecting ducts of Bellini has been renamed collecting duct carcinoma.

### Group 5: 4+5=9, 5+4=9, 5+5=10
- Xp11 translocation carcinoma has been renamed MiT family translocation renal cell carcinoma to include not only TFE3 translocation tumors but also TFEB tumors.

### KIDNEY
- Grading for clear cell renal cell carcinoma (RCC) uses the WHO / International Society of Urologic Pathology system based upon nucleolar prominence instead of the very similar Fuhrman grading scheme.
- Multilocular cystic renal cell carcinoma has been renamed multilocular cystic renal neoplasm of low malignant potential.
- Succinate dehydrogenase deficient renal cell carcinoma is a newly added entity. Patients have mutations in an SDH gene, usually SDHB. Pheochromocytoma / paraganglioma also occurs. The renal tumors have solid growth with uniform cells that have eosinophilic vacuolated cytoplasm that can be glassy to granular, with a bubbly appearance. IHC for SDHB is negative while the entrapped mast cells are positive.
- Tubulocystic renal cell car-


**Acquired cystic disease associated renal cell carcinoma** is a new type of RCC which is associated with dialysis treatment. Cells have intracytoplasmic lumina, creating a sieve-like appearance, and calcium oxalate crystals.

- **Clear cell papillary renal cell carcinoma** is the final new entity; it is probably the most significant as it is relatively common. A tubular architecture is present with nuclei that are all aligned with supranuclear or subnuclear vacuoles. Clear cytoplasm mimics clear cell RCC. IHC reveals positivity for CK7 and CAIX, while CD10 and AMACR are focal to negative.

  - The upper size limit for papillary adenoma has been changed from ≤ 0.5 cm to ≤ 1.5 cm.
  - **Cystic nephroma** has been separated into pediatric cystic nephroma, which contains DICER mutations, and adult cystic nephroma, which does not.
  - **Carcinoid** has been renamed **well differentiated neuroendocrine tumor**. Neuroendocrine carcinoma has been split into small cell neuroendocrine carcinoma and large cell neuroendocrine carcinoma.

**TESTIS**

- **Intratubular germ cell neoplasia (unclassified)** has been renamed **germ cell neoplasia in situ (GCNIS)** because the neoplastic cells are located in the spermatogonial niche and are not intratubular proliferations. Space filling proliferations include intratubular seminoma and intratubular embryonal carcinoma.
  - Post-pubertal yolk sac tumor and teratoma are now termed yolk sac tumor, postpubertal-type and teratoma, postpubertal-type.
  - In addition to placental site trophoblastic tumor, true non-choriocarcinomatous trophoblastic tumors now include epithelial trophoblastic tumor and cystic trophoblastic tumor. These lesions are most common in metastatic lesions or post-chemotherapy.
  - Pre-pubertal teratoma, mixed teratoma and yolk sac tumor are now further specified with the suffix **prepubertal-type**. These pre-pubertal tumors do not have chromosome 12p amplification.
  - **Spermatocytic seminoma** was finally renamed, and is now called **spermatocytic tumor**. It has no relationship to seminoma.

**BLADDER (UROTHELIAL TRACT)**

- The term **urothelial hyperplasia** has been replaced by **urothelial proliferation of uncertain malignant potential**. The lesion is the same, with thickened urothelium that shows minimal cytologic atypia, no true papillary fronds, but often has epithelial tenting or undulations. There is usually a history of noninvasive papillary urothelial carcinoma, low grade.
  - The signet ring cell and clear cell variants of adenocarcinoma have been removed.
  - In the bladder, clear cell adenocarcinoma was renamed clear cell carcinoma, although the terminology remains clear cell adenocarcinoma in the prostate.
  - **Carcinoid tumor** has been renamed **well differentiated neuroendocrine tumor**. Small cell carcinoma has been renamed small cell neuroendocrine carcinoma.
  - **Intratubular large cell hyalinizing Sertoli cell neoplasia** has been added as a distinct entity.

**MEET THE AUTHOR**

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