REFERRAL OF ANTENATALLY DIAGNOSED RENAL ABNORMALITIES - CHW

PRACTICE GUIDELINE©

KEY POINTS

- Unilateral mild to moderate hydronephrosis can generally be managed with local ultrasound surveillance
- Severe unilateral hydronephrosis should be referred to Nephrology/Urology
- Consider antibiotic prophylaxis especially in children with bilateral severe hydronephrosis or in the presence of hydroureter
- Significant renal anomalies such as bilateral severe hydronephrosis and bilateral renal dysplasia should be referred for antenatal renal consultation

CHANGE SUMMARY

- N/A – New document

READ ACKNOWLEDGEMENT

- The following staff are to read and be aware of this document:
  - Fetomaternal staff and Obstetricians
  - Neonatal intensive care staff
  - Postnatal ward staff
  - Paediatricians

This document reflects what is currently regarded as safe practice. However, as in any clinical situation, there may be factors which cannot be covered by a single set of guidelines. This document does not replace the need for the application of clinical judgement to each individual presentation.

Approved by:	SCHN Policy, Procedure and Guideline Committee	
Date Effective:	1st June 2022	**Review Period:** 3 years
Team Leader:	Staff Specialist	**Area/Dept:** Renal CHW

Introduction

Purpose

The purpose of this guideline is to provide guidance for referral of babies with antenatally diagnosed renal abnormalities

Expected Outcomes

- Facilitate referral to The Children's Hospital at Westmead for high-risk abnormalities for counselling by the paediatric nephrology department and genetic testing
- Reduce parental anxiety about outcomes of children with antenatally diagnosed renal tract abnormalities
- Provide guidance for follow up referrals for children with renal abnormalities

Definitions

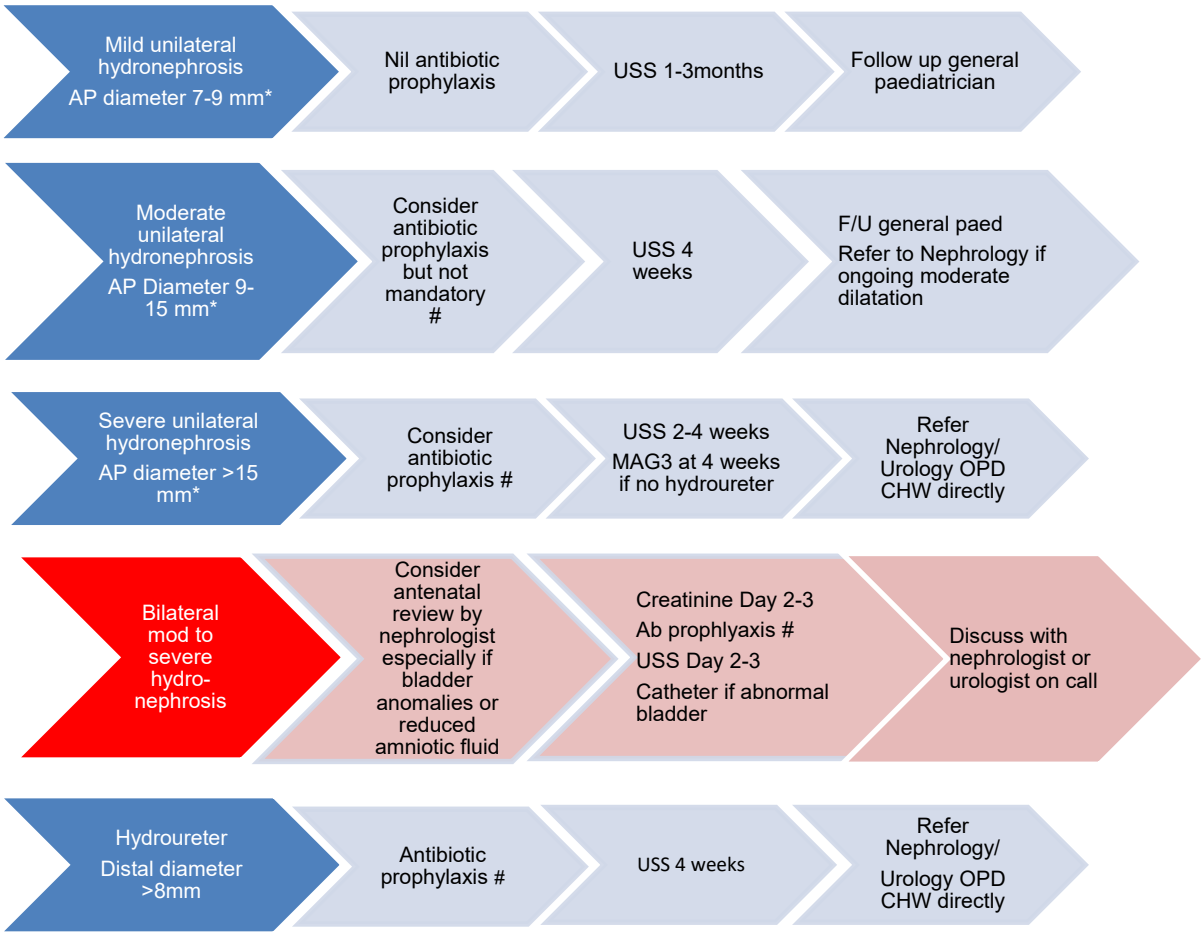
- AP diameter – anteroposterior diameter
- CAKUT – congenital abnormalities of the kidney and urinary tract
- USS – ultrasound
- OPD – outpatient department
- MCDK – multicystic dysplastic kidney

Procedure

CAKUT (congenital abnormalities of the kidney and urinary tract) are one of the most commonly diagnosed anomalies on prenatal ultrasound, present in 1-5% of pregnancies. CAKUT anomalies range from mild renal pyelectasis to severe renal dysplasia and the worst abnormalities will result in Potter's sequence. The prognosis and the management of the different renal abnormalities vary significantly, and the purpose of this guideline is to provide guidance for the referral of babies found to have renal abnormalities.

Given that the risk of urinary tract infection is often higher in babies with CAKUT, all families should be counselled about the signs and symptoms of UTI in infants and advised about where to seek appropriate management.

Figure 1: Management of antenatal hydronephrosis##



Notes

*on third trimester USS

Consider prophylaxis with:

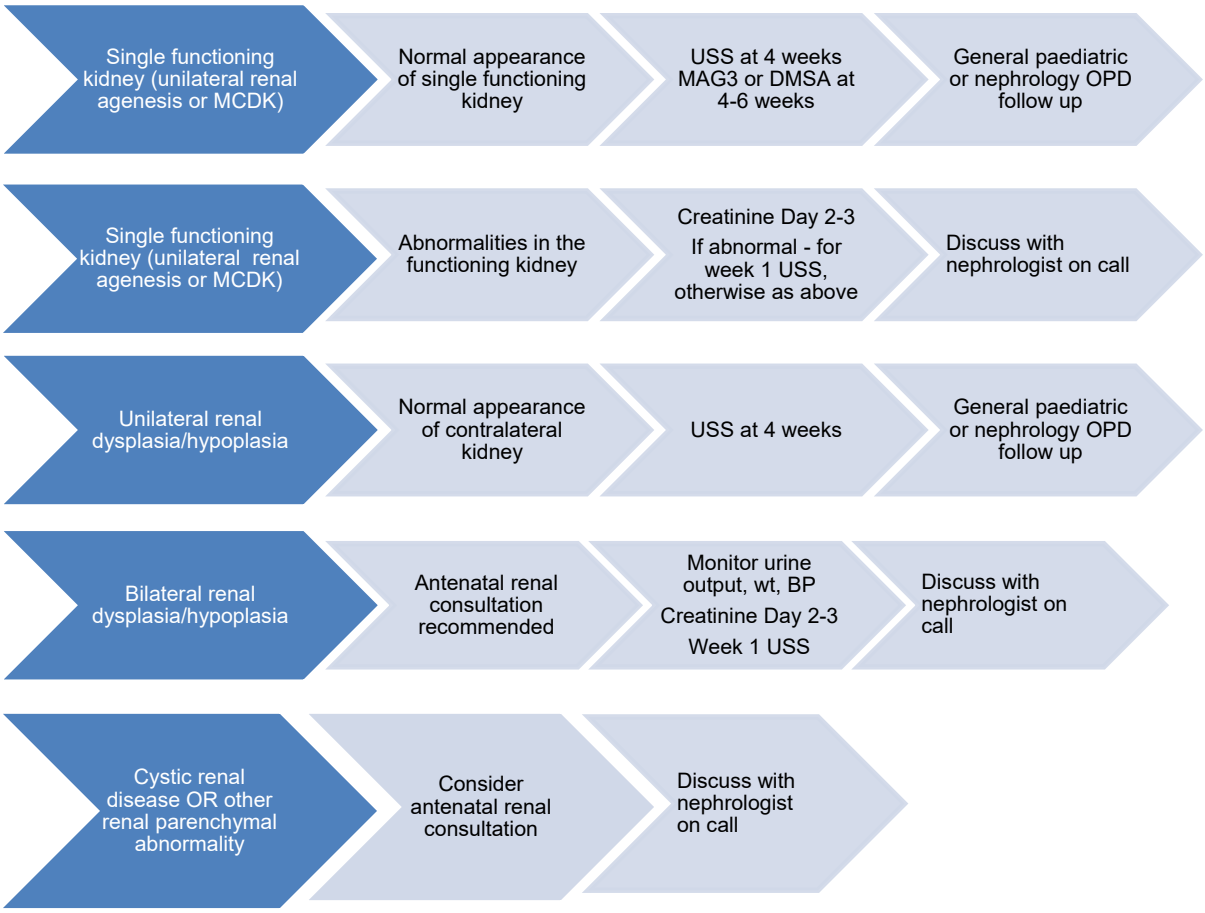
1st line: cefalexin (12.5 mg/kg/dose at night) at night before 2 months of age.

2nd line: Trimethoprim with sulfamethoxazole 2 mg/kg/dose (trimethoprim component) at night from 2 months of age.

Cefalexin is preferred due to risk of kernicterus in children <2 months of age

##CHW Switchboard: 02-98450000 for nephrologist/urologist on call

Figure 2: Management of renal dysplasia/hypoplasia[##]



Guidelines for antibiotic prophylaxis

Antibiotic Prophylaxis		
AGE	**Antibiotic**	**Dose**
Neonates (less than 28 days corrected gestational age)	**Cefalexin**	12.5mg/kg/dose orally at night (max dose:125mg)
Infants greater than 28 days	**Cefalexin**	12.5mg/kg/dose orally at night (max dose: 125mg)
OR		
Infants greater than **2 months** *Trimethoprim/Sulfamthoxazole should not be given to premature babies or to infants less than 2 months of age due to the risk of kernicterus.*	**Trimethoprim / Sulfamthoxazole**	2mg/kg/dose orally at night (dose based on trimethoprim) (max dose: 160 mg trimethoprim/800 mg sulfamethoxazole)

Prophylaxis around Micturating Cystourethrogram (MCUG)		
AGE	**Antibiotic**	**Dose**
Neonates (less than 28 days corrected gestational age)	**Cefalexin**	12.5 mg/kg/dose 8-hourly for 3 days (day prior, on the day and one day after MCU) (max dose: 125mg)
Infants greater than 28 days	**Cefalexin**	12.5 mg/kg/dose 8-hourly for 3 days (day prior, on the day and one day after MCU) (max dose: 125mg)
OR		
Infants greater than **2 months** *Trimethoprim/Sulfamthoxazole should not be given to premature babies or to infants less than 2 months of age due to the risk of kernicterus.*	**Trimethoprim / Sulfamthoxazole**	2mg/kg/dose orally twice a day for 3 days (day prior, on the day and one day after MCU) (dose based on trimethoprim) (max dose: 80 mg trimethoprim/400 mg sulfamethoxazole)

Educational Notes

Antenatal hydronephrosis

Hydronephrosis is the most common renal tract abnormality detected antenatally, with post-natal pathology identified in approximately 36%. The degree of hydronephrosis, as characterised by the maximal anterior posterior (AP) diameter of the renal pelvis predicts the likelihood of finding pathology on subsequent investigation (Table 1)[1]. The most common pathologies identified are pelvic-ureteric junction (PUJ) obstruction and vesicoureteric reflux (VUR). Other less common pathologies identified include posterior urethral valves (PUV) and ureteral obstruction.

There are several systems in use for the classification of antenatal hydronephrosis. The most commonly utilised is based solely on the renal AP diameter and is summarised in Table 1. This classification system does not take into account renal parenchymal changes, such as thinning or increased echogenicity, and consequently alternative classification systems have been created including that used by the Society of Fetal Urology[2]. Significant pathology is more commonly identified in those with moderate or severe hydronephrosis.

Urinary tract infection is more common with high grade hydronephrosis and prophylactic antibiotics in these infants may be beneficial.

Degree of APD	Second Trimester	Third Trimester	Pathology identified
Mild	4 to 7 mm	7 to 9 mm	11.9% (4.5 to 28.0)
Moderate	7 to 10 mm	9 to 15 mm	45.1% (25.3 to 66.6)
Severe	≥10mm	≥15mm	88.3 (53.7 to 98.0)

Table 1. Correlation of renal AP diameter in second and third trimester with significant postnatal uro-nephrological pathology[1].

Hydroureter

Hydroureter is usually identified antenatally with associated hydronephrosis and renal pelvic dilatation, but it can be a solitary finding. Hydroureter suggests the possibility of VUR (most commonly), vesicoureteric junction obstruction or a non-refluxing, non-obstructive megaureter. If bilateral, the bladder should be fully assessed on USS to look for evidence of trabeculation and in male infants posterior urethral valves should be excluded by MCUG. There is a higher risk for urinary tract infections and therefore prophylactic antibiotics should be considered[3].

Multicystic dysplastic kidney and single kidney

Multicystic dysplastic kidney (MCDK) is commonly seen as multiple non-communicating cysts of variable size. MCKD kidneys are non-functioning and may be palpable at birth but some will have already started to involute at birth and may be smaller than expected. There is a high rate of spontaneous involution with complete atrophy in 35% at two years, 47% at five years and 62% at 10 years.[4] There is often, but not always, associated compensatory hypertrophy of the remaining kidney. Up to 20% of children have associated reflux in the contralateral kidney[5] but in general there is no increase in the rates of UTI or renal scarring.

Consequently, MCUG is not routinely performed and antibiotic prophylaxis is not recommended, unless there is abnormal ultrasound imaging of the contralateral kidney.

Unilateral renal agenesis may also be diagnosed on antenatal ultrasound. There is typically compensatory hypertrophy of the remaining kidney and often mild renal pelvic dilatation. Consider doing a DMSA or MAG3, as sometimes an ectopic kidney may be present in the pelvis., that has not been detected on USS.

Children with either MCDK or unilateral renal agenesis with no associated hydronephrosis in the contralateral kidney are followed up with serial ultrasound through infancy (6, 12 and 24 months) to ensure renal growth of the normal kidney and can be referred non-urgently to the Nephrology Department for follow up.

The concomitant finding of hydronephrosis in a child with single functioning kidney warrants serum creatinine check prior to discharge from hospital and discussion with the Nephrology Team to facilitate early review.

Renal dysplasia/hypoplasia

Renal dysplasia/hypoplasia is occasionally seen associated with antenatal hydronephrosis. All children with diagnosed bilateral renal dysplasia/hypoplasia should have renal function checked on day 3 of life. If renal function is normal, patients can be referred non-urgently to Nephrology outpatients. If the creatinine is abnormal, the patient should be discussed with the Nephrology Team.

Cystic renal disease

Renal cysts are not a common finding on ultrasound in children. A single cyst may not be significant and simply warrants ultrasound surveillance. Multiple cysts are pathogenic and may be due to renal dysplasia or one of the many cystic renal disorders.

Enlarged echogenic kidneys diagnosed antenatally are suspicious for autosomal recessive polycystic kidney disease (ARPKD). Discrete cysts may or may not be seen. There is a highly variable course of ARPKD with significant antenatal and perinatal mortality but others may not progress to end stage renal disease until adulthood. Antenatal paediatric nephrology consultation should be offered to all parents with a foetus with suspected ARPKD. Infantile nephronophthisis and other genetic conditions can also present with echogenic kidneys antenatally and again paediatric nephrology antenatal counselling should be offered. These infants will also require renal genetics follow up (will be arranged by the nephrologist involved).

Autosomal dominant polycystic kidney disease (ADPKD) causes end stage kidney disease, usually in mid adult life. Presentation can occur in the neonatal and infant period, typically with bilateral renal cysts but usually normal sized kidneys in the early stages.

**Neonates with a parent with ADPKD should not undergo routine renal ultrasound without prior consultation by a paediatric nephrologist, as there are ethical considerations regarding the diagnosis of a condition in childhood that cannot be treated, but will not have impact until adulthood*.

References

1. Lee RS, Cendron M, Kinnamon DD, Nguyen HT. Antenatal Hydronephrosis as a Predictor of Postnatal Outcome: A Meta-analysis. Pediatrics. 2006;118(2):586-93.
2. Fernbach SK, Maizels M, Conway JJ. Ultrasound grading of hydronephrosis: introduction to the system used by the Society for Fetal Urology. Pediatr Radiol. 1993;23(6):478-80.
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4. Hayes WN, Watson AR. Unilateral multicystic dysplastic kidney: does initial size matter? Pediatr Nephrol. 2012;27(8):1335-40.
5. Ismaili K, Avni FE, Alexander M, Schulman C, Collier F, Hall M. Routine voiding cystourethrography is of no value in neonates with unilateral multicystic dysplastic kidney. J Pediatr. 2005;146(6):759-63.