Paediatric Nephro-Urology: guideline for management of the child with a solitary functioning kidney

Summary
This guideline is for the use of any colleague looking after a child with solitary kidney at Evelina London Children's Hospital. An estimated 1 in 2,000 babies are born each year with kidney agenesis and the frequency of solitary functioning kidneys is as much as 1 in 500-1 in 800.

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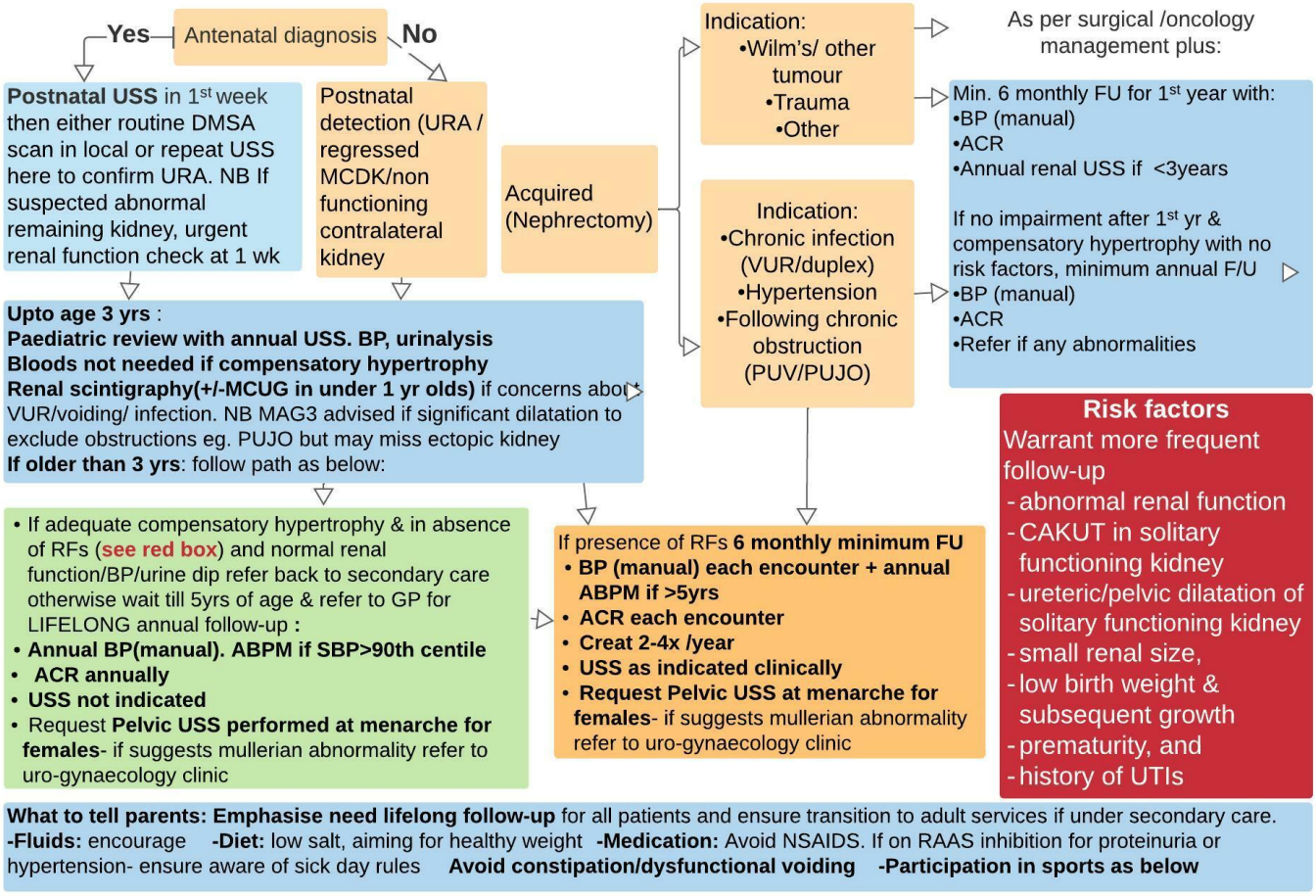
INTRODUCTION: A solitary functioning kidney occurs as the result of a contralateral (1) renal agenesis (2) multi-cystic dysplastic kidney, (3) atrophic or dysplastic kidney or (4) nephrectomy. It may predispose to hypertension, proteinuria and chronic kidney disease (CKD) as a consequence of hyper-filtration. Therefore, increased surveillance to introduce timely preventative strategies are required.

MANAGEMENT AFTER DETECTION



Key : URA =unilateral renal agenesis; MCDK=multicystic dysplastic kidney; CAKUT= congenital anomolies of kidneys and urinary tract; PUJO=pelvi-ureteric junction obstruction; PUV:=posterior urethral valves; MCUG=micturating cystourethrogram; USS-=ultrasound scan; ACR= albumin creatinine ratio; ABPM= ambulatory blood pressure monitoring; RAAS= renin angiotensin aldosterone system

KEY POINTS FOR INVESTIGATIONS AND MANAGEMENT (PLEASE SEE ALGORITHM ABOVE). The incidence of solitary kidneys is relatively common and estimated between 1 in 500 and 1 in 800 births**.** Not all children need follow-up at ELCH. In the absence of risk factors and in context of normal renal function, absence of hypertension and proteinuria, and adequate compensator hypertrophy, annual review at the general practice or with local paediatrician is sufficient. Emphasis is that follow-up should be **lifelong**. During puberty, there is risk of decline in kidney function, which persists into adulthood including during pregnancy in females (pre-eclampsia, gestational proteinuria or hypertension, UTIs). Avoid nephrotoxic medication as far as is possible and monitor drug levels where applicable.

ASSESSING FOR ADEQUATE COMPENSATORY HYPERTROPHY

Adequate hypertrophy is when kidney length is ≥ 2 standard deviations (>95th centile) above the mean for their size. Serial plotting of the growth of the solitary kidney on a kidney growth chart or an online renal growth calculator (e.g. https://www.prevmed.sunysb.edu/jjc/MrNomogram/default2.aspx) will help identify patients whose kidneys are not showing compensatory hypertrophy.

MONITORING COMPLICATIONS INCLUDING REDUCTION IN GFR, ALBUMINURIA AND HYPERTENSION

- **Plasma creatinine measurement:** As proteinuria and hypertension act as portents of CKD, surveillance blood tests are not necessary unless risk factors develop or are present. Children are particularly vulnerable during the rapid growth of adolescence
- **BP monitoring:** Annual blood pressure measurement is recommended and should be life-long. If elevated, recheck on two further separate episodes (as recommended by European Society of Hypertension guidelines) and if persistently elevated, ambulatory blood pressure measurement (ABPM) or home Doppler measurements to confirm hypertension, and refer to Nephrology if abnormal. If elevated >95th centile for age/sex/height, refer without delay. Antihypertensive medication to be initiated only in conjunction with nephrology input.
- **Monitoring for proteinuria:** Besides hypertension, albuminuria is another early marker of glomerular hyper-filtration. The accuracy of stick urine testing correlates poorly with UACs when assessing outcomes. Perform a first morning void UAC ratio as a screening tool for this during yearly life-long screening visits. If elevated refer to Nephrology. Treatment should be considered in case of modestly elevated UAC ratios (3.39-33 mg/mmol) and is strongly advised in case of a UAC ratio of >33mg/mmol). Again this should be initiated in conjunction with nephrology input.

GENETIC SCREENING: Limit genetic screening to children with additional anomalies or a positive family history of renal tract abnormalities. Other associated anomalies include anorectal, bladder, uterine, pulmonary hypoplasia, talipes or cardiac (atrial or ventricular septal defects). Enquire about maternal diabetes mellitus or use of specific drugs (immunomodulatory, anticonvulsants, and RAAS inhibitors) during pregnancy.

SCREENING FOR MÜLLERIAN TRACT ABNORMALITIES: The paramesonephric (Müllerian) and mesonephric (Wolffian) ducts are embryologically related. In males, this might manifest as seminal vesicle hypoplasia/absence of the vas deferens but this often goes unrecognised. In females, abnormalities may need surgical intervention. After the onset of menarche, enquire about cyclic abdominal pain. Mullerian abnormalities should be excluded by ultrasound (including obstructed hemi-vagina and ipsilateral renal anomaly (OHVIRA) syndrome and Mayer-Rokitansky-Küster-Hauser syndromes).

- **USS of pelvis assessing internal organs in 1st month of life** as baseline and may help understand Müllerian anatomy. This could be combined with 1st postnatal renal scan.
- **USS of pelvis at menarche** to detect OHVIRA group and non-obstructive Müllerian anomaly.
- **USS pelvis if menarche not achieved by 14 years age** to detect MRKH anomaly.

WHAT TO TELL PARENTS:

As their child has only one functioning kidney, renal injury, of any aetiology, increases the risk/degree of renal insufficiency. Loss of that kidney might result in the need for dialysis or a renal transplant, and lifelong medications.
What they can do:

- **Ensure child attends for LIFELONG annual review of blood pressure, urinalysis plus any further appointments if abnormalities identified.**
- **Ensure transition to adult services if under secondary care follow-up.**
- **Fluid intake** should be encouraged.
- **Diet should be healthy with the aim to keep the child's weight in a healthy range.** Avoid excessive salt. No evidence for limiting protein intake (and children need sufficient protein to grow) but also advised to avoid excess to reduce risk of glomerular hyperfiltration.
- **Constipation and dysfunctional voiding** should be flagged to ensure addressed promptly.
- **Medication**
    - Avoid NSAIDS**.** Other drugs may be nephrotoxic and should always check before administering.
    - If on ACE inhibitors for proteinuria or hypertension, to be aware to cease when reduced fluid intake
    - Antibiotic prophylaxis: There is no evidence that antibiotic prophylaxis is of any value in children that have a solitary kidney unless it is associated with suspected vesico-ureteric reflux and/or recurrent UTIs.
- In females parents should be counselled about the possibility of co-occurring Müllerian duct anomalies. Advised to attend GP for USS at onset of menarche (as above).
- **Participation in sports** Children with a solitary kidney are ***not at higher risk*** **for renal injury during contact sports**, but if an injury occurred it could result in loss of kidney function and the need for dialysis. Renal injuries from motor vehicle accidents are much more common than injuries from sports activities. Therefore, their child should always be in appropriate car restraints and be aware of pedestrian/bicycle road safety. Renal injury can result from contact/collision or limited contact sports, but the risks of head injury are greater to keep things in perspective. Wearing protective padding during contact/collision sports +/- limiting contact sports may decrease the risk of renal injury. Pedal bicycling, sledding, downhill skiing/snowboarding and horse-related activities may carry a higher risk than other activities.

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