Genitourinary
Congenital Malformations of the Kidneys
• renal agenesis complete absence of the kidneys
  – occurs when ureteric buds fail to develop
  – Or degenerate before they induce the metanephric mesoderm to form nephrons
• congenital polycystic kidneys
  – the failure of the uriniferous tubules to join

Genitourinary
Congenital Malformations of the Kidneys
• double or divided kidney
  – ureteral bud closest to the sinus drains the lower renal pole and enters the bladder at the trigone
  – the ureter that drains the upper pole enters the bladder in a more medial and caudal position (ectopic ureter).
  – Ureterocele

Genitourinary
Congenital Malformations of the Kidneys
• horseshoe kidney
  – forms when the inferior poles of the kidney fuse while they are in the pelvis

Genitourinary
Urinary Bladder
• derived from the hindgut derivative known as the urogenital sinus.
• Exstrophy of the Bladder
  – characterized by the protrusion of the posterior wall of the urinary bladder, which contains the trigone of the bladder and ureteric orifices

Genitourinary
Congenital Malformations of the Kidneys
• defective closure of the inferior part of the anterior abdominal wall during the fourth week of gestation
• no muscle or connective tissue forms in the anterior abdominal wall to cover the urinary bladder
Genitourinary
The Urachus

• Early in development the urinary bladder is continuous with the allantois
• Allantois regresses to become a fibrous cord known as the urachus.

Genitourinary
The Urachus

• Urachal fistula
  – If the lumen of the allantois persists
  • Causes urine to drain from the bladder to the umbilicus

Genitourinary
The Urachus

• Urachal cyst
  – Small part of the lumen of the allantois persists

Genitourinary
The Urachus

• Urachal cyst
  – small part of the lumen of the allantois persists

Genitourinary
Bladder

• Normally visualized in all fetuses
  – Too large evaluated again at the end of the study
  – Normal emptying occurs every 30 minutes

• Failure to observe the bladder may indicate a severe renal abnormality when accompanied by oligohydramnios.
• Bladder wall should be thin in a normal fetus

Genitourinary

• Amniotic fluid is a critical marker in the assessment of renal function.
• Fetal kidneys begin to excrete urine after the 11th week
• Become the major contributor of fetal urine (hence, amniotic fluid volume) after the 14th to 16th weeks of pregnancy.

Renal agenesis

• The virtual absence of the kidneys.
• Kidneys and bladder are not visualized.
• Amniotic fluid is absent or severely decreased.
• In renal agenesis the adrenal glands may be large and may mimic the kidneys.
Renal agenesis

- Severe oligohydramnios after 13 to 15 weeks menstrual age
- Persistent absence of urine in fetal bladder
- Failure to visualize kidneys (use color flow to outline renal arteries)
- Abnormally small thorax.
- Most associated malformations are undetected prenatally because of poor visualization anhydramnios.

Renal Cystic Disease

- Heterogenous group of heritable, developmental, and acquired disorders.
- Potter classification covers most of the renal cystic conditions seen in the prenatal period.

Potter’s Syndrome

- Is characterized by renal agenesis, oligohydramnios, pulmonary hypoplasia, abnormal facies, and malformed hands and feet.
- Potter’s Syndrome Type I: Infantile Polycystic Kidney Disease
  - Progressive renal enlargement
  - Echogenic renal parenchyma
  - Empty bladder and oligohydramnios

Potter’s Syndrome Type II: Multicystic Dysplastic Kidney Disease
- Multiple noncommunicating cysts of variable size
- No distinct renal pelvis
- No distinct renal parenchyma
- Renal size may be normal, hypoplastic, or enlarged
- Severe oligohydramnios if bilateral

Potter’s Syndrome Type III: Adult Dominant Polycystic Kidney Disease
- Large kidneys with hyperechoic parenchyma
- Size may be asymmetric
- Genetic link autosomal-dominant

Potter’s Syndrome Type IV: Obstructive Cystic Dysplasia
- The fetal kidneys appear small and echogenic with cortical peripheral cysts.
  - If bilateral, look for early bladder outlet obstruction (“keyhole bladder”), bilateral hydronephrosis, a thick-walled bladder, and severe oligohydramnios.
  - The renal cortex is dysplastic and replaced with the multiple cortical cysts.
Obstructive Urinary Tract Abnormalities

- Hydronephrosis
  - the most common fetal anomaly
  - occurs when there is an obstruction in the ureter, bladder, or urethra.

Sonographic findings:
- AP renal pelvic diameter greater than 5 to 10 mm
- Rim of renal parenchyma preserved
- Calyceal distention with central pelvis communication

Secondary Obstruction Ureterocele and Ectopic Ureter

- Ureterocele
  - a cystic dilation of the intravesical (bladder) segment of the distal ureter.
- ectopic ureter
  - Ureter that does not insert near the posterolateral angle of the trigone of the bladder.
- Unless hydronephrosis is present, this condition is difficult to diagnose in utero.

Posterior Urethral Valve Obstruction

- occurs only in male fetuses
  - manifested by the presence of a valve(s) in the posterior urethra.
- results in hydronephrosis, hydroureters, or dilation of the bladder and posterior urethra.

Posterior Urethral Valve Obstruction

- Dilated bladder
- Dilated posterior urethra (keyhole appearance)
- Oligohydramnios
- Hydroureters
- Hydronephrosis and dysplasia
- Fetal ascites (some cases)
  - Distention of fetal abdomen (urethral obstruction malformation complex prune-belly syndrome)
  - Male fetus

Prune-Belly Syndrome

- The condition consists of:
  - Cryptorchidism
  - agenesis of abdominal wall muscle,
  - megaureters, and bladder outlet obstruction caused by urethral anomalies such as atresia, stenosis valves, or diverticulum.
Prune-Belly Syndrome

- Undescended testicles
- Large urinary bladder
- Dilated prostatic urethra
- Dilated & tortuous ureters
- Hydronephrotic, dysplastic or Normal, kidneys

Masses in the kidney

- **mesoblastic nephroma.**
  (hamartoma most common renal tumor is a)
- large, single, solid masses originating from the kidney.
- collection of oddly arranged tissue indigenous to the area

Masses in the kidney

- neuroblastoma - adrenal tumor
- Bilateral renal enlargement with calcifications with shadowing was observed

Congenital Malformations of the Genital System

- Hypospadias
  - Incomplete fusion of the urogenital folds cause abnormal openings of the urethra along the ventral aspect of the penis.

Malformations of the Uterus and Vagina

- incomplete fusion of the two paramesonephric ducts (müllerian ducts)
  - various forms of duplication of the uterus and/or vagina may occur.
- uterus didelphys
  - double uterus and double vagina
- bicornuate uterus
  - duplication of the uterus with one vagina also may occur.
- unicornuate uterus
  - single uterine tube and horn - only one paramesonephric duct develops, a () is formed.