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11 Tweets • 2021-04-24 22:54:48 UTC • See on Twitter

rattibha.com

1/ Hello #MedTwitter

This month's @ASPNeph Imaging Webinar was about #cystickidneydisease
Here are some nuggets!
#Medtweetorial #nephtwitter #kidneycysts #ciliopathies

Let's start with a poll!

Which of the following is true about kidney cysts?

2/ All the above

If cysts in children are rare, why do we care?!

Cysts can be-

🔸 symptomatic (pain, infection, bleeding, obstruction)

🔹 a marker of underlying disease (PKD)

🔸 can be associated with progressive worsening of kidney function

🔹 can have malignant potential

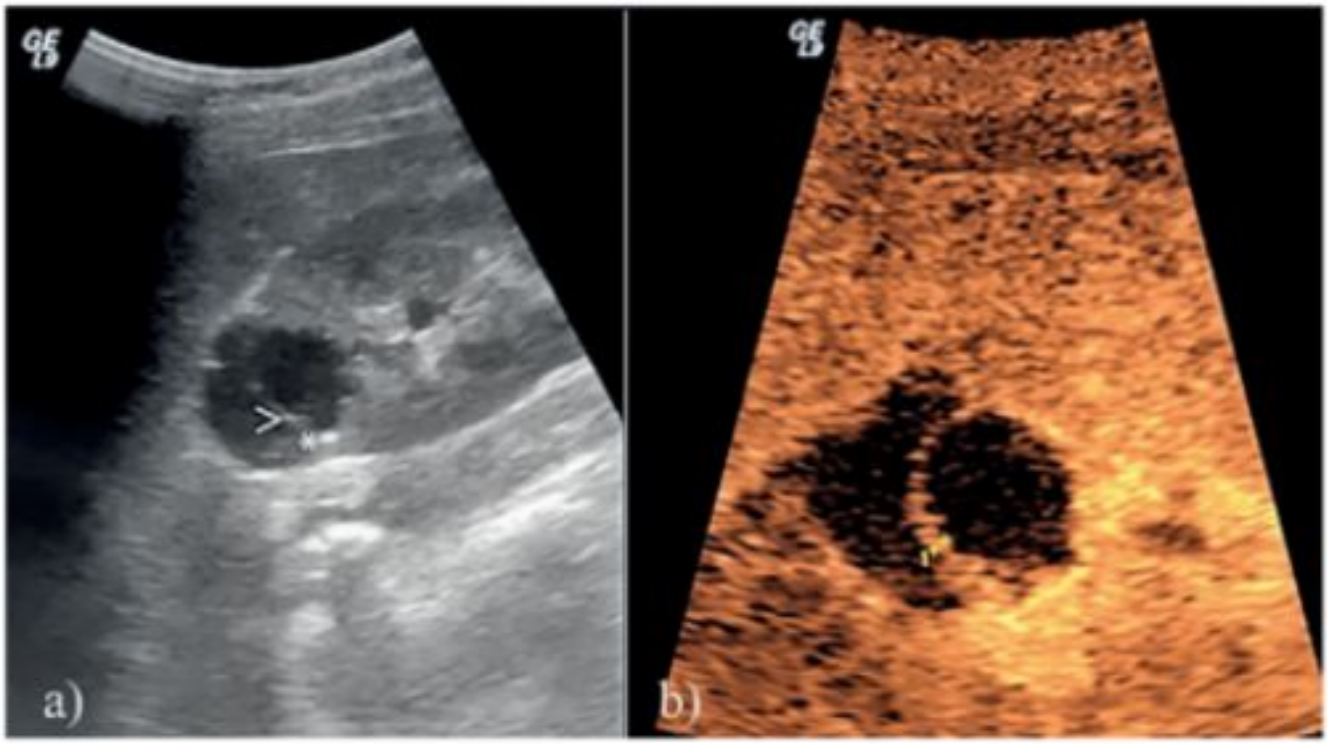
PMC6704226

3/ Imaging:

🔸 Ultrasonogram (USG) +/- doppler → best in children

🔹 Contrast enhanced USG (fig)- delineates blood blow and microcirculation, differentiates solid vs cystic components, communication with subcapsular space if present etc

https://www.cureus.com/articles/24403-the-use-of-contrast-enhanced-ultrasound-in-pediatrics-a-case-series



Traditional USG contrast enhanced USG

Radu et al 2015

4/

- CT & MRI→eval of complex cysts

Cons- sedation & contrast

- IVP/ Urography→connection to calyceal system
- Angiography- vascularization
- DMSA Scintigraphy- evaluate fn (multiple cysts)
- MR Urography: functional & comprehensive assessment, quicker

Cons: 💰,availability

Journal of Ultrasound
Official Journal of the Italian Society of Ultrasonology in Medicine and Biology

J Ultrasound. 2019 Sep; 22(3): 381–393.
Published online 2019 Jan 1. doi: 10.1007/s40477-018-0347-9

PMCID: PMC6704226
PMID: 30600488

Language: English | Italian

Pediatric cystic diseases of the kidney

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Abstract

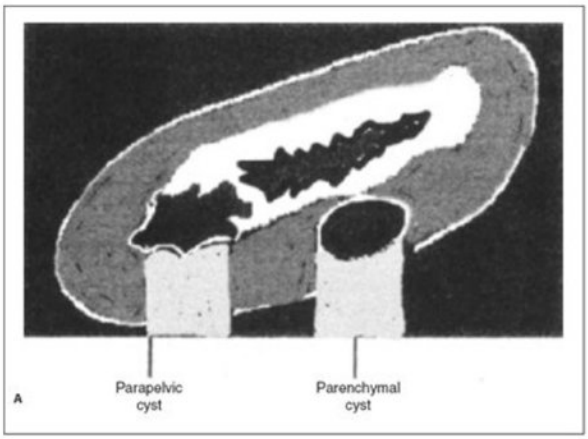
Go to:

Pediatric renal cystic diseases include a variety of hereditary or non-hereditary conditions. Numerous classifications exist and new data are continuously published. Ultrasound is the primary technique for evaluating kidneys in children: conventional and high-resolution US allows a detailed visualization of renal parenchyma and of number, size and location of the cysts, hence representing the most important diagnostic imaging technique for the first diagnosis and follow-up of these young patients. The purpose of this pictorial essay is to review the spectrum of renal cystic lesions in children from simple, complex or malignant single cysts to the several poly/multicystic kidney diseases.

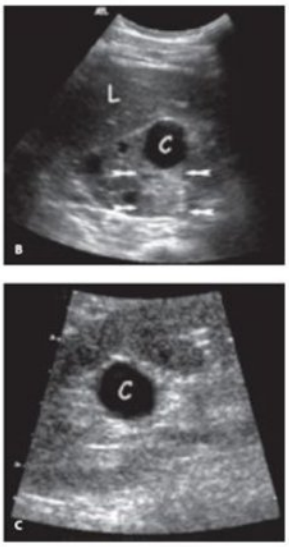
Keywords: Cystic renal disease, Kidney, Neonatal, Pediatric, Ultrasound

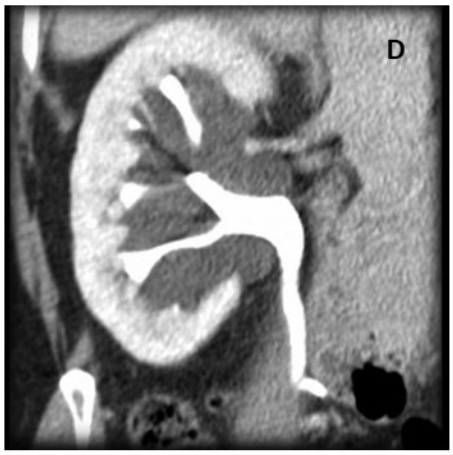
5/ DDx solitary cyst:

- Epithelial (Simple cortical) cyst
- Para & peri pelvic cyst
- Dilated minor/ major calyx- hydronephrosis or diverticulum
- Renal abscess
- Aneurysm/ Pseudoaneurysm- doppler differentiates
- Cystic Wilms in children
- Adults- Cystic/ papillary RCC, lymphoma



Parapelvic cyst: originate from parenchyma & protrudes into renal sinus, and it can cause obstruction



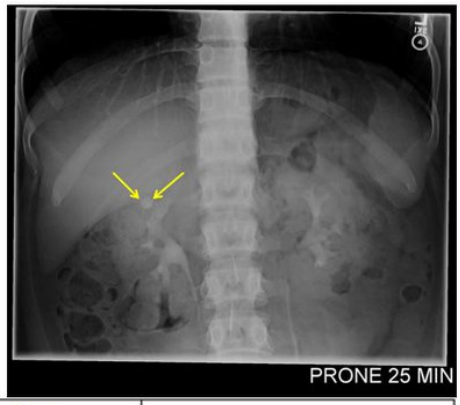


Peripelvic cyst or renal lymphangiectasia: originate within the sinus itself and have a lymphatic origin

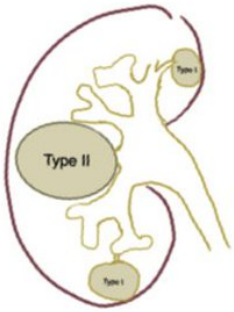
Parenchymal cyst at the midpole of the kidney shows good through transmission, smooth borders, and absence of internal echoes. Parapelvic cyst at the upper pole has an irregular outline. Parapelvic cysts can be mistaken for dilated calyces. **B.** Sonogram: cortical parenchymal cyst (C). Note enhanced through transmission behind it (arrows). Other smaller cysts elsewhere in this kidney are also noted. **C.** Sonogram: Parapelvic cyst (C) in a different patient shows a well-defined anechoic mass centered in and distorting the renal sinus fat. **D**. Peripelvic cyst (D)

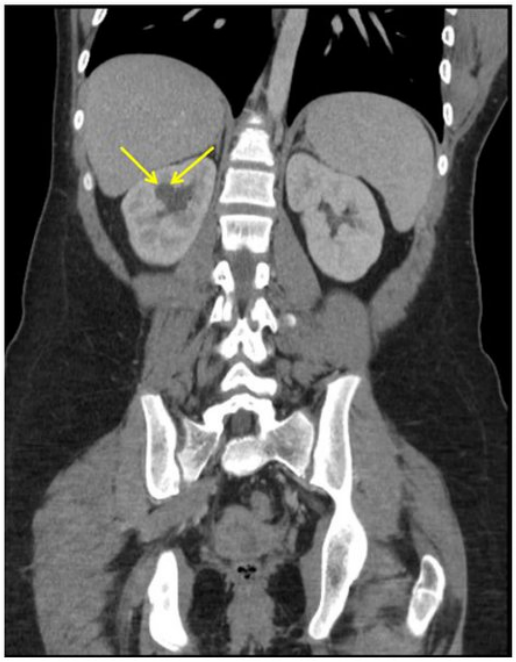
https://radiopaedia.org/cases/peripelvic-cysts-1
https://radiologykey.com/possible-renal-mass/

Calyceal diverticulum: prone image from an intravenous pyelogram 25 minutes after intravenous iodine contrast injection reveals a small pooling of contrast in an upper pole calyx in the right kidney (yellow arrows) connected by a narrow channel to the renal pelvis, compatible with a calyceal diverticulum.



Characteristics	Types	
	I	II
Site of communication with Pelvicalyceal system	Minor calyx	Renal Pelvis/Major Calyx
Location of lesion	Polar	Midzone
Symptoms	Rare	Common





Calyceal diverticulum: Coronal CT of the abdomen and pelvis with contrast reveals a small outpouching in an upper pole calyx of the right kidney (yellow arrows), compatible with a calyceal diverticulum.

https://radiologypics.com/2013/03/02/solution-to-unknown-case-5-calyceal-diverticulum/
https://www.jpurol.com/article/S1477-5131(20)30017-6/abstract

6/ Differential diagnosis for Multiple Cysts/ Ciliopathies:

- ADPKD
- ARPKD
- MCDK
- NPHP
- HNFβ1- asso. PKD
- Glomerulocystic disease
- TSC

Adults: above, and ADTKD, VHL, MSK, Glomerulocystic kidney disease, Dialysis asso. cysts, Lithium-related cysts, cystic nephroma, RCC, etc

Genetic Kidney disease with cystic phenotype

Loftus 2013
PMID: 22736301

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Name	gene	Age at presentation	Clinical features	
			Renal	**Extra-renal**
ADPKD	*PKD1* (16p13.3-p13.12)-85% *PKD2* (4q21-q23)- 15% *PKD3*- rare (Chr11)	Usually 20-40 yrs	Abdominal masses, polyuria, polydipsia, UTIs, ESKD (earlier with *PKD1* mut– mean 53 yrs)	Liver and Pancreatic Cysts, Sub-Arachnoid hemorrhage
ARPKD	A.R. *PKHD1* (6p21.1-p12)	Neonatal, childhood	Abdominal pain. UTIs. ESKD in infancy or childhood	Oligohydramnios if severe, ascending cholangitis, HTN. ~30% Perinatal mortality, mostly due to lung hypoplasia & complications.
Nephronophthisis (NPHP)	A.R. >20 genes. *NPHP1-13*	3 forms. Infancy, childhood, adolescence	Polyuria, polydipsia, enuresis, ESKD	Growth retardation, anemia, (visual loss, liver fibrosis, cerebellar ataxia if associated with another syndrome)
MCKD/ ADTKD	A.D. UMOD, REN & MUC1	Early adulthood	ESKD	Gout
HNF1β-related diseases	*HNF1β* (17q12)	Any age	Variable. Hyperechogenic kidneys, MCDK, agenesis, hypoplasia, cystic dysplasia, or hyperuricemic TIN not a/w *UMOD* mutation	Anomalies of the GUT, pancreas atrophy, liver abnormalities, **MODY 5** (Maturity Onset Diabetes in Young type 5) & genital malformations
VHL	A.D. *VHL* (3p25.3)	Any age (mean 26)	Kidney cysts, renal cell carcinoma (RCC)	CNS or retinal hemangioblastomas, pheochromocytoma, pancreatic cysts
TSC	A.D. *TSC1* (9q34), *TSC2* (16p13.3). High rate of spontaneous mutations	Childhood	Renal cysts-seen in contiguous deletion of ADPKD1 & TSC2 genes. Renal AML. ESKD.	Facial angiofibromas, cardiac rhabdomyomas, retinal hamartomas, lymphangioleiomyomatoses
Renal cysts in syndromes	Varies according to syndrome (including Meckel–Gruber, Bardet-Biedl, Ehlers-Danlos, Trisomy 13, 18 and 21, & Zellweger syndromes)			

PKD- polycystic kidney disease, MCKD: Medullary Cystic Kidney Disease, ADTKD: AD tubulointerstitial kidney disease, HNF: Hepatic Necrosis Factor, MCDK: Multicystic dysplastic kidney, UMOD: Uromodulin, GUT: genitourinary tract, VHL: Von Hippel Lindau disease, TSC: Tuberous sclerosis complex.

Developmental Kidney disease with cystic phenotype				@drM_sudha
Name	gene	Age at presentation	Clinical features	
			Renal	**Extra-renal**
Medullary sponge kidney	*GDNF* mutations. May be part of other syndromes	20–50 years but may present younger	Hematuria, UTI and calculi	-
Multicystic renal dysplasia	Usually sporadic. Familial- *PAX 2*. A/w many syndromes	Usually detected prenatally or soon after birth	Abdominal mass, flank pain, UTI. UPJ obstruction: small kidneys Familial/ syndromic: large kidneys	Hypertension
Acquired Kidney disease with cystic phenotype				
Name	Incidence	Age at presentation	Clinical features	
			Renal	**Extra-renal**
Acquired renal cystic disease	Acquired. 7–22 % of pre-dialysis patients. >90 % 10 years post-dialysis	Any age depending on age of development of ESKD	Flank pain, bleeding, RCC	-
Simple renal cysts	Very common. Incidence increases with age. M:F 2:1	Any age. Usually incidental finding	Rarely pain, bleeding, infection	-
Multilocular renal cysts	Rare	Any age but often early childhood	Often asymptomatic. abdominal mass, abdominal pain, or hematuria	Hypertension
Hypokalemic renal cysts		Any age	Usually cysts do not cause symptoms	- Loftus 2013 PMID: 22736301

Glomerulocystic kidney - Genetic and non-genetic forms				@drM_sudha
Name	Gene	Age at presentation	Clinical features	
			Renal	**Extra-renal**
GCK in PKD	ADPKD	Any age	As per primary disease	As per primary disease
Hereditary GCKD	ADGCKD and HNF1β mutations	Any age	Abdominal masses, renal insufficiency, flank pain, hematuria	Hypertension
Syndromic GCK	As per syndrome	Any age	As per syndromes, e.g., X-linked dominant oral-facial-digital syndrome type 1, tuberous sclerosis	-
Obstructive GCK	-	Any age	Associated with renal dysplasia. Urinary tract infections	-
Sporadic GCK	de novo mutation, ischemic, or drug-induced	Any age	Abdominal masses, renal insufficiency, flank pain, hematuria. Described post-hemolytic uremic syndrome	Hypertension Loftus 2013 PMID: 22736301

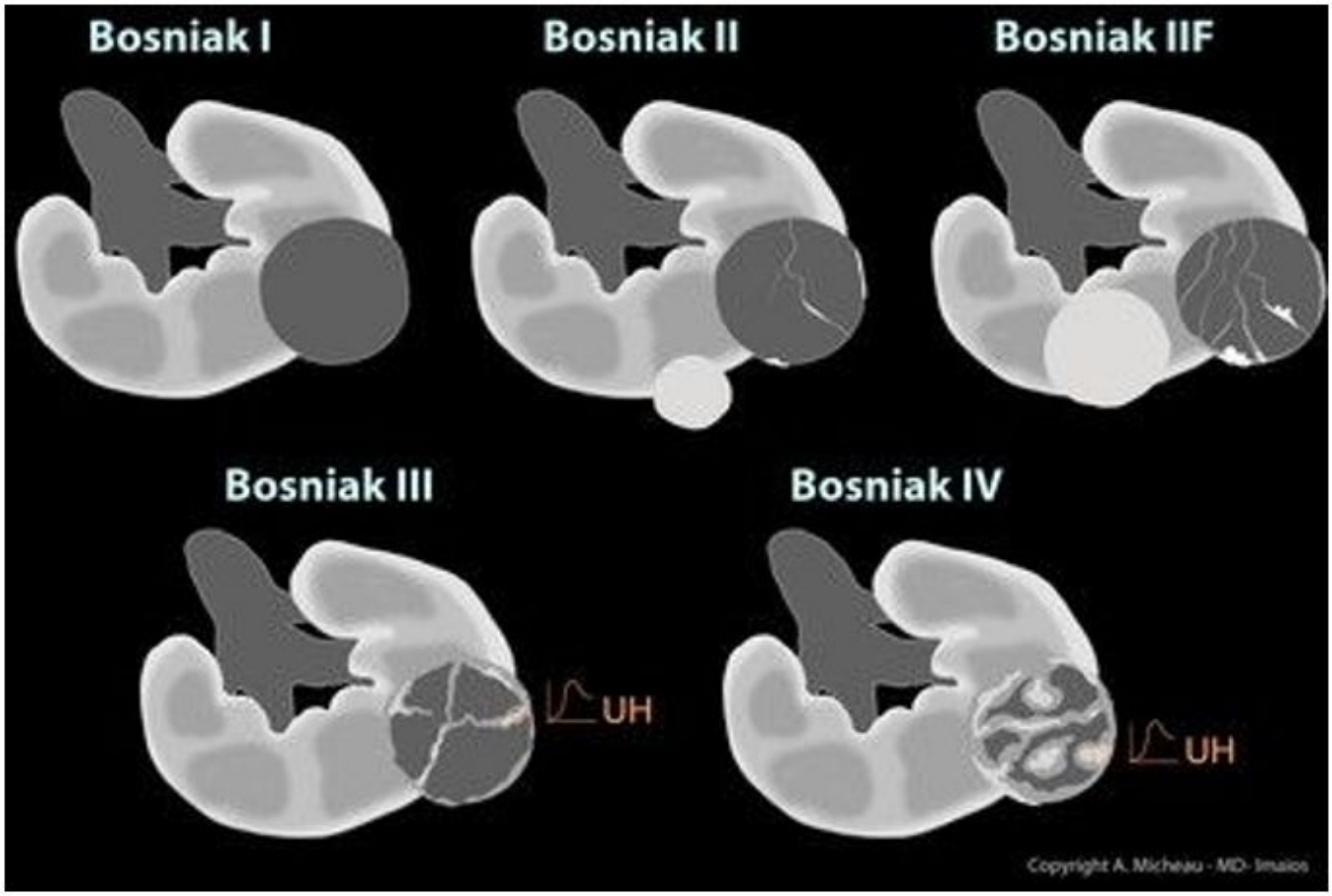
7/ How to assess the malignant potential of cystic kidney lesions in children?

⚡The modified Bosniak classification system⚡

🔹Class 1 or 2- most often benign

Class 2- periodically monitored

🔸class 3 or 4- >90% has intermediate / malignant pathology -excision warranted



https://www.imaios.com/en/e-Cases/Channels/Radiology/Radiological-classifications-commonly-used-in-medical-imaging/Renal-Cysts

Table 1 The modified Bosniak classification system for renal cysts, based on ultrasound findings.

Grade	Shape	Wall			Septations				Calcification	Content
		Thickness	Nodules	Doppler flow	Number	Thickness	Nodules	Doppler flow		
I	Round	≤1 mm	No	No	0	N/A	No	No	No[a]	Anechoic
II	Lobulated	≤1 mm	No	No	Few	≤1 mm	No	No	No	Debris
III	N/A	≥1 mm	No	Yes	Multiple	≥1 mm	No	Yes	Yes	N/A
IV	N/A	N/A	Yes	N/A	N/A	N/A	Yes	N/A	N/A	Soft tissue

[a] Not including movable cyst stone.

Published in Journal of pediatric urology 2015

Ultrasound classification of solitary renal cysts in children.

B. Karmazyn, A. Tawadros, +5 authors M. Kaefer

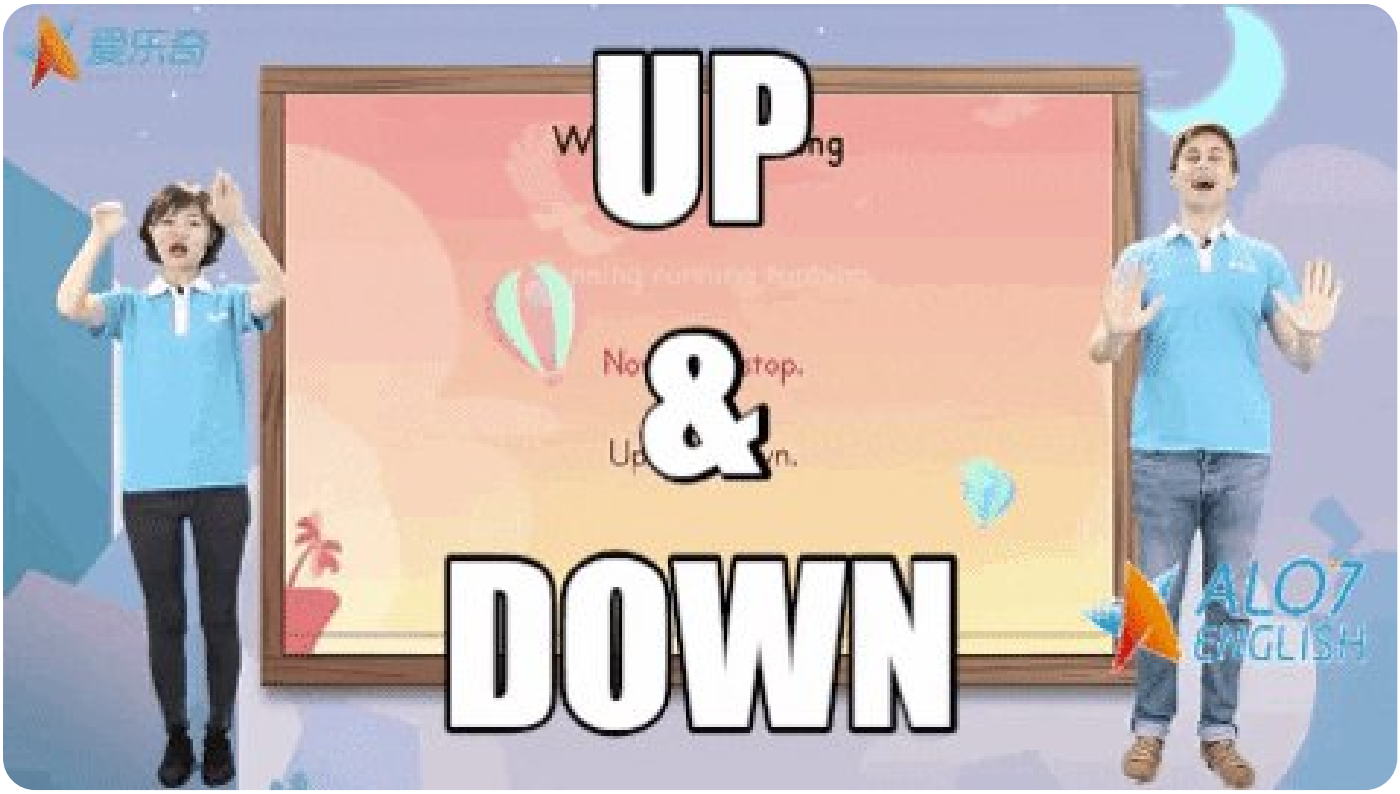
8/ Let's pit MRI/ CT vs USG for classifying kidney cysts.

For Eg if you have Bosniak 2 category of cyst by USG-

9/ B.

I was fascinated to know from @ASPNeph webinar that the sonogram is better at classifying the cyst. MRI/CT scan can downgrade the class but not upgrade. What is your experience?

Share it with us in the comments.



10/ That's all folks!
Here's a tweetorial on pathogenesis of cysts and ciliopathies...

11/ For a case-based clinical discussion with radiology expert login to @ASPNeph website, April webinar. Answer questions to get #MOC2credits #Membereducation #ASPNFOAMgroup

Until next time...
@RoshanPGeorgeMD @SwastiThinks @basalely @priti899