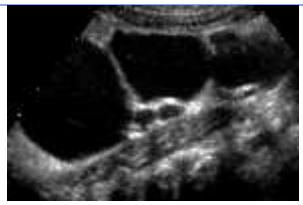
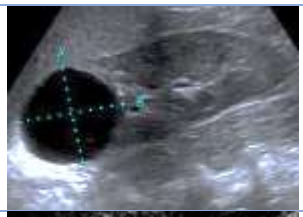
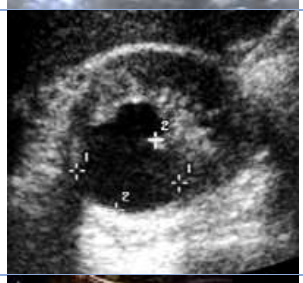
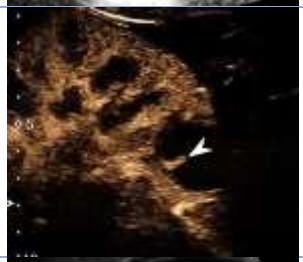
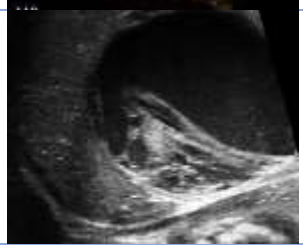
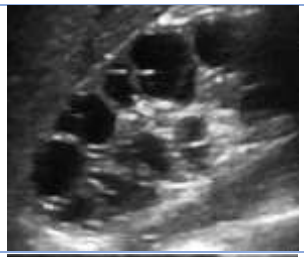
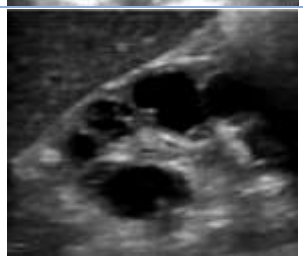
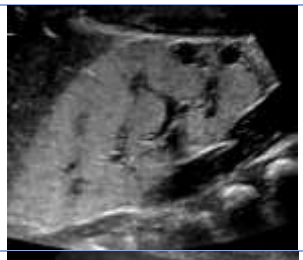
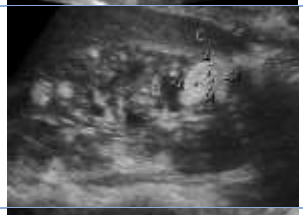
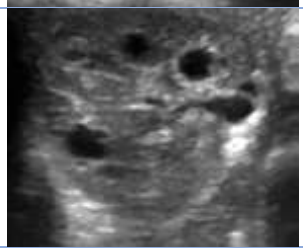
ESR Essentials: renal imaging in children - practice recommendations by the European Society of Paediatric Radiology

Electronic Supplementary Material

Table S1. Paediatric cystic kidney disease – common and rare entities to consider, typical manifestation and appearance, and follow-up needs (Table adapted from Gimpel C et al. [9], also using the chart from Riccabona M et al. [2; 3; 6; 10])

Entities	Manifestation	Follow-up or other imaging	Appearance
Non-genetic entities	May still be congenital, or manifest later in life ('acquired cyst')		
Cystic dysplasia (can be bilateral) / MCDK(no renal parenchyma, unilateral as bilateral form is lethal)	Usually congenital, secondary to ureteric obstruction Often associated with (ipsilateral) genital malformation – actively search for it	Depends on if bilateral and if with or without contralateral renal hypertrophy, risk of progression –US used for follow-up though often with little contribution to the further course and for monitoring the healthy kidney growth if unilateral	
Simple cyst	Often incidentally	Detailed medical and family history, thorough clinical examination, follow-up US – no CEUS, MRI or CT	
Acquired CKD	Usually depicted on US after event, e.g., after trauma	no follow-up, except for annual US in children after renal transplant and replacement therapy / chronic renal insufficiency DDx caliceal diverticulum - ce-MRU	
Complicated cyst	US, possibly CEUS (off label use)	Ce-MRI, if US (including CEUS) is unable to classify Apply US-or MRI-adapted 'Bosniak' classification	
Cystic tumour	Often palpable, sometimes as part of screening in tumour risk syndromes e.g. haemorrhagic Wilms tumour, (segmental) cystic nephroma …	CEUS + cross-sectional ce- imaging (preferably MRI) for diagnostic work-up, staging and follow-up (unless benign – then US will suffice)	
Genetic CDK[*1]	Inherited – however may only manifest later in life, without congenital changes		

ARPKD	US (already prenatally), typically micro-cysts in enlarged kidneys Typical ciliopathy	Renal and abdominal US (liver cysts or fibrosis, dilated bile ducts, signs of portal hypertension); follow-up annually	
ADPKD I and II	Familial or imaging suspicion, vessel and liver/pancreas/spleen involvement common Typical ciliopathy	Confirm by US, follow-up by serial US, MRI if US restricted, suspicion of malignancy Look for cysts in liver, spleen and pancreas, in older patient check for vessel complications (e.g. cerebral aneurysm)	
Nephronophthisis	Seen on US Association GCKD, MCKD, and other CKDs ciliopathy	No regular (US) follow-up Look for liver involvement in conditions with known extra-renal manifestations MCKD - medullary cysts GCKD - dilated bowman capsule resulting in mostly subcapsular and cortical cysts	
ADTKD UMOD and MUC1 (former MCKD)	Usually seen on US, includes also (segmental) 'medullary sponge kidney' Uromodulin defect	Depends on size and clinical presentation, under discussion, at least US	
ADTKD HNF1b	On US kidneys may just be hypo- or dysplastic, do not need to exhibit depictable cysts Uromodulin defect	Associated genital, pancreatic, hepatic and thymus anomalies - follow-up depending on structural kidney involvement At least once evaluate genitalia, liver and pancreas (often with early onset diabetes)	
TS-complex (classical and others) **BBS**	Seen on US, BBS consists of microcysts arising at the corticomedullary junction (dilated tubules) ciliopathy	Regular annual US follow-up, possibly initial MRI and if unclear	
Syndromic	Joubert, Bardet-Biedl, Meckel-Gruber Rare with Ivemark Sy, Hajdu-Cheney Sy, Zellweger Sy, Beckwith-Wiedemann Sy	Follow-up US imaging to assess progression of disease if of clinical impact	

Note: (US) imaging appearance initially often unspecific and indecisive, may become more specific by typical patterns when disease evolves. Do not confuse caliceal dilatation or clubbing, a dilated upper pole system in a duplex kidney, an urinoma or adrenal cystic changes with renal cysts!

[*1] In ciliopathies (hereditary defect of cilia) heterogeneous phenotypes and many organs involved (as well as in syndromic cysts and other genetic disorders) – even thymus, genitalia central nervous system or musculo-skeletal system affected
Often genetic conditions manifest already initially bilaterally – particularly ARPKD

Abbreviations: ARPKD - autosomal recessive polycystic kidney disease, ADPKD - autosomal dominant polycystic kidney disease, ADTKD - tubulo-interstitial CKD, BBS - Bardet-Biedel Syndrome, CEUS - contrast-enhanced ultrasound, ce - contrast-enhanced, ESPR - European Society for Paediatric Radiology, GCKD - glomerulocystic kidney disease, MCDK - multicystic dysplastic kidney, MCKD - medullary cystic kidney disease, TS - tuberous sclerosis, US – ultrasound

Table S2. List of common and rare solid renal lesions in childhood

Entity	Age group	Description	Remark
Variations			
Hypertrophic column of Bertin	Any age	See table 'Normal findings and variations''	DDx - hypertrophic parenchyma in scaring
Parenchymal bridge in a duplex system	Any age	On US: Central echogenicity disrupted by normal renal parenchyma without mass effect, 2 renal pelvises	Possible accessory renal vessels
Dromedary hump	Any age	Lateral contour alteration in mid-third of kidney with normal parenchyma	Only left kidney
Inflammatory			
Focal nephritis	Any age	On US: hypo- or hyperechoic 'pseudo-mass' confined to the medulla	Diagnosis made clinically and by follow-up, usually no other imaging other than US necessary
Inflammatory pseudotumour / lobar nephroma	Any age Rather rare	On US: hypoechoic homogeneous mass with sharp margins	Don't start chemotherapy unless an inflammatory process is excluded
Xanthogranulomatous pyelonephritis	Any age Rare	Typically, inhomogeneous and destructive aspect with clubbed and dilated pelvi-caliceal system, staghorn stone / calcifications	Often additional CT or MRI indicated
Abscess, necrosis, haemorrhage	Any age Rather rare	Varying echogenicity, no central vascularisation	If US is clear, no other imaging is needed
Benign and intermediate malignant			
Mesoblastic nephroma	Neonate / congenital (<8 months) Rather rare	Solid homogenous or heterogenous mass; can invade the perinephric space Hypercalcemia	'semi'-malignant, treatment - nephrectomy without further chemotherapy or staging / follow up (if histology confirms initial suspicion)
Hamartoma	Any age Extremely rare	Variable appearance on US, isoechoic, may contain calcifications and fat	Often associated with syndromes
Angiomyolipoma (AML)	Any age Very rare in patients without tuberous sclerosis	On US: mixed echogenicity, often rather hyperechoic, often multiple; echogenicity at least equal to fat in the hilum	May bleed, also seen in other abdominal parenchymal organs, particularly in the liver If little or no fat consider epithelioid AML, (prognosis different - need for biopsy)
Ossifying renal tumour of infancy	Early infancy Very rare	Gross calcification, little visible other tumour stroma, may compress collecting system	
Nephroblastomatosis / nephrogenic rests	Can be pre-malignant, > 2 years	Can easily be missed on US, CEUS may be helpful	MRI with DWI is recommended
Cystic nephroma / multilocular cystic renal tumour	Usually older (> 4 years) but can occur at any age Relatively rare	Tumour containing numerous small cysts separated by thin septae; different from adult type	If thick and irregular septa or nodular component: caveat malignancy, indistinguishable from WT
Malignant			
Wilms tumour (WT)	Mean age 3 years (and older),	90% of paediatric renal tumours, solid usually homogeneous mass, mostly	99% of WT tumours are treated without biopsy based only on imaging findings –

	most common renal tumour in this age group, but can be present even before birth and in neonates	exophytic, often with a pseudo-capsule; calcifications are rare.	According SIOP protocol biopsy recommended only with atypical clinical, imaging or biological features
Rhabdoid tumour	Young infants, mean age 18 months Relatively rare	Often subcapsular fluid collections, no pseudo-capsule, ill-defined margins, calcifications, hypercalcemia, early systemic metastasis (lung, bone, brain),	Highly aggressive, poor prognosis Note: synchronous or metachronous intracranial tumour
Clear cell sarcoma	Mean age 3 years, > 6 months Rare	Often partially cystic. Bone metastasis	Quite aggressive
Lymphoma (NHL)/ leukaemia (mostly AML)	> 3 / 5 years Relatively rare	Focal hypoechoic mass or diffuse renal enlargement by infiltration	Intermediate prognosis
Juvenile renal cell carcinoma	> 10 years Relatively rare, though more common than Wilms tumour in this age group	Same features as in adults	Intermediate prognosis

Abbreviations: AML – angiomyolipoma, CEUS – contrast-enhanced ultrasound, CT – computed tomography, DDx – differential diagnosis, DWI – diffusion-weighted imaging, MRI – magnetic resonance imaging, NHL – non-Hodgkin lymphoma, US – ultrasound, SIOP - International Society of Paediatric Oncology, WT – Wilms tumour.

Warning: Do not biopsy a renal mass; Refer affected children to a specialised centre for further imaging, staging, further workup, and treatment according to the European study protocols