Case 1

What is the diagnosis?
1. Multicystic dysplastic disease.
2. Reflux nephropathy
3. Pyelonephritis
4. Autosomal recessive polycystic kidney disease
8 month old with right sided VUR

International Reflux study committee grading system

- I: Reflux into ureter not reaching renal pelvis
- II: Reflux reaching pelvis but not blunting calyces
- III: Mild Calyceal blunting
- IV: Progressive calyceal and ureteral dilation
- V: Very dilated and tortuous collecting system, intrarenal reflux
SFU grading for hydronephrosis

<table>
<thead>
<tr>
<th>Grade</th>
<th>Central renal complex</th>
<th>Renal parenchymal thickness</th>
<th>Ultra sound scan</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>intact</td>
<td>Normal</td>
<td><img src="image1" alt="Image" /></td>
</tr>
<tr>
<td>I</td>
<td>Slight splitting of pelvis</td>
<td>Normal</td>
<td><img src="image2" alt="Image" /></td>
</tr>
<tr>
<td>II</td>
<td>Evident splitting of pelvis and calices</td>
<td>Normal</td>
<td><img src="image3" alt="Image" /></td>
</tr>
<tr>
<td>III</td>
<td>Wide splitting of pelvis and calices</td>
<td>Normal</td>
<td><img src="image4" alt="Image" /></td>
</tr>
<tr>
<td>IV</td>
<td>Further splitting of pelvis and calices</td>
<td>Reduced</td>
<td><img src="image5" alt="Image" /></td>
</tr>
</tbody>
</table>

15 yr old renal Tx pt with h/o fever and abdominal pain
17 yr old with h/o persistent fevers and right flank pain
16 yr old with acute left flank pain
Case 1

What is the diagnosis?
1. Multicystic dysplastic disease.
2. Reflux nephropathy
3. Pyelonephritis
4. Autosomal recessive polycystic kidney disease
Case 2

What is the diagnosis:
1- Chronic kidney disease
2- Multicystic dysplastic kidneys
3- Autosomal recessive polycystic kidney disease
4- Autosomal dominant polycystic kidney disease

Liver US
ARPKD

- Single gene disorder characterized by bilateral, symmetrical cystic renal disease involving distal convoluted tubules and collecting ducts
- Hyperechoic kidneys, poor cortico-medullary differentiation
- Small cysts <1 cm in diameter, diffuse microcysts
ADPKD

- Multiple renal cysts > 1cm in diameter
- Cystic organ involvement: kidneys (100%), liver (50%), pancreas, brain, gonads
- Non-cystic organ involvement: cardiac valves (25%), hernias, colonic diverticula, cerebral aneurysms
- Cysts start appearing within first decade of life, chronic renal insufficiency by 4th decade of life

4 month old with MCDK
MCDK

- Non-functioning kidney, dysplastic tissue
- Multiple cysts ranging in size from 1- >10 cm
- Second most common abdominal mass in neonate
- Involutes with time
- 40% have contralateral renal abnormality:
  - UPJO
  - VUR

Case 2

What is the diagnosis:
1. Chronic kidney disease
2. Multicystic dysplastic kidneys
3. **Autosomal recessive polycystic kidney disease**
4. Autosomal dominant polycystic kidney disease
Case 3

What is the cause of the abnormality?
1- Urethral injury
2- Anterior urethral valves
3- Posterior urethral valves
4- Primary megaureter

Newborn with antenatal diagnosis of bilateral hydronephrosis
Posterior urethral valves

- 3 types
  - I: Most common, anterior fusion of plicae colliculi
  - II: Rarest, longitudinal folds from verumontanum to bladder neck
  - III: Windsock tissue distal to verumontanum

- Distinct caliber change of urethra at level of valves
- VCUG is still “gold standard”
- US: Angling the transducer towards the bladder neck may reveal dilated posterior urethra
- “Key-hole” appearance is classic finding in PUV
4 yr old with h/o unilateral hydroureteronephrosis
Primary megaureter

- Obstructive ureteral dilation above adynamic, normal caliber, short ureteral segment above ureterovesical junction (UVJ)
- Diagnosis best on IVP or MAG3 renal diuretic scan
- Etiology: Unknown, paucity of ganglion cells?, Hypoplasia of muscle fibers?

Case 3

What is the cause of the abnormality?
1- Urethral injury
2- Anterior urethral valves
3- Posterior urethral valves
4- Primary megaureter
What is the diagnosis?
1. Testicular torsion
2. Scrotal cellulitis
3. Trauma
4. Epididymo-orchitis

Epididymo-orchitis

- Infectious inflammation of epididymis, testicle or both
- Classic finding: Enlargement of the testes and/or epididymis with increased blood flow and small reactive hydroceles
- Tx: Antibiotics
- Recurrent episodes may lead to decreased fertility
12 yr old with h/o acute scrotal pain

Testicular Torsion

- Spontaneous or traumatic twist of testes and spermatic cord, resulting in occlusion/infarction
- Usually testes twists medially, and can be manually de-torsed
- Decreased or absent blood flow on color Doppler US
What is the diagnosis?
1- Testicular torsion
2- Scrotal cellulitis
3- Trauma
4- Epididymo-orchitis
Case 5

Most common solid abdominal mass?
1- Neuroblastoma
2- Hepatoblastoma
3-Wilm’s tumor
4- Multilocular cystic nephroma
Wilm’s Tumor
Wilm’s Tumor

Wilm’s tumor
Wilm’s Tumor

- Most common solid abdominal mass and most common renal tumor
- Embryonic tumor with epithelial, blastemal and stromal elements
- Peak incidence 2-3 years, African American at higher risk, 1% familial
- Same incidence as Neuroblastoma

Wilm’s Tumor

- Firm non-tender, painless mass in abdomen
- Hematuria
- Hypertension
Wilm’s Tumor

- Bulky and replaces most of the kidney
- Usually solid with pseudocapsule, calcification rare
- Local and regional extension to lymph nodes and extra-renal space
- Renal vein/IVC, urothelial spread
- Distant mets to lung and liver
Neuroblastoma

- 10% of all childhood cancers
- Most common extra-cranial solid malignancy of childhood
- Second most abdominal cancer in older infant and child after Wilm’s tumor
- Average age of diagnosis is 2 years
- 50 to 60% have metastases at time of diagnosis
Neuroblastoma

- Tumor of primitive neural crest cells (neuroblasts) that may arise anywhere in the sympathetic ganglion chain or adrenal medulla
- Increased secretion of catecholamines and metabolites (HVA, VMA, norepinephrine or dopamine)
- Usually silent till it invades adjacent structures, metastasizes or produce paraneoplastic syndromes (MEI, diarrhea with hypokalemia)
- Mets to skeleton, bone marrow, liver, lymph nodes and skin

Neuroblastoma

- Gross or microscopic Ca+2, well-demarcated but no capsule
- Two-thirds of NB arise in abdomen, of which 2/3 arise in the adrenal gland
- Differences from Wilm’s tumor
Case 5

Most common abdominal solid abdominal mass?
1- Neuroblastoma
2- Rhabdomyosarcoma
3- Wilm's tumor
4- Multilocular cystic nephroma

Case 6

Diagnosis?
1- Sacrococcygeal teratoma
2- Perforated appendicitis with appendicolith
3- Ovarian teratoma
4- Mesenteric cyst
14 yr old with pelvic pain
18 yr old with yolk sac tumor
Case 6

Diagnosis:
1. Sacrococcygeal teratoma
2. Perforated appendicitis with appendicolith
3. Ovarian teratoma
4. Mesenteric cyst

Thank You!

Questions?