NENC LMNS Clinical Guideline.

Guideline for the investigation and management of congenital abnormalities of the kidneys and renal tract

Title	Clinical Guideline for the investigation and management of congenital abnormalities of the kidneys and renal tract
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Clinical Expert Group	Fetal Medicine
Applicable to:	All Trusts in the NENC LMNS geographical area: This guideline is for all maternity staff: Obstetric consultants, all other obstetric doctors, Midwives and, Ultrasonographers.
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Document owner	North East and North Cumbria (NENC) Local Maternity and Neonatal System (LMNS) Fetal Medicine Group.

**Although the term "women" and new "mums/mothers" is used throughout this document, we recognise that not all birthing people identify as such.*

Introduction

Renal tract abnormalities are among the commonest congenital malformations and affect around 1.6 per 1000 births. They can be diagnosed antenatally as part of routine antenatal care or diagnosed postnatally for example following urinary tract infection or as an incidental finding.

The majority do not cause adverse complications in childhood but some patients can experience recurrent infections and also long term problems including hypertension, proteinuria and renal impairment.

The role for antibiotic prophylaxis is often discussed due to the increased risk of urinary tract infection with some abnormalities of the renal tract. When required, this can be either trimethoprim or cephalexin as per the children's BNF.

This is a guideline contains flowcharts for the initial management and investigation of the following common conditions;

Unilateral hydronephrosis

Bilateral hydronephrosis

Suspected lower urinary tract obstruction (LUTO)

Unilateral renal agenesis

MCDK

Ectopic/ pelvic kidney

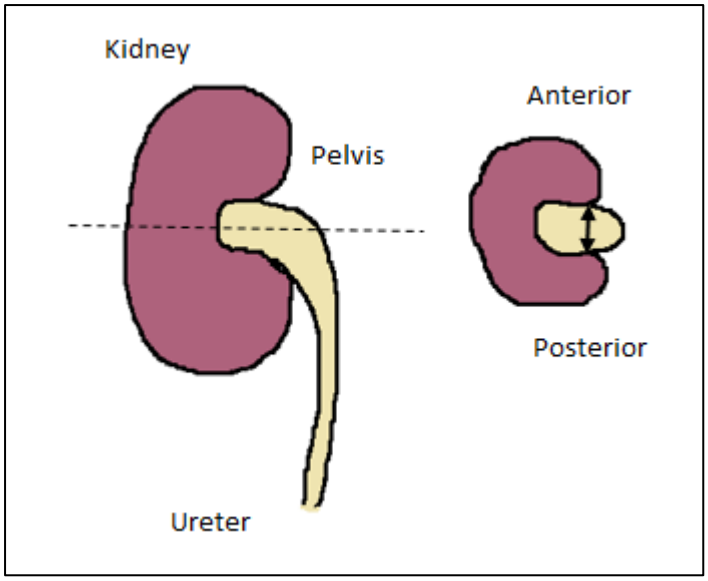
Duplex kidney

Horseshoe Kidney

These are not exhaustive guidelines and if there are any questions or concerns during the patient's initial investigations then please discuss with the paediatric nephrology or urology team at GNCH, Newcastle Upon Tyne Hospital Trust.

Hydronephrosis

Hydronephrosis describes a dilation of the collecting system within the kidney and is commonly defined as an AP diameter >9mm at any time after 30 week gestation. It is a common finding on antenatal scans and in the majority of cases, it is not due to any underlying abnormality and spontaneously improves. As the degree of hydronephrosis worsens, the likelihood of an underlying cause increases and so the intensity of postnatal investigation and management is based around the hydronephrosis severity. This is reported on US scan as the antero-posterior diameter (APD).



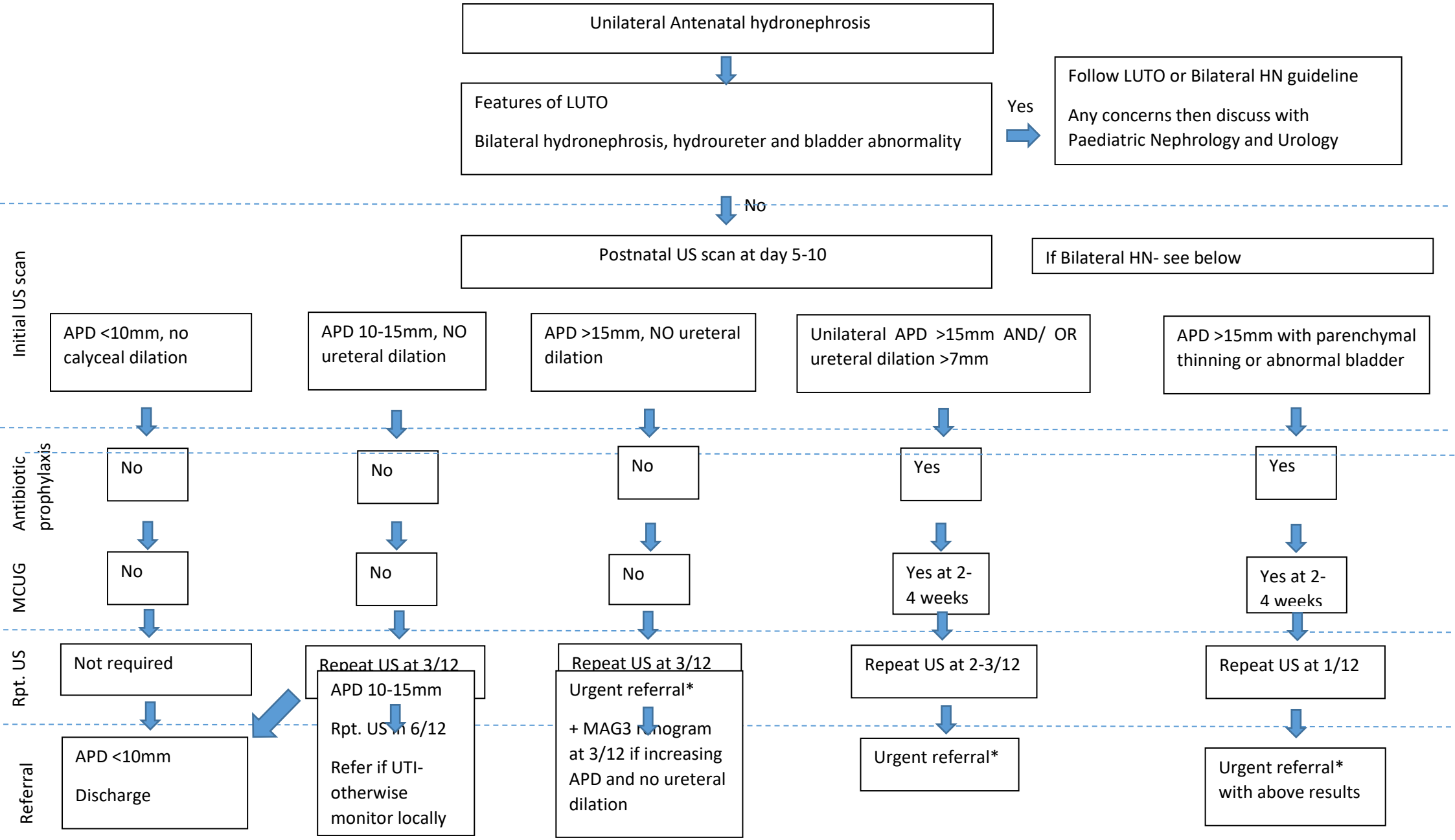
Common causes of Antenatal hydronephrosis	Prevalence
Transient- resolves on postnatal scans	50%
Hydronephrosis with no evidence of obstruction; or extra-renal pelvis	15%
PUJ obstruction	11%
Vesico-Ureteric Reflux	9%
Megaureter	4%
Dysplasia	3%
MCDK	2%
Duplex kidney	2%
PUV	1%

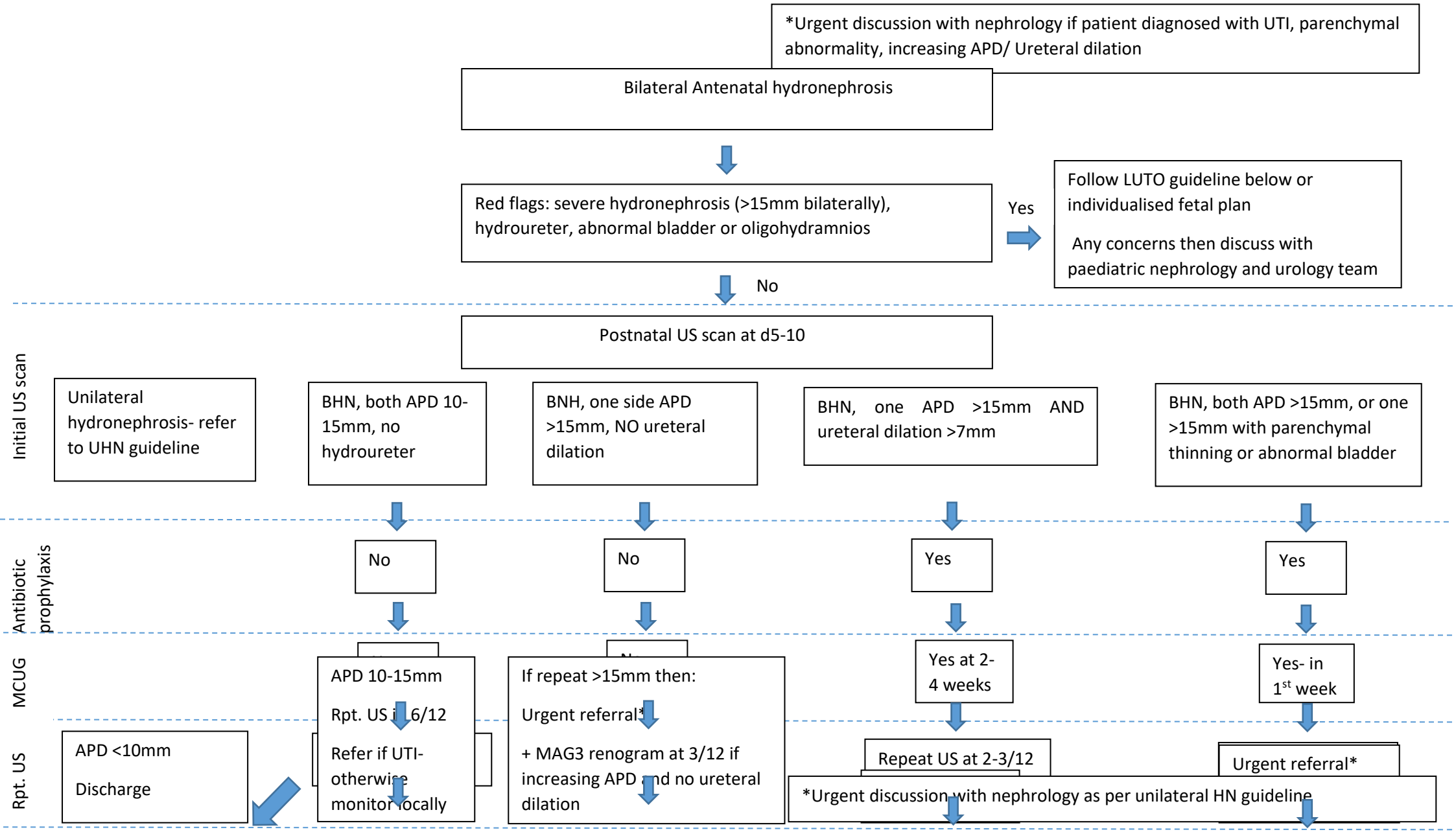
Prophylactic antibiotics

Prophylactic antibiotics are recommended from birth for any patient with:

- Antenatal APD >15mm (Either unilateral or bilateral)
- Ureteric dilation or bladder abnormality
- Any hydronephrosis associated with other renal parenchymal abnormality

The antibiotics can then be reviewed and, if appropriate, stopped following the postnatal scan as per the guidelines below


Unilateral Antenatal hydronephrosis
Features of LUTO
Bilateral hydronephrosis, hydroureter and bladder abnormality
Yes
Follow LUTO or Bilateral HN guideline
Any concerns then discuss with Paediatric Nephrology and Urology
No
Postnatal US scan at day 5-10
If Bilateral HN- see below
Initial US scan
APD <10mm, no calyceal dilation
APD 10-15mm, NO ureteral dilation
APD >15mm, NO ureteral dilation
Unilateral APD >15mm AND/ OR ureteral dilation >7mm
APD >15mm with parenchymal thinning or abnormal bladder
Antibiotic prophylaxis
No
No
No
Yes
Yes
MCUG
No
No
No
Yes at 2-4 weeks
Yes at 2-4 weeks
Rpt. US
Not required
Repeat US at 3/12
Repeat US at 3/12
Repeat US at 2-3/12
Repeat US at 1/12
Referral
APD <10mm
Discharge
APD 10-15mm
Rpt. US in 6/12
Refer if UTI- otherwise monitor locally
Urgent referral*
+ MAG3 renogram at 3/12 if increasing APD and no ureteral dilation
Urgent referral*
Urgent referral* with above results


*Urgent discussion with nephrology if patient diagnosed with UTI, parenchymal abnormality, increasing APD/ Ureteral dilation
Bilateral Antenatal hydronephrosis
Red flags: severe hydronephrosis (>15mm bilaterally), hydroureter, abnormal bladder or oligohydramnios
Yes
Follow LUTO guideline below or individualised fetal plan
Any concerns then discuss with paediatric nephrology and urology team
No
Postnatal US scan at d5-10
Initial US scan
Unilateral hydronephrosis- refer to UHN guideline
BHN, both APD 10-15mm, no hydroureter
BNH, one side APD >15mm, NO ureteral dilation
BHN, one APD >15mm AND ureteral dilation >7mm
BHN, both APD >15mm, or one >15mm with parenchymal thinning or abnormal bladder
Antibiotic prophylaxis
No
No
Yes
Yes
MCUG
APD 10-15mm
Rpt. US in 6/12
Refer if UTI- otherwise monitor locally
If repeat >15mm then:
Urgent referral*
+ MAG3 renogram at 3/12 if increasing APD and no ureteral dilation
Yes at 2-4 weeks
Yes- in 1st week
Rpt. US
APD <10mm
Discharge
Repeat US at 2-3/12
Urgent referral*
*Urgent discussion with nephrology as per unilateral HN guideline

Features of LUTO:

Bilateral hydronephrosis (>15mm)/ hydroureter/ abnormal bladder

+/- Megacystis

Delivery should be at RVI Hospital

Is there an individual plan in the notes?

Yes

Follow individual plan

No

Initial management

Early assessment by neonatal team and consideration for urethral catheter.

Initially for 12hourly U&Es- discuss with on call Nephrology consultant if any concerns

Antibiotic prophylaxis

Yes

US scan

Urgent US scan at 24hours- discuss results with Paediatric Urology and Nephrology

MCUG

Urgent MCUG- within first 72hours

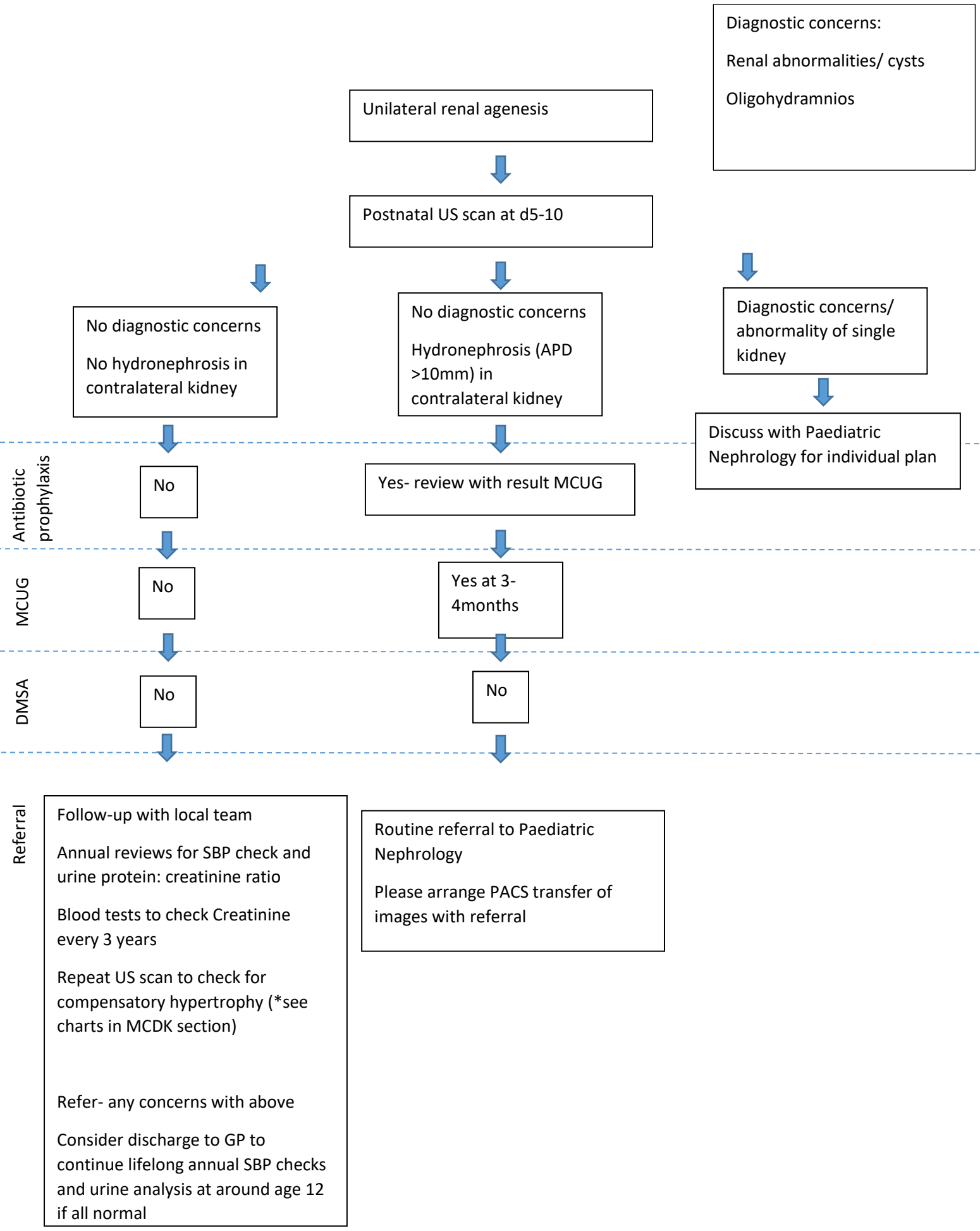
NB. This guideline is for the management of the renal aspect of LUTO. Management may be changed if significant respiratory concerns

Unilateral renal agenesis

Unilateral renal agenesis (URA) occurs when one kidney fails to develop. This can be in isolation or may be part of a spectrum of other congenital abnormalities such as VACTERL and so patients may require further investigation for associated abnormalities.

It is quite common and is estimated to occur in 1:1000 live births. The majority are not thought to be caused by any genetic mutations and occur spontaneously.

The majority of children with this condition do not suffer from any complications but they are at increased risk of high blood pressure and also renal impairment especially if the contralateral kidney is also not normal. For this reason, we advise life-long follow-up and monitoring


Diagnostic concerns:
Renal abnormalities/ cysts
Oligohydramnios
Unilateral renal agenesis
Postnatal US scan at d5-10
No diagnostic concerns
No hydronephrosis in contralateral kidney
No diagnostic concerns
Hydronephrosis (APD >10mm) in contralateral kidney
Diagnostic concerns/ abnormality of single kidney
Discuss with Paediatric Nephrology for individual plan
Antibiotic prophylaxis
No
Yes- review with result MCUG
MCUG
No
Yes at 3-4months
DMSA
No
No
Referral
Follow-up with local team
Annual reviews for SBP check and urine protein: creatinine ratio
Blood tests to check Creatinine every 3 years
Repeat US scan to check for compensatory hypertrophy (*see charts in MCDK section)
Refer- any concerns with above
Consider discharge to GP to continue lifelong annual SBP checks and urine analysis at around age 12 if all normal
Routine referral to Paediatric Nephrology
Please arrange PACS transfer of images with referral

MCDK- Multi-cystic dysplastic kidney

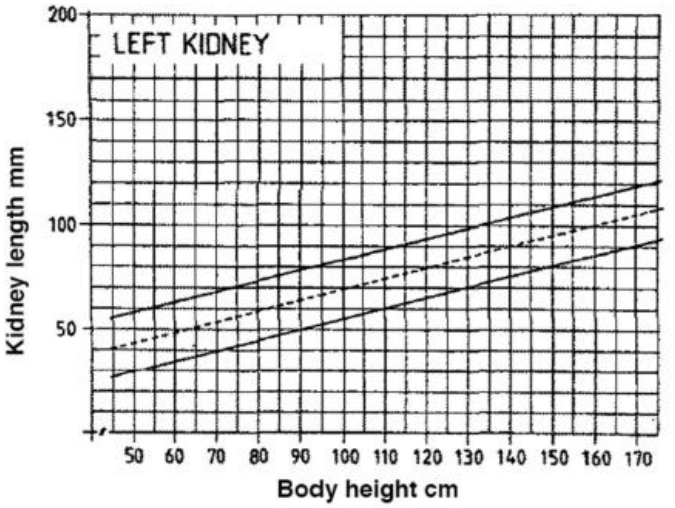
This is a non-functioning kidney that has not developed properly. Instead of normal kidney tissue, there is just a collection of cysts; fluid filled sacs. These can be quite large at birth but normally shrink during the first few years of life.

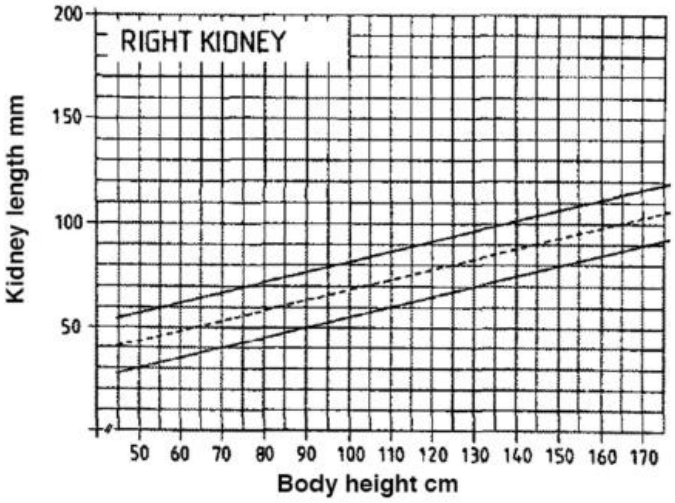
The majority of people with this condition are not affected by it but some may experience:

- High blood pressure
- Urinary tract infections
- Reduced kidney function

These complications are normally caused by the other kidney also not being normal.

The postnatal scans are aimed to confirm the diagnosis and to confirm the contralateral kidney is normal and demonstrates compensatory hypertrophy (increased growth to compensate for single kidney- see charts below for normal kidney size with increasing height)





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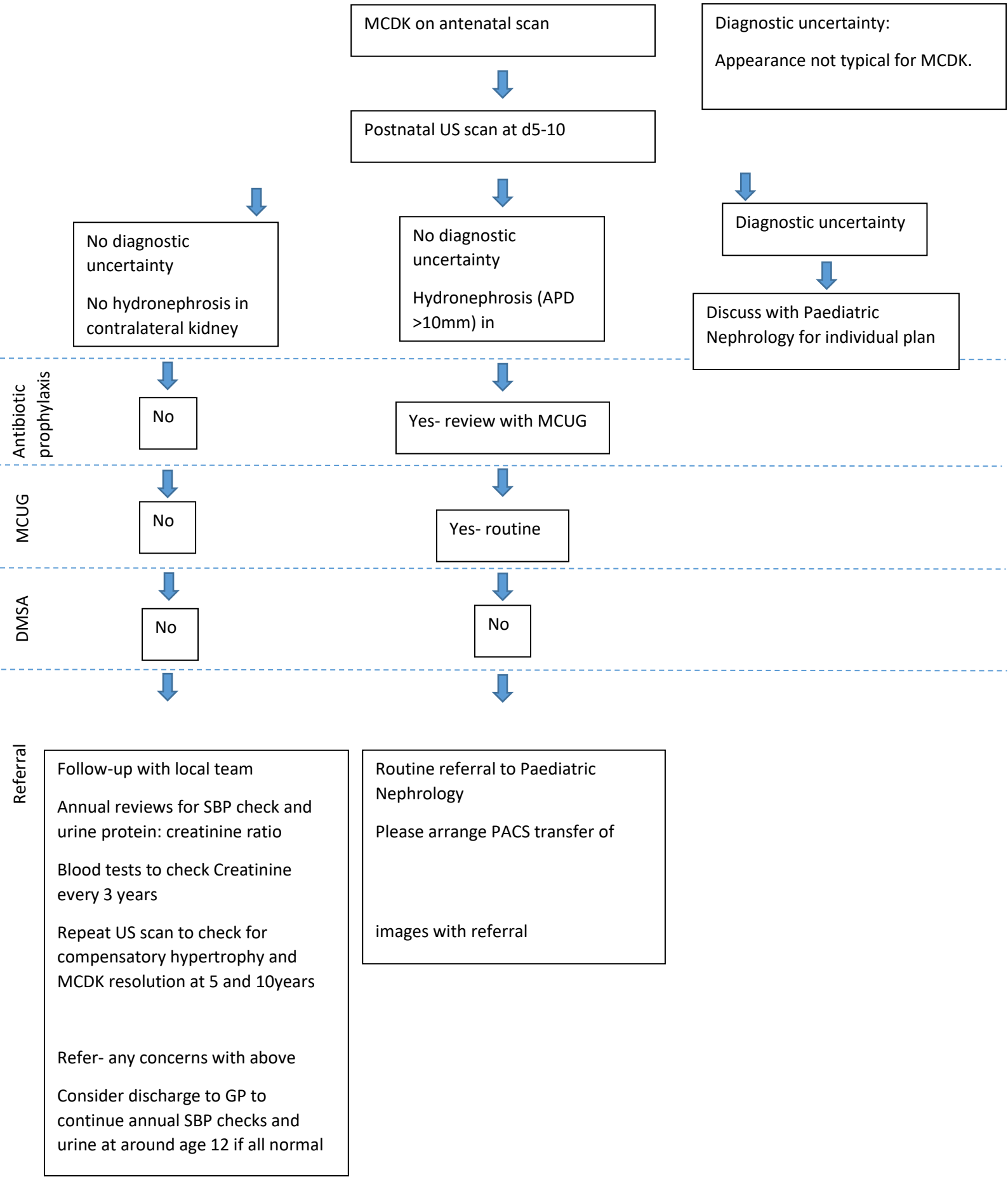

MCDK on antenatal scan
Postnatal US scan at d5-10
Diagnostic uncertainty:
Appearance not typical for MCDK.
No diagnostic uncertainty
No hydronephrosis in contralateral kidney
No diagnostic uncertainty
Hydronephrosis (APD >10mm) in
Diagnostic uncertainty
Discuss with Paediatric Nephrology for individual plan
Antibiotic prophylaxis
No
Yes- review with MCUG
MCUG
No
Yes- routine
DMSA
No
No
Referral
Follow-up with local team
Annual reviews for SBP check and urine protein: creatinine ratio
Blood tests to check Creatinine every 3 years
Repeat US scan to check for compensatory hypertrophy and MCDK resolution at 5 and 10years
Refer- any concerns with above
Consider discharge to GP to continue annual SBP checks and urine at around age 12 if all normal
Routine referral to Paediatric Nephrology
Please arrange PACS transfer of
images with referral

Duplex Kidney

A duplex kidney is a kidney that has not developed properly and has two drainage parts to one kidney. It may also have two drainage tubes (ureters) coming out of the kidney and down into the bladder.

The majority of people with a duplex kidney are not affected by it but it can be complicated by

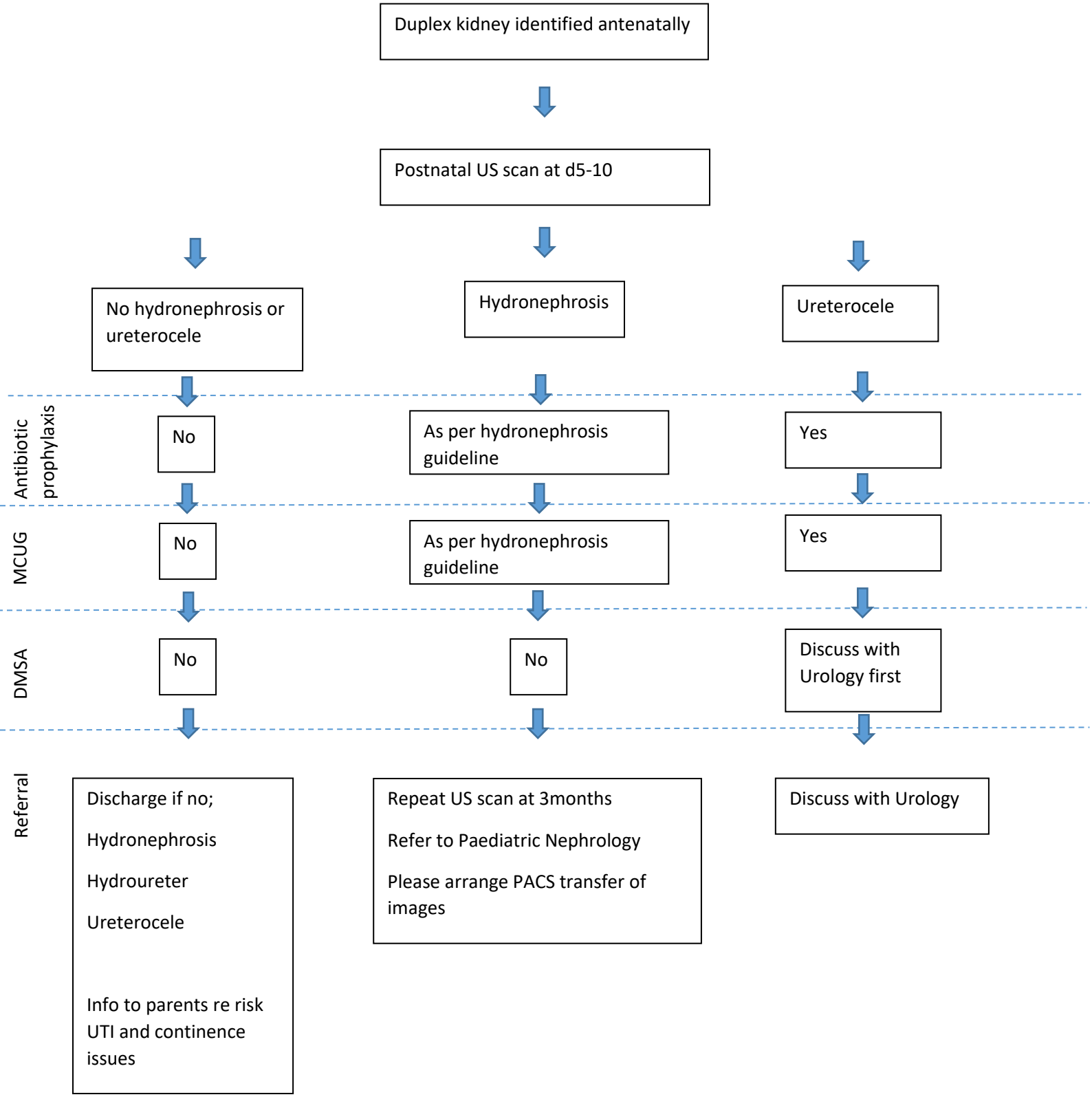
Vesicoureteric reflux- urine that travels back up from the bladder into the ureter and kidney. This typically affects the lower moiety.

Obstruction- this can be at the level of the PUJ or in association with an ureterocele (upper moiety)

Continence issues related to ectopic insertion of a ureter

Further information on duplex kidneys can be found on the infokid website:

https://www.infokid.org.uk/duplex-kidneys


Duplex kidney identified antenatally
Postnatal US scan at d5-10
No hydronephrosis or ureterocele
Hydronephrosis
Ureterocele
Antibiotic prophylaxis
No
As per hydronephrosis guideline
Yes
MCUG
No
As per hydronephrosis guideline
Yes
DMSA
No
No
Discuss with Urology first
Referral
Discharge if no;
Hydronephrosis
Hydroureter
Ureterocele
Info to parents re risk UTI and continence issues
Repeat US scan at 3months
Refer to Paediatric Nephrology
Please arrange PACS transfer of images
Discuss with Urology

Ectopic/ Pelvic kidney

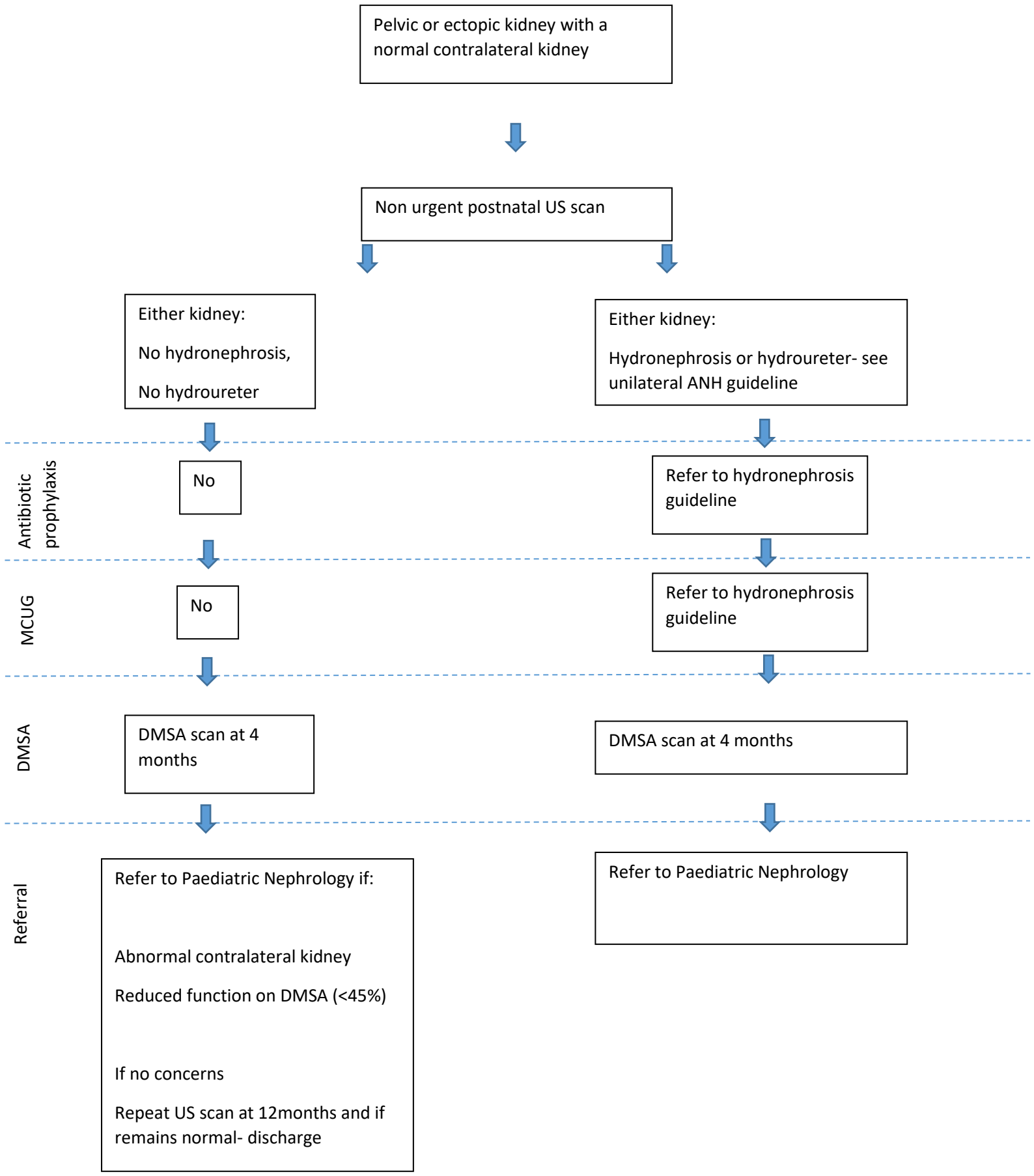
This is caused by failure of the normal ascent of the kidney during embryogenesis. The abnormal kidney can be anywhere along the path of normal embryological ascent but pelvic kidneys are the most common. It is estimated to occur in 1:1000-5000 live births.

The most common complications are

Renal hypoplasia/ dysplasia of the ectopic kidney

Obstruction

The aim of the follow-up scans is to identify those at risk of these complications and determine who needs ongoing follow-up throughout their childhood.


Pelvic or ectopic kidney with a normal contralateral kidney
Non urgent postnatal US scan
Either kidney:
No hydronephrosis,
No hydroureter
Either kidney:
Hydronephrosis or hydroureter- see unilateral ANH guideline
Antibiotic prophylaxis
No
Refer to hydronephrosis guideline
MCUG
No
Refer to hydronephrosis guideline
DMSA
DMSA scan at 4 months
DMSA scan at 4 months
Referral
Refer to Paediatric Nephrology if:
Abnormal contralateral kidney
Reduced function on DMSA (<45%)
If no concerns
Repeat US scan at 12months and if remains normal- discharge
Refer to Paediatric Nephrology

Horseshoe kidney

This is the commonest fusion abnormality of the kidneys with an estimated incidence of 1:400 and is more common in males. Both kidneys typically fuse at the lower pole but fusion at the upper pole is possible though much less common.

Importantly, this can be associated with genetic diseases such as Turner's syndrome and Down syndrome as well as other congenital abnormalities such as VACTERL.

The main renal complications are;

Obstruction- typically PUJO

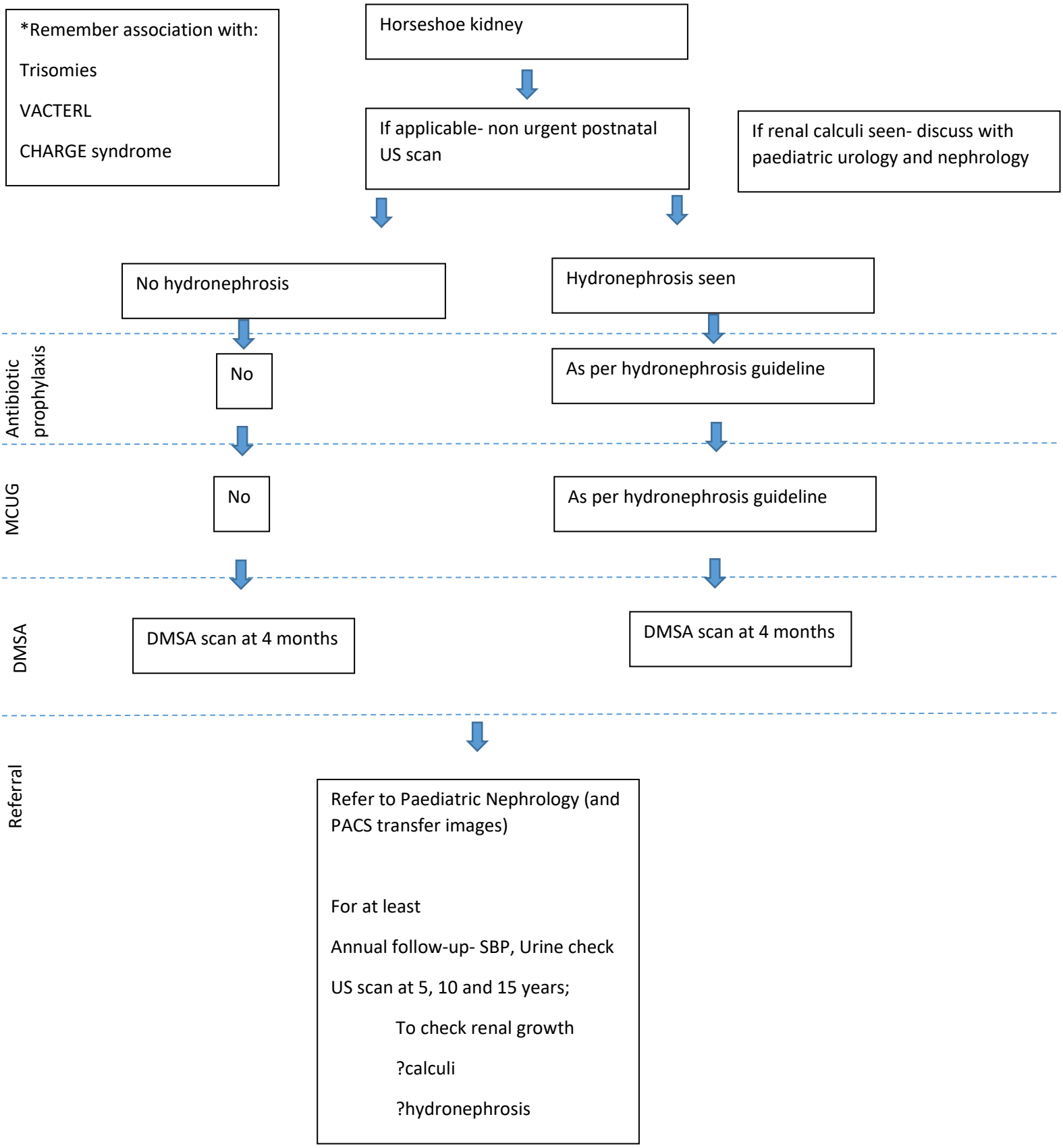
Infection

Calculi

The lifetime risk of these complications has been reported to be high (26%, 19%, 25% respectively by Kang et al (2021) and during childhood you also can see increased rates of hypertension and proteinuria ((Yavuz et al (2015). It is important to arrange follow-up and to give a follow-up plan for the patient and their GP to follow at the time of their discharge from paediatric services.

Further information on horseshoe kidneys can be found at:

https://www.infokid.org.uk/horseshoe-kidney


*Remember association with:
Trisomies
VACTERL
CHARGE syndrome
Horseshoe kidney
If applicable- non urgent postnatal US scan
If renal calculi seen- discuss with paediatric urology and nephrology
No hydronephrosis
Hydronephrosis seen
Antibiotic prophylaxis
No
As per hydronephrosis guideline
MCUG
No
As per hydronephrosis guideline
DMSA
DMSA scan at 4 months
DMSA scan at 4 months
Referral
Refer to Paediatric Nephrology (and PACS transfer images)
For at least
Annual follow-up- SBP, Urine check
US scan at 5, 10 and 15 years;
To check renal growth
?calculi
?hydronephrosis

Addendum for DGH hospitals with potential challenges to accessing urgent paediatric USS services. Pragmatic application of the guideline should be applied for the local provider trust and process through their local governance process. For lower risk renal abnormalities scans up to 2-4 weeks may be more appropriate (BMUS guidance). This includes renal agenesis with no structural concerns with the contralateral kidney, Isolated unilateral renal pelvis dilatation with no suspicious renal features, Antenatally suspected congenital renal anomaly e.g. duplex kidney with no hydronephrosis and Unilateral multicystic kidney disease with normal contralateral kidney.

Suspicious features

- *Calyceal dilatation*
- *Ureteric dilatation*
- *Solitary kidney*
- *Abnormality of the renal parenchyma e.g. abnormal echogenicity, cysts*.
- *Abnormality of the urinary bladder e.g. dilated, thick walled, ureterocele*
- *Abnormal volume of amniotic fluids"*

References

BMUS guidance :
https://www.bmus.org/media/resources/files/Guidelines_for_postnatal_US_referrals_for_followup_of_antenatal_detected_ZV5fDDb.pdf

Hydronephrosis

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Duplex Kidney

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Horseshoe Kidney

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