SOLITARY renal CYSTS/cystic mases

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Overview

- Cystic Renal Mass imaging
- How seriously should we take solitary renal cysts?
- Sudarsana has given the medical perspective
- Abhijit Dixit the genetics
- This is an imaging perspective
- I will demonstrate a number of different patterns of cysts
- Explain how we assess cysts (size, location, characteristics etc).
- Show some things that look like cysts but aren’t.

Solitary cysts ; how seriously we should take them.

-Most cystic renal masses encountered incidentally are benign and can be diagnosed confidently on imaging and require no follow-up.

-Either a cyst is simple or it is Not

-If it is not and minimally complex we would probably follow-up with US in a year or so (there is no consensus on the time interval and MR IS NOT MORE SPECIFIC THAN US IN CHILDREN). Most end up not changing.

-If a cyst has any solid components /complex septae /enhancement then this is taken more seriously. Wilms which is the most common renal paediatric tumour can have cystic change. The only other tumour, nephroma, is rare and benign. Metastasis can have cystic change.

Simple renal cysts in children: diagnosis and follow-up with US. K McHugh, D A Stringer, D Hebert, C A Babiak Published Online:Feb 1 **1991**

A study done a while back by a reknowned paediatric radiologist to assess the US frequency of simple renal cysts reviewed the results of 16,102 children over a 5-yr period. 37 simple cysts in 35 patients were reviewed. Follow-up US of 23 cysts in 22 patients for up to 5 yrs showed no change in size in 17 cysts (74%). The largest cyst was drained percutaneously; all other cysts were managed conservatively. No patient showed deterioration of RFTs. The authors concluded that in a paediatric patient demonstrating normal RFTs, no further intervention is necessary when a simple renal cyst is identified at US.

More recently, a committee of international experts in paediatric nephrology, radiology, and US formulated recommendations at a consensus meeting. The final statement was endorsed by the European Society of Pediatric Radiology, the European Federation of Societies for Ultrasound in Medicine and Biology, the European Society of Pediatric Nephrology, and reviewed by the European Reference Network for Rare Kidney Diseases.

Imaging of Kidney Cysts and Cystic Kidney Diseases in Children: An International Working Group Consensus Statement Article in Radiology 290(3):181243 ·

December 2018

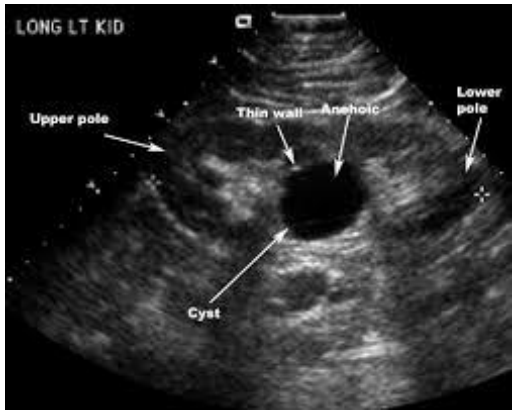
International Consensus Statement – has provided clinical **guidance** on standardization of imaging tests to evaluate kidney cysts in children.

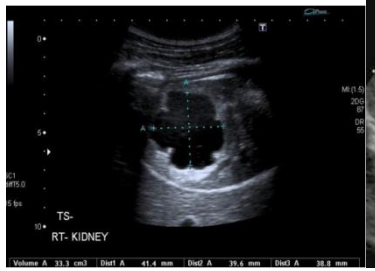
Dec 2018

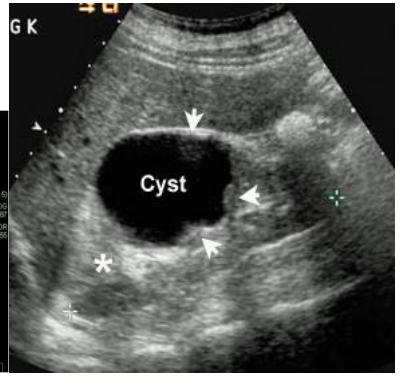
- Main recommendations are as follows:-
  - US is the method of choice when assessing paediatric kidney cysts, with selected indications for MRI and contrast-enhanced US.
  - CT should be avoided whenever possible because of ionizing radiation. Renal US yields essential diagnostic information in many cases.
  - US is usually sufficient for follow-up kidney imaging, but MRI can be valuable for clinical trials in patients with ADPKD or in older children with tuberous sclerosis complex to evaluate both kidney cysts and angiomyolipomas.

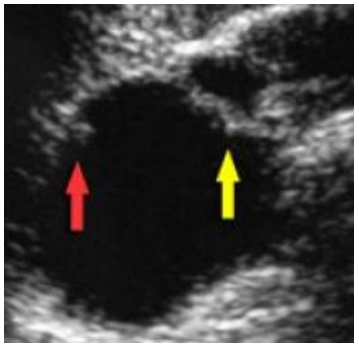
How we assess cysts (size(>5cm =large and may need drainage), location(parapelvic parenchymal, cortical), characteristics etc).

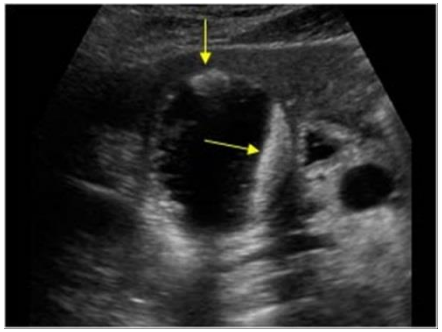
- If is thin-walled, anechoic, nonseptated, separate from the collecting system, and has no Doppler blood flow related to the cyst
- Cysts with additional features, such as solid components, septations, or thickened walls, should be considered complex cysts.. ...











Management of simple renal cyst in children: French multicenter experience of 36 cases and review of the literature Article Apr L. Joyeux E Sapin Nicolas Koutlidis 2015

Bosniak classification
good to know but not vital in paeds

- Bosniak criteria has remained the reference standard for diagnosis and classification of cystic renal masses for >30yrs
- However this criteria were 1st described and are intended for use primarily with CT in adults
- There is limited data to suggest this can be adapted to US in paediatrics.

Bosniak classification of renal cysts

Bosniak 1

- Simple cyst with pencil thin wall. No soft tissue, septa, or calcifications. No enhancement.

Bosniak 2

- Benign simple cyst with **pencil thin septa** / **thin calcifications.**

Followup

Bosniak 2F –very low malignant potential 1%

- Multiple thin septa or minimal smooth wall thickening. Thick and nodular calcifications may be present. **There must not be any measurable enhancement or soft tissue.** This category also includes totally intrarenal lesions >3 cm.

Surgery

Bosniak 3- low malignant potential 10%

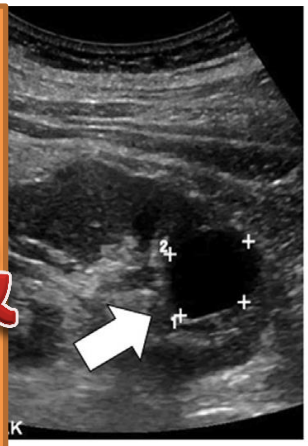
- Complex cysts with thickened wall and/or septa with **measurable** enhancement.

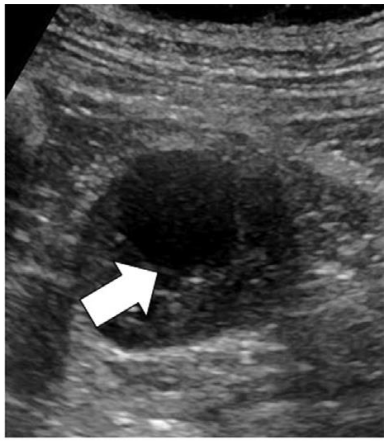
Bosniak 4

- Clearly malignant cystic masses that can have all of the criteria in category 3, but also contain distinct soft-tissue component separate from the wall or septa

Different patterns of cyst- using Bosniak classification

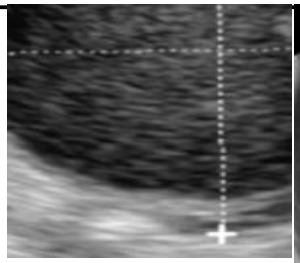
1 & 2

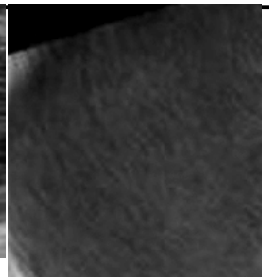


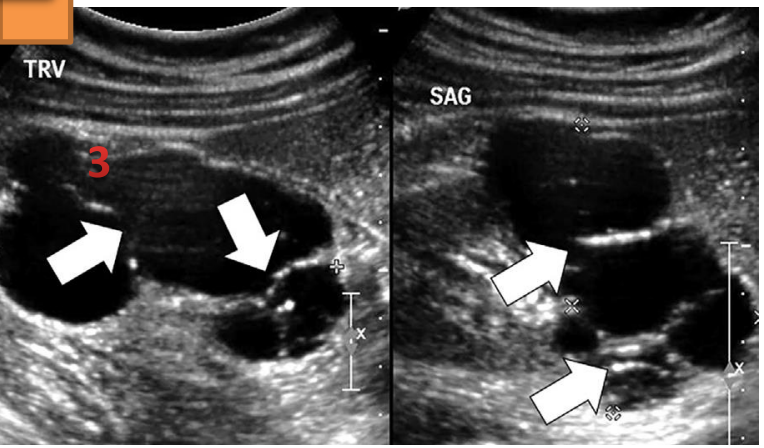


1)Simple cyst (Bosniak 1) Sag&Trvs US anechoic simple cyst with an imperceptible wall and increased through transmission

2)Proteinaceous or Haemorrhagic cyst (B2)



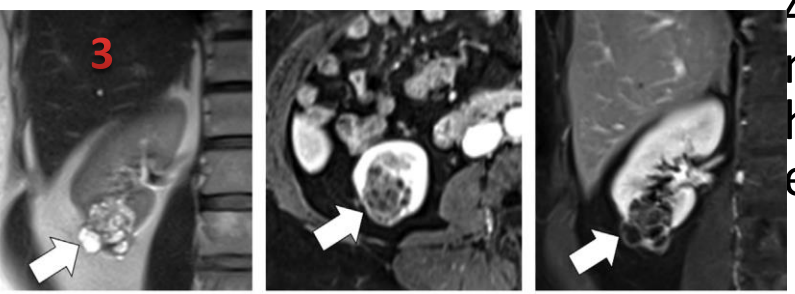




3)Complex cyst (thick/nodular septae,measurable enhancement,calcification, soft tissue components) Bosniak 3 and

Incidentally detected complex renal cyst on US.

Trvs&Sag -multiple complex thick septa 2F or 3.



4)Localised cystic disease of the kidney - multiple cysts (resembling ADPKD at histopathology) involve a segment of or even an entire kidney

How about - Simple cyst with other renal findings

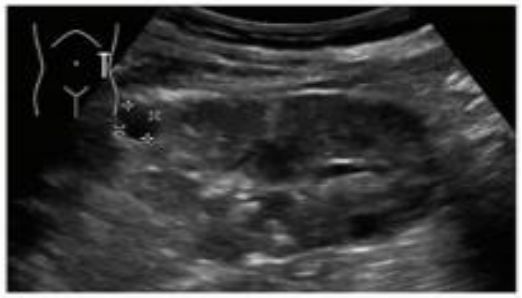
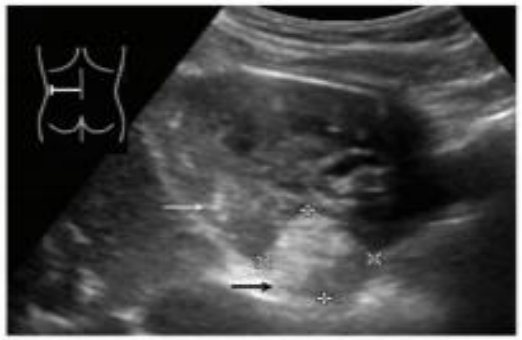
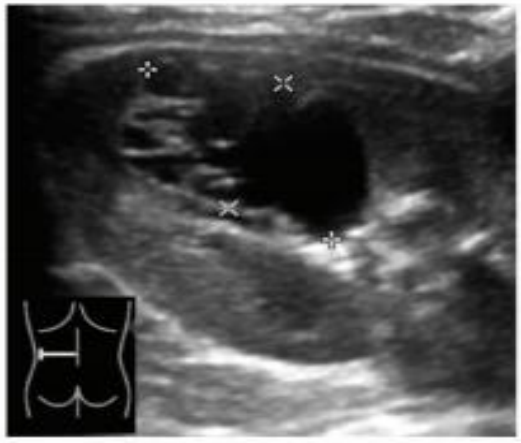
US Diagnosis	Image
Tuberous sclerosis complex (TSC) • single or multiple cysts • usually also with angiomyolipomas (AML) • increase with age	 
Complex cysts Cyst with any of: • thickened wall • septations • calcifications • enhanced perfusion on color Doppler	

Image 1a and b
Cyst and angiomyolipomas
If the AML are > 3cm MR followup
If the AML are < 3cm yrly US

Image 2 – Bosnisk 3 and beyond- surgical cyst- Biopsy/ partial nephrectomy

Some centres with expertise or in research
Contrast enhanced MR/US

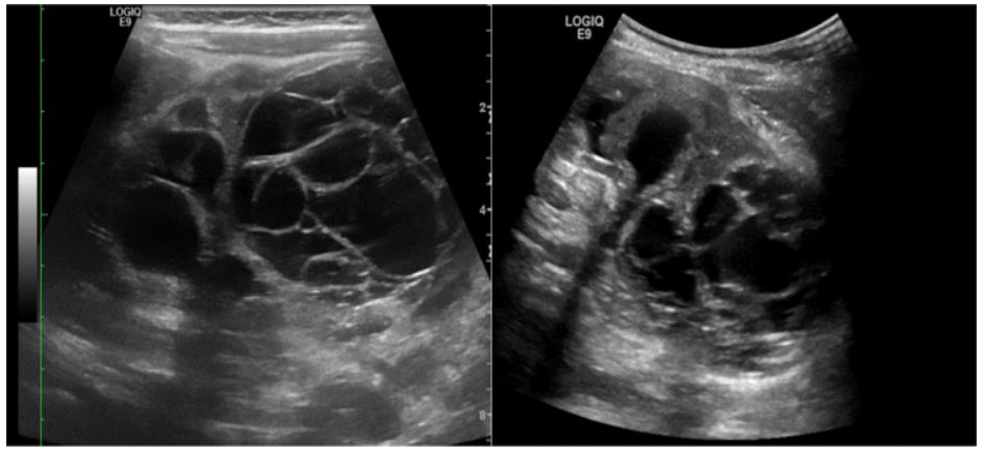
Complex cyst

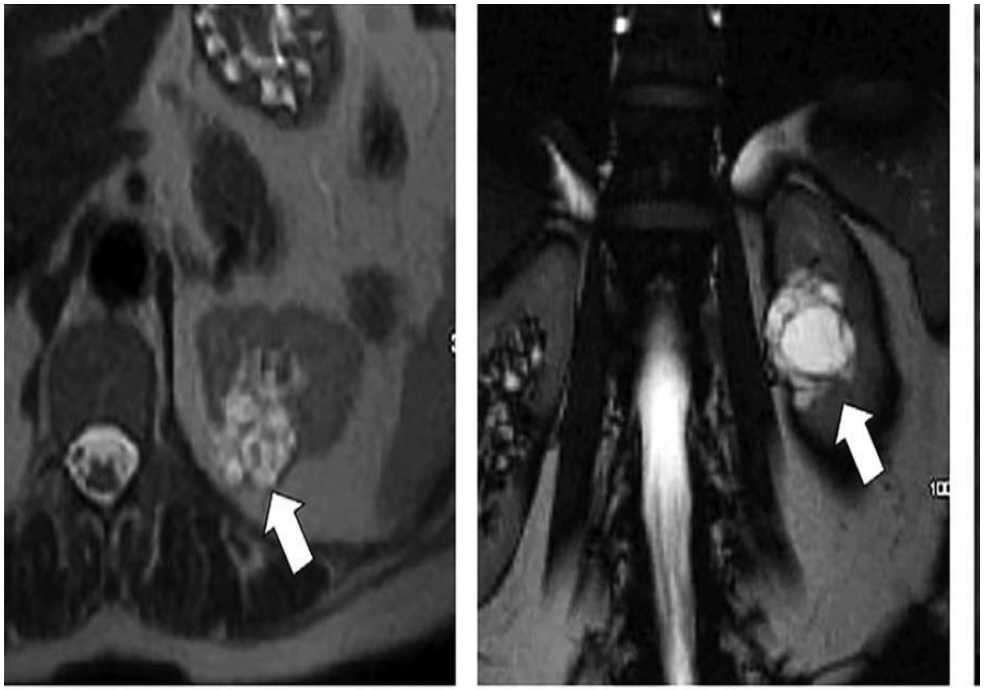
More on Complex cystic masses in children- Bosniak 3

- Cystic nephroma, is a rare benign tumour with bimodal age and sex distribution. It occurs in boys <5 years of age and in perimenopausal women. Multiple fluid filled non-communicating cysts surrounded by a thick capsule.

- No risk of malignant transformation has been reported. Childhood cystic nephroma must be differentiated from a necrotic Wilm's tumour and the presence of any solid component essentially excludes a cystic nephroma.

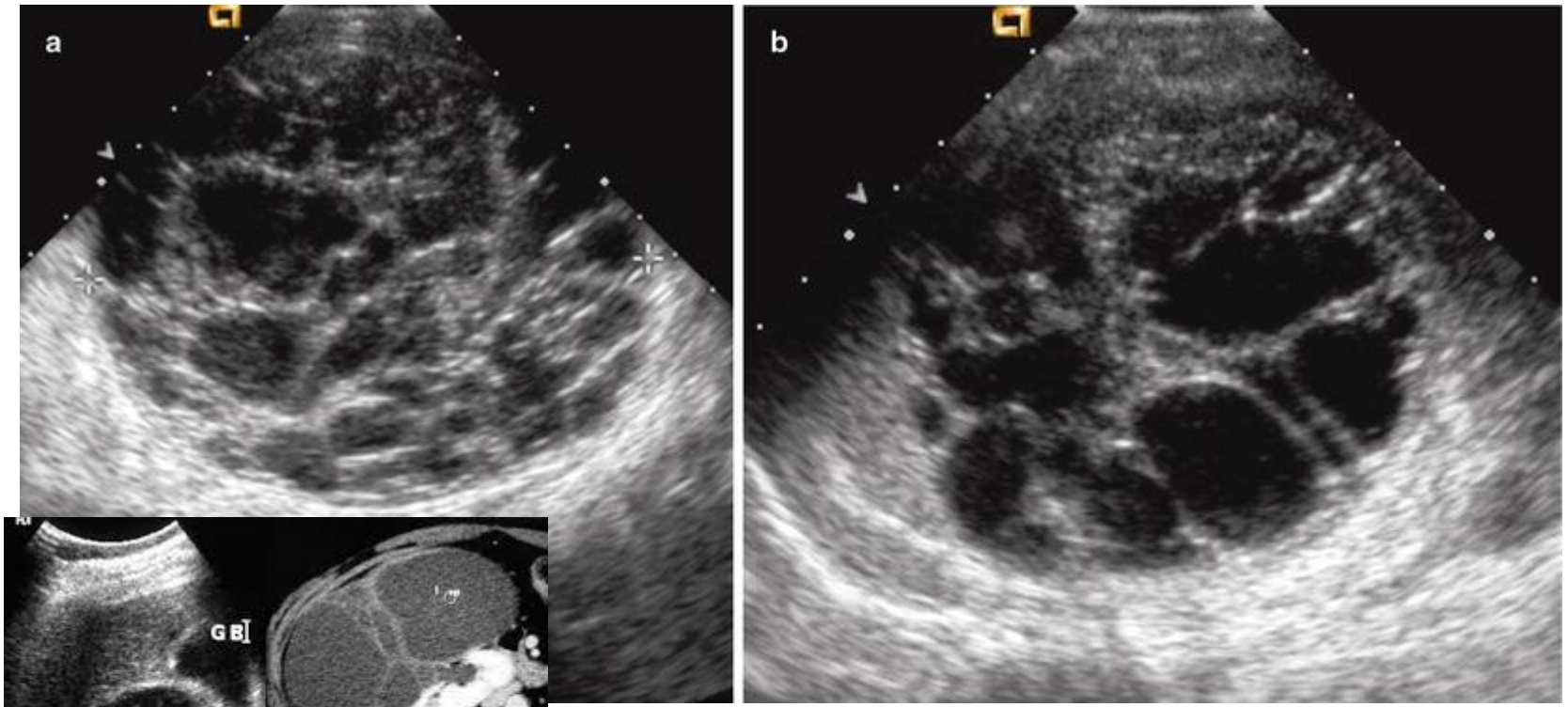
Cystic nephroma US &MR





Wilms with Cystic change- most are clearly Bosniak 4- need MR/CT and staging.

- Cystic poorly differentiated nephroblastoma (CPDN) is a rare variant of nephroblastoma.



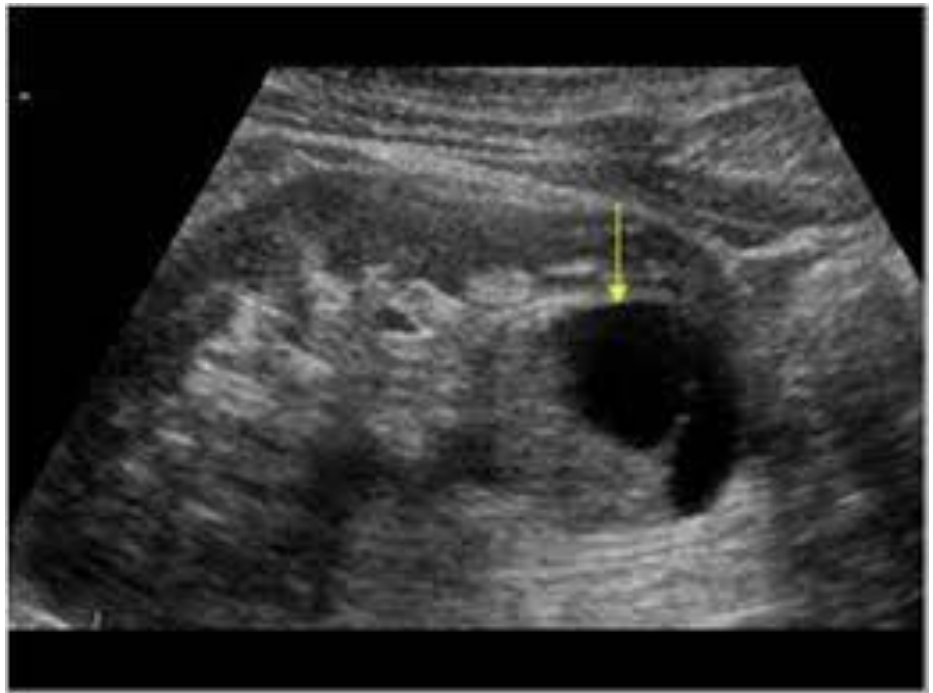
Added value of CT for surgical planning

Other less common
Malignant cystic mass

RCC IS EXTREMELY RARE IN CHILDREN

The incidence of RCC tumor in childhood is estimated to be from 1.8% to 6.3% of all malignant renal tumors

Cystic RCC



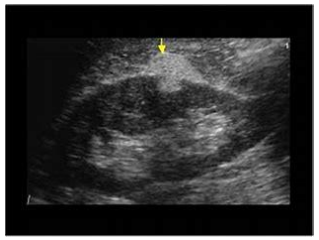
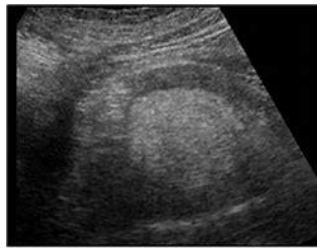
- **It may be very difficult (if not impossible) to differentiate necrosis from cystic changes on imaging.**

Followup- Solitary cysts ; how seriously we should take them.

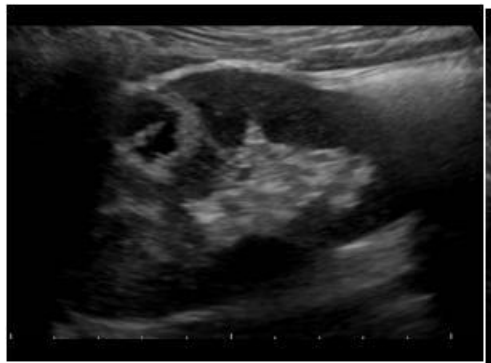
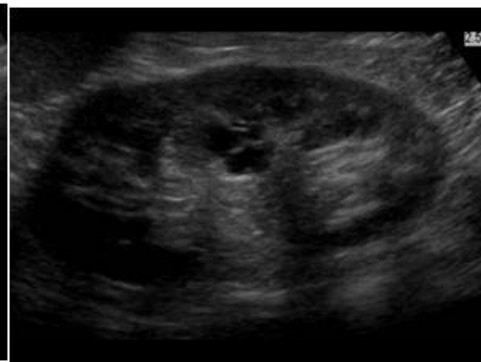
- Simple cysts- can be ignored. At most an US at 1 yr eg for haemorrhagic or proteinaceous cyst to confirm it is stable/ has resolved and there is no other change. However depending on clinical picture, renal function test derangement , size (>5cm = large ? Complications like pressure on adjascent structures /leakage etc ? Drainage.
- Complex cysts
  - Nephroma would need a biopsy- it is benign.
  - Wilms and RCC(rare)- obviously need further characterisation usually MR but sometimes CT for presurgical planning for vascular assessment.

Angiomyolipomata may appear cystic

- AML is generally a solid mass without cystic or epithelial components.

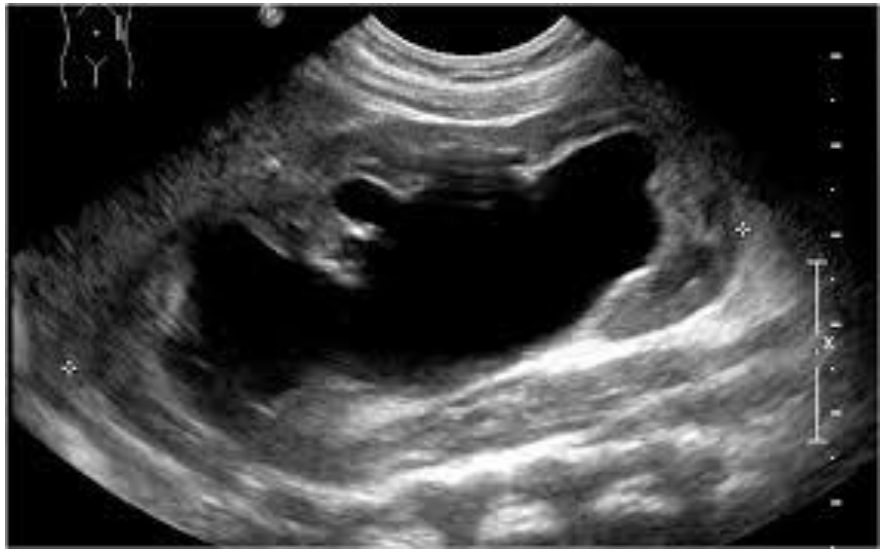



- Renal angiomyolipoma (AML) with epithelial cysts (AMLEC) is a comparatively rare benign renal tumor. AMLEC has been described as a cystic variant of AML.

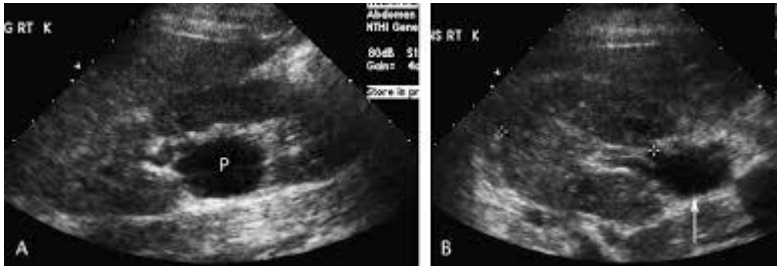



Other entities that are commonly misinterpreted as cysts

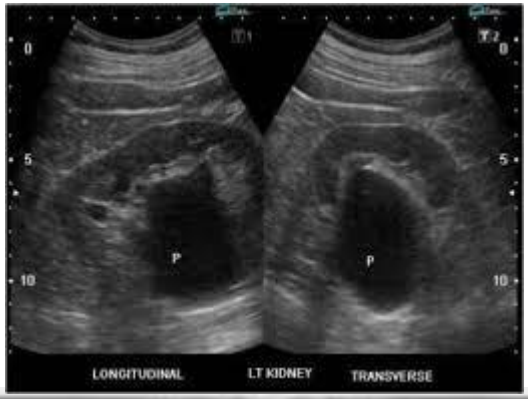
Dilated calyx- communication between the cystic components- not a cyst

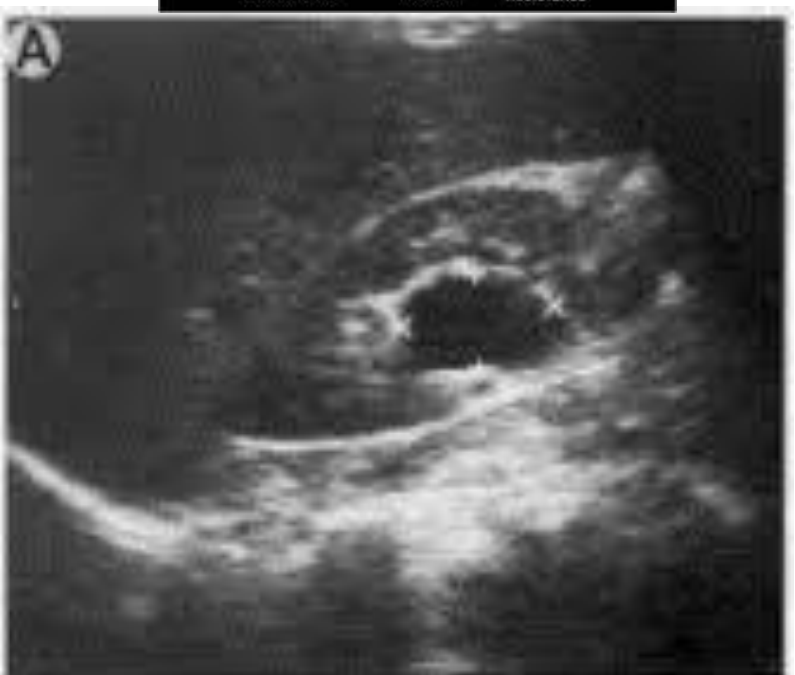


Extrarenal pelvis- best appreciated during the scan. Should be communicated in report as confusion can occur on review of images at eg MDT



Parapelvic cyst can be misinterprated as hydronephrosis





- It is important for the person performing the scan to recognise this.
- And it is equally important for separate MDT interpreters to raise this possibility if a report has made a misinterpretation

Followup- The end

- There is no established follow-up schedule in terms of timing or imaging methods.

- Follow-up or surveillance is generally performed according to local or institutional urological practice.

CURRENT RESEARCH

- Although the present standard of care is biopsy+/- surgery for Bosniak 3 cystic masses, there is an increasing body of evidence to support active surveillance instead of surgery. Follow-up every 6 months up to 2 years and then yearly follow-up is considered appropriate. An interval increased in size of the solid component to>3 cm and accelerated growth of lesion has been suggested as criteria for resection. Some authors have even suggested that Bosniak 4 lesions with solid component <3 cm can be managed with active surveillance.

- Bosniak 2F lesions are generally followed yearly for signs of radiological progression to Bosniak 3 and the duration of imaging follow-up, although also subject to institutional variation and surgeon or patient preference, has been suggested to be 4 years in the study by Hindman et al., which showed that approximately 10.9% progressed to malignancy with time to progression between 6months to 3.2 years.

References

1) Narayanasamy S et al. Clinical Radiology. Review. Contemporary update on imaging of cystic renal masses with histopathological correlation and emphasis on patient . Article history: Received 16 May 2018. Accepted 6 September 2018.
2) Bosniak MA. The current radiological approach to renal cysts. Radiology1986;158(1):1e10.
3) Bosniak MA. The use of the Bosniak classification system for renal cysts and cystic tumors. J Urol 1997;157(5):1852e3.
4) Sevcenco S, Spick C, Helbich TH, et al. Malignancy rates and diagnostic performance of the Bosniak classification for the diagnosis of cystic renal lesions in computed tomographyda systematic review and metaanalysis. Eur Radiol 2017;27(6):2239e47.
5) Eble JN, S G, Epstein JI, Sesterhenn IA. Pathology and genetics of tumors of the urinary system and male genital organs. In: World Health Organization classification of tumors. Lyon: WHO; 2004.
6) Hindman NM, Hecht EM, Bosniak MA. Follow-up for Bosniak category 2F cystic renal lesions. Radiology 2014;272(3):757e66.
7) Jhaveri K, Gupta P, Elmi A, et al. Cystic renal cell carcinomas: do they grow, metastasize, or recur? AJR Am J Roentgenol 2013;201(2):W292e6.
8) Donin NM, Mohan S, Pham H, et al. Clinicopathologic outcomes of cystic renal cell carcinoma. Clin Genitourin Cancer 2015;13(1):67e70.
9) Bielsa O, Lloreta J, Gelabert-Mas A. Cystic renal cell carcinoma: pathological features, survival and implications for treatment. Br J Urol 1998;82(1):16e20.

- Thank you
- Comments and questions are welcome during discussion time