Congenital anomalies of the kidney and urinary tract (CAKUT). How to proceed for diagnosis?

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Learning objectives

- Congenital abnormalities of the kidney and urinary tract (CAKUT) occur 1 of 500 newborns
- CAKUT predispose to the development of hypertension, cardiovascular disease and end-stage renal failure
- Quick management++

Learning objectives:
1. To illustrate a wide spectrum of CAKUT.
2. To look for the causes of congenital obstructive uropathy.
3. To describe a strategy for diagnosis of CAKUT with different imaging modalities.

Background

- CAKUT are variable and can affect the kidney(s) alone and/or the lower urinary tract.
- Radiologic investigation is essential for diagnosis. Ultrasonography (US) is ideally suited for the evaluation of children with a suspected urinary tract abnormality.
- Voiding cystourethrography (VCUG) is essential for the evaluation of the anatomy and abnormalities of the bladder and urethra and should determine which additional examinations are required: intravenous urography (IVU), CT scan or MR imaging.
- 99mTc-DTPA dynamic renal scintigraphy is one of the most important diagnostic tools in the evaluation of neonates with hydroureteronephrosis.
- CAKUT: spectrum of diseases:
  1. Kidney anomalies (renal malformations): Renal agenesis (renal aplasia)• Renal hypoplasia• Multicystic dysplastic kidneys (renal dysplasias) (MRDs)• Duplex kidneys
  2. Ureteric anomalies: Ureteropelvic junction obstruction (UPJO)• Ureterovesical junction obstruction• Ureterovesical junction incompetence• Duplications
  3. Vesical anomalies
  4. Urethral anomalies
- US is commonly the initial diagnostic study. If dilatation of the urinary tract is confirmed, VCUG is performed to identify vesicoureteral reflux (VUR) or other causes of upper tract dilatation. If VUR is excluded, nuclear diuresis renography is the primary test for identifying obstructed tract anomalies. IUV or
CT scan can also be used to determine the presence of anatomic abnormality. MR urography can combine superior anatomic and functional information in a single test that does not use ionizing radiation.

**Imaging findings OR Procedure details**

**Developmental anomalies of the kidney**

**Renal agenesis:**
Failure of one or both kidneys to develop
Bilateral: rare, associated with other congenital anomalies, incompatible with life
Unilateral: common (one in 4,000 infants, more boys than girls), asymptomatic;
Other kidney enlarges to compensate#Absent ipsilateral renal artery.

**Renal hypoplasia:**
Small congenital kidney
Complete (global) renal hypoplasia
Segmental renal hypoplasia

**Renal malposition:**
# One or both kidneys
# Ectopic kidney (fig1)
# Crossed renal ectopia (one kidney has crossed to the contralateral side) (Fig2)
# Associated with fusion of kidneys

Horseshoe kidney
Fusion of upper pole
# Abnormalities of rotation are often associated with ectopic kidneys

**Multicystic dysplastic kidney (MCDK)**
Usually happens in only one kidney
• **US**: multiple internal cysts of varying sizes and shapes.

The renal parenchyma is usually fibrous and echogenic

Exclude any communication with the ureter and between cysts++

• **RS**: no excretion

**Duplex collecting system (fig.3):**

Complete or incomplete duplication of the collecting system.

1% of normal population

**Radiographic features:**

• Duplicated ureters extending a variable distance down to the bladder
• Obstruction of the upper pole moiety down to the bladder, often with a ureterocoele (US++)
• Vesico-ureteral reflux into the lower pole moiety
• Ectopic insertion of the upper pole moiety into the prostatic urethra in males or vaginal vault in females

**Shape abnormalities of the kidney**

-Persistent fetal lobulation (fig.4):

Normal variant seen in adult kidneys

-Hypertrophied column of Bertin:

• May be mistaken for a renal mass+++ 
• Usually located in the mid portion of the left kidney
• CT an MRI: Enhance uniformly with renal cortex, and remain isodense to normal parenchyma on delayed images.

-Dromedary hump :

Normal variant of renal contour, caused by the splenic impression onto the superolateral left kidney.

**Developmental anomalies of urinary tract**

1) Pelvi-ureteric junction (PUJ) obstruction

1 per 1000 - 2000 newborns.
Commonly unilateral. Left side. Male predominance.

May present in adult populations.

Recurrent urinary tract infections. Stone formation.

Causes: often unknown. Ureter fold, compressive aberrant vessel.

Associations with others anomalies.

**US** : dilated renal pelvis with a collapsed proximal ureter. Cortical and renal pelvis measurement (fig.5)

**Doppler** : higher RI, lower polar artery.

**IVU** : dilation and "chubby or baggy appearance" of renal pelvis. (fig.6 and 7)

**Scintigraphy** : quantitate the degree of obstruction.

**Uro MRI or CT** : hydronephrosis, crossing vessel.

Particularity: Congenital PUJ caused by urothelial ureteral fold (fig.8 and 9)

2) **Congenital Megaureter**

Obstructed primary megaureter

Refluxing primary megaureter (caused by VUR due to paraureteric diverticulum, ureterocele, duplicated collecting system)

Non-refluxing unobstructed primary megaureter

Association: renal dysplasia, megacalyces

**Radiographic features** (fig.10 and 11):

On all modalities the ureter is enlarged ( >7 mm ).

In obstructed form the distal ureter is most dilated++ (fig12 and 13)

In refluxing primary megaureter, VUR is demonstrated with other abnormalities.

3) **Vesicoureteric reflux (VUR)**

- The most frequent congenital anomalies of urinary tract.
- Urinary tract infections++
- VUR may be an isolated or associated with other congenital anomalies: posterior urethral valves, duplex collecting system, neurogenic bladder…
- **Voiding cystourethrogram (VCUG)**: primary diagnostic procedure +++
• Presence and grade of VUR (fig.14 and 15)
• Micturition or during bladder filling VUR
• Associated anatomical anomalies
  • Ultrasound: Renal parenchyma,
  • Contrast ultrasound: Viewing VUR using micro-bubbles
  • Nuclear cystography:
    • Lower radiation dosage++
    • Lack of spatial resolution
    • Difficult to recognize bladder abnormalities
• MR voiding cystourethrogram: ?

4) Ureterocoele: (fig.17)

Congenital dilatation of the distal-most portion of the ureter

Abnormal vesicoureteric junction: Herniation into the bladder

Simple ureterocoele: with normal VUJ

Ectopic ureterocoele: most frequent, the site of VUJ is abnormal (duplex collating system++) (fig.18).

US: cystic structure into the bladder with associated dilated distal ureter (fig.16)

IVU: Substraction image into the bladder (head cobra sign) (fig.19 and 20)

Everted ureterocoele: appear like bladder diverticulum

5) Posterior urethral valves (PUV's)

The most common congenital obstructive lesion of the urethra.

Male infants.

Fetal US: oligohydroamios, bilateral renal dilation, ureteral dilation, hypertrophy of the bladder, "keyhole sign"

US: thick-walled and trabeculated bladder, dilation of posterior urethral (more than 6mm ++), " keyhole sign"

VCUG: Is the best imaging technique for the diagnosis (fig.21).

  • Micturition phase in a lateral or oblique views++
  • Dilatation and elongation of the posterior urethra
  • The valve: radiolucent band (occasionally seen)
  • VUR: associated in 50% of cases

6) Other, less common:
• Bladder diverticulum (fig.22)
• Neurogenic bladder (fig.23)
• Pre-ureteral vena cava (fig.24 and 25)
• Iliac ureteral diverticulum (fig.26)

Images for this section:

Fig. 1
**Fig. 2:** Crossed renal ectopia: Malrotation and dilatation of the right kidney. Gross dilation and thickened cortex of the crossing left kidney.
Fig. 3: IVU: Complete duplication of the left collecting system with gross ureterocoeola (arrows)
Fig. 13: Anatomopathological aspect of congenital megaureter: narrowed distal ureter
Fig. 12: Congenital megaureter on IVU: narrowed distal ureter
Fig. 11: IVU : Obstructive megaureter with important dilation of distal ureter
**Fig. 10**: US: Obstructive megaureter with important dilation of distal ureter

**Fig. 9**: Anatomopathological aspect
Fig. 8: IVU: ureter fold: radiolucent band
**Fig. 7:** Anatomopathological aspect
Fig. 15: Grade of VUR: From 1 to 5
**Fig. 25:** Pre-ureteral vena cava: the ureter passing posterior to the inferior vena cava causing hydronephrosis

**Fig. 24:** Pre-ureteral vena cava: the ureter passing posterior to the inferior vena cava causing hydronephrosis
Fig. 23
**Fig. 21:** VCUG: Micturition phase. Dilatation and elongation of the posterior urethra.
Fig. 20: CT SCAN : left Ureterocoele
Fig. 19: CT SCAN : left Ureterocoele
non functional upper moiety

functional lower moiety

ureterocoele
**Fig. 18:** IVU: Ureterocoele with duplex collecting systeme

**Fig. 17:** Left Ureterocoele
Fig. 16: Typical aspect on US
Fig. 26: Iliac ureteral diverticulum
Conclusion

RENAL PELVIS DILATION

HOW TO PROCEED? SENN FIG. 27

INFANTS WITH MILD POSTNATAL RENAL DILATATION

HOW TO PROCEED? SENN FIG. 28

CAKUT are common in children. They are a significant cause of morbidity in infancy. A right strategy and a familiarity with urologic anomalies are essential for correct diagnosis and appropriate management.

Images for this section:

Decision algorithms

Grades de dilatation (ESPR–ESUR)
References


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