Imaging of Congenital Anomaly of the Urinary System

Dr Wan Irnawati Wan Ab Rahman,
Radiologist HQE1
Radiography Update Course 8/6/2015
Outline of Presentation

- Overview: Spectrum of Congenital Anomaly of Kidney and Urinary Tract (CAKUT)
- Diagnostic strategies for diagnosis
CAKUT-Background

• Affect 1:500 newborn
• Can affect kidney alone +/- LUT
• Significant cause of morbidity
• Predispose to infection, development of HPT, CVS disease and ESRF
• Correct dx is essential for appropriate mx
### Overview

#### Kidney anomalies
- Dysgenesis
- Renal agenesis
- Hypoplasia
- Dysplasia
- AbN shape/site
  - Ectopic
  - Fusion
  - Horseshoe
  - Cross-fused

#### Ureteric anomalies
- PUJ obstruction
- Ureterocele
- VUJ obstruction
- Vesico-ureteric reflux (VUR)

#### Vesical & urethral anomalies
- Bladder exsotrophy
- Patent urachus
- PUV
Radiological Examination

Best screening tool for suspected CAKUT

Anatomical abN which helps to determine next step of Ix (IVU, MCU, CT, MRI)

IVU: Functional assessment of hydroureteronephrosis
RENAL ANOMALIES

• Most are asymptomatic
• Incidental findings when we image the abdomen
• Some presented with early renal impairment, young HPT and UTI
Renal Agenesis

Failure of one or both kidney to develop

Bilateral: incompatible with life

Unilateral
1:4000
20-36% genetic
Renal Hypoplasia

Small kidney d/t partial development

Usually unilateral

Small kidney = small arteries → predispose HPT
Multicystic dysplastic kidney (MCDK)

- Formation of multiple, irregular cysts of various sizes replacing normal renal parenchymal.

- Not treatable: require u/s follow up
  - Cyst <5cm: high chance of spontaneous involution
  - If no involution after 2y.o: resection
  - Risk of malignant transformation

- U/S:
  - multiple internal cysts of varying sizes and shapes
  - fibrotic/echogenic parenchymal

- Renal scintigraphy: No excretion
Abnormality in shape & position

• Ectopic kidney
• Fusion anomaly
  – Horseshoe kidney
  – Crossed fused ectopia
Ectopic kidney

• Kidney that is not located in its usual position
• Due to failure to ascend from its origin the pelvis or ascend superiorly to be located in the thorax
• Mostly resides in pelvis – presented with abdominal mass and repeated UTI
Fusion anomaly: Horse shoe

• Most common type of fusion anomaly
• 1:400-500 adults
• LP of the kidney fused in the midline
• Prone to develop recurrent UTI, increase susceptibility to trauma
• Independent risk factor to develop calculi and TCC
Crossed fused ectopia

- Both kidneys are located on the same side with 2 ureters arising from respective kidneys
- The ureter arising from the crossed kidney travels back to the opposite site to inserts in the bladder
Abnormality of collecting system

- Duplex systems
- Hydronephrosis
  - Obstruction of urine flow to the distal urinary tract (PUJ obstruction)
  - Or reflux of urine up into the ureter d/t VUR or urethral obstruction (PUV)
- Bladder extrophy
- Patent urachus
Imaging strategies

U/S: Confirmed dilatation of urinary tract

Performed MCUG to rule out VUR

If VUR excluded: IVU or dynamic scintigraphy to identify obstructed anomaly
Duplex collecting system

• 0.7% of the normal adult population and in 2-4% of patients investigated for urinary tract symptoms
• characterized by an incomplete fusion of upper and lower pole moieties resulting in a variety of complete or incomplete duplications of the collecting system.
• Most are asymptomatic and diagnosed incidentally.
• When symptoms occur (infection, reflux or obstruction) the patient is likely to have completely duplicated ureters.
DUPLEX SYSTEM

• Imaging findings:
  – Duplicated ureters extending a variable distance down to UB
  – Obstruction upper moiety (with ureterocele)
  – VUR of lower moiety
Ureterocele

- Congenital dilatation of intra-mural distal-most portion of ureter
- u/s: cystic structures at VUJ with associated dilated distal ureter
Ureterocele

- 2 types
  - Orthotopic: when they occur in a normally positioned ureteral orifice in a single system (20%)
  - Ectopic ureteroceles occur in the upper pole moiety of a complete ureteral duplication (80%)
- Infection is the most common presentation of ureteroceles in children
- Obstruction of the kidney or upper pole moiety may lead to renal dysplasia or nephropathy.
- VUR is common, occurring in the contralateral ureter in 25% of cases and the lower pole moiety in 50%.
- Reflux into the lower pole ureter is thought to be caused by a shortened intramural tunnel at the ectopic location of the orifice.
- In females, the upper pole insertion may be distal to the external sphincter or in the wall of the vagina resulting in incontinence.
Hydronephrosis

• Dilatation of the renal pelvis (unilateral/bilateral) may be due to
  – Obstruction of urine flow to the distal urinary tract (PUJ obstruction)
  – Or reflux of urine up into the ureter d/t VUR or urethral obstruction (PUV)
PUJ obstruction

• 1:1000-2000 newborns
• Commonly unilateral
• Male predominance
• Cx: recurrent UTI, stone formation
• Causes: ureter fold, compressive aberrant vessels
• Radiological findings:
  – u/s: dilated renal pelvis with collapsed prox ureter
  – IVU: dilatation and baggy appearance of renal pelvis
Ureter fold: radiolucent band
IVU: LEFT PUJ OBSTRUCTION
Vesico-ureteric reflux (VUR)

• Most frequent anomaly
• May be isolated or associated with PUV, duplex system and neurogenic bladder
• Presentation: repeated UTI
• Primary Dx tool: MCUG
• Compression of the tunneled portion of the distal ureter by intraluminal bladder pressure acts as a one-way valve that prevents VUR.
• Failure of maturation of the VUJ or a short submucosal tunnel alters the function of this valve mechanism and allows reflux.
<table>
<thead>
<tr>
<th>Grade</th>
<th>Ureter</th>
<th>VCUG Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Reflux only</td>
<td>Normal</td>
</tr>
<tr>
<td>II</td>
<td>Reflux, no dilatation</td>
<td>Reflux, no blunting</td>
</tr>
<tr>
<td>III</td>
<td>Mild to moderate tortuosity</td>
<td>Mild to moderate dilatation of renal pelvis</td>
</tr>
<tr>
<td>IV</td>
<td>Moderate dilatation</td>
<td>Moderate dilatation, obliteration of the sharp angle of the fornices</td>
</tr>
<tr>
<td>V</td>
<td>Gross dilatation and tortuosity</td>
<td>Gross dilatation of pelvis and calyces, loss of papillary impression</td>
</tr>
</tbody>
</table>
Management of VUR

• the goal is to prevent infection and renal scarring.
• 80% of grade II and 92% of grade I VUR cases have resolved by 5 years after diagnosis and are usually managed conservatively
• indications for surgical intervention includes:
  – breakthrough pyelonephritis despite antibiotics
  – increasing renal scarring
  – persistent high-grade VUR
  – associated UVJ abnormality
Posterior Urethral Valve (PUV)

• Most common obstructive lesion
• Male infant (1:4000-7500 infants)
• Fetal u/s: oligohydromnios, bilateral HN, hypertrophy of UB wall
• MCUG: gold standard
  – Dilatation and elongation of posterior urethra
  – Radiolucent band (occasionally seen)
  – a/w VUR in 50% cases
MCUG: Dilatation and elongation of the posterior urethra
Patent urachus

• Failure of the allantois to obliterate will result in a urachal remnant.
• The urachus may persist
  – patent in its entirety (patent urachus)
  – remain open at the bladder and closed at the umbilicus (diverticulum)
  – patent at the umbilicus and closed at the bladder (sinus)
  – close at both ends with a central patent lumen (cyst)
• The patent urachus is the most common symptomatic variant in children and is associated with posterior urethral valves (PUVs) or urethral atresia in one third of the cases
• Patent urachus is clinically evident early in life as leakage of urine from the umbilicus.
• The remaining remnants are generally asymptomatic unless complicated by infection
Embryology of GUT system

**Fig. 2**—Diagram of lower urinary tract development. By end of seventh week of gestation, cloaca (not shown) is separated into dorsal (D) and ventral (V) parts by urogenital sinus (US), which will develop into perineal body. A = allantois, M = mesonephric duct.

**Fig. 3**—Diagram of bladder development. Urogenital sinus further differentiates into cranial and vesical (brown), middle and pelvic (blue), and caudal and phallic (yellow) divisions. Cranial division forms bladder dome, ventral bladder wall, and urachus (U). Bladder trigone (not shown) is formed by caudal ends of mesonephric ducts distal to ureteral bud (not shown). Urachus involutes and becomes median umbilical ligament by week 16. Developing genitals (G) are normally positioned cephalad to opening of urogenital sinus. US = urogenital sinus.
Patent Urachus

- Urachal infection in children can lead to sepsis and requires antibiotic therapy, drainage, or both followed by excision.
- The most serious complication of a urachal remnant in the adult population is the development of malignancy—most commonly adenocarcinoma.
Thank you!!