## CHAPTER 35: SYNDROMES

This is a rapid review of syndromes that impact the genitourinary system. See respective chapters for further details on selected syndromes. Many of these syndromes may appear as "distractor" answer choices.

## PRUNE BELLY SYNDROME (EAGLE-BARRETT OR TRIAD SYNDROME)

- Cryptorchidism
- · Abdominal wall musculature defect
- Urinary tract anomalies (e.g. renal dysplasia, hydroureter, dilated prostatic urethra)

## WAGR SYNDROME

- · Defect on WT1 gene (11p13)
- · Wilms' tumor
- Aniridia
- · Genitourinary abnormalities (hypospadias/UDT)
- · Retardation (mental)

## BECKWITH-WIEDEMANN SYNDROME

- Congenital defect on WT2 gene (11p15) resulting in overgrowth disorder
- Macrosomia, macroglossia, hemihyperplasia, omphalocele, visceromegaly, hypoglycemia
- · Increased risk of Wilms tumor, rhabdomyosarcoma, hepatoblastoma
- Approximately 10-15% have medullary sponge kidney