Renal – Antenatal Urinary Tract Abnormalities

Summary

This guideline refers to the management of antenatal urinary tract abnormalities (AUTA). Antenatally detected renal abnormalities are common. It is important that the most appropriate investigation is performed at the most appropriate time and that parents are fully informed and offered appropriate follow up.

Principle pathologies:

- Multicystic kidney
- PUJ obstruction
- UV obstruction +/- reflux
- 1° VU reflux
- Duplex ureters
- Ureterocele
- Neurogenic bladder (rarely seen at birth)
- Associated with other anomaly e.g. anorectal malformation

Important points

1. Except in the presence of posterior urethral valves or gross dilatation due to reflux, prophylactic antibiotics are not indicated. However if an MCUG is to be undertaken then the infant should be on full antibiotic course for 48 hours before the procedure to reduce the risk of MCUG related infection. There is not complete consensus on the use of antibiotics. Some specialists recommend that “antibiotic prophylaxis (Trimethoprim 2 mg/kg nocte) should be started on all infants from birth. If there is no abnormality detected by the 2/52 US scan then these can be stopped. If however dilatation returns and an MCUG is planned then they should be restarted. Prior to an MCUG a full treatment dose (4 mg/kg bd) should be given 48 hours before the procedure”. It is recommended that units discuss with their referral centre and establish the locally recommended practice.

2. The majority of conditions are associated with a good outcome and follow up should be arranged through appropriate clinics. More than half of the antenatally diagnosed urinary tract abnormalities are transient. However some severe conditions may be associated with a significant risk of renal damage or risk to the infant and urgent surgical referral may be indicated.

3. All urgent scans and subsequent scans should be requested using appropriate request forms. All forms should include as much antenatal information as possible. They should also include, if available, parental telephone numbers. The information should be clear and legible.

4. If any investigations are organized as an out patient it is very important that a mechanism is in place to make sure that urgent investigations are performed within an appropriate time scale and that results are reviewed as soon as they become available.

5. For details of the principal pathologies please see the glossary.

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Antenatally Detected Renal Anomalies – boys

Remember that on occasions unilateral dilatation may indicate outlet problems such as urethral valves, and that gross renal dilatation may be mistaken for a cystic kidney. A normal stream of urine may be passed by a boy with PU valves.

Antenatally Detected Renal Anomalies - girls

Remember that on occasions unilateral dilatation may indicate outlet problems though less commonly than in boys, and that gross renal dilatation may be mistaken for a cystic kidney.

Section 1: Antenatal renal pelvis dilatation – Unilateral

Unilateral assumes that the contra lateral kidney is normal

On occasions unilateral dilatation may indicate outlet problems such as urethral valves and that gross dilatation may be mistaken for a cystic kidney. A normal stream of urine may be passed by a boy with PU valves. Bladder wall thickness may be difficult to determine unless the bladder is at least half full.

<table>
<thead>
<tr>
<th>Abnormality</th>
<th>Investigations</th>
<th>Follow up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isolated renal dilatation</td>
<td>U/S at 2/52</td>
<td>Observe to 1 year, U/S 3 monthly</td>
</tr>
<tr>
<td></td>
<td>A/P diameter &lt; 10mm</td>
<td>Refer to urologist</td>
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<tr>
<td></td>
<td>&gt; 10mm</td>
<td></td>
</tr>
<tr>
<td>With dilated ureter</td>
<td>U/S at birth (within 3 days) and 2 weeks</td>
<td><em>May need MCUG if PU valves suspected</em></td>
</tr>
<tr>
<td></td>
<td>A/P diameter &lt; 10 mm</td>
<td>Neonatal renal/follow up clinic; US at 6/52 and 3/12; further investigations after NU conference review</td>
</tr>
<tr>
<td></td>
<td>&gt; 10mm</td>
<td>Refer to urologist</td>
</tr>
<tr>
<td>With thick walled bladder</td>
<td>Urgent U/S and MCUG after discussion with Urology</td>
<td>Refer to urologist</td>
</tr>
<tr>
<td>Evidence of ureterocele or duplex</td>
<td>U/S at 2 weeks</td>
<td>Refer to urologist</td>
</tr>
<tr>
<td>MCDK</td>
<td>U/S at 2 weeks, DMSA at 3 months</td>
<td>Refer to nephrologist and inform tertiary renal centre</td>
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<tr>
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<tr>
<td>Isolated bilateral renal dilatation</td>
<td>U/S at birth (within 2-3 days) and at 2/52 &lt;br&gt; A/P diameter &lt; 10mm &lt;br&gt; &gt; 10mm</td>
<td>Neonatal renal/follow up clinic; US at 6/52 and 3/12; further investigations after NU conference review &lt;br&gt; Refer to urologist</td>
</tr>
<tr>
<td>Bilateral dilatation with dilated ureter(s)</td>
<td>Urgent US (12-48 hours) and MCUG &lt;br&gt; Posterior urethral valves found &lt;br&gt; Posterior urethral valves not found &lt;br&gt; A/P diameter &lt; 10 mm &lt;br&gt; &gt; 10mm</td>
<td>Refer to urologist &lt;br&gt; Neonatal renal/follow up clinic; US at 6/52 and 3/12; further investigations after NU conference review &lt;br&gt; Refer to urologist</td>
</tr>
<tr>
<td>With thick walled bladder (rare in girls)</td>
<td>Urgent U/S and MCUG after discussion with Urology (± suprapubic catheter)</td>
<td>Refer to urologist</td>
</tr>
<tr>
<td>Evidence of ureterocele or duplex</td>
<td>U/S at birth (within 3 days) and at 2 weeks</td>
<td>Refer to urologist</td>
</tr>
<tr>
<td>MCDK</td>
<td>U/S at 2 weeks, DMSA at 3 months</td>
<td>Refer to nephrologist and inform tertiary renal centre &lt;br&gt; Note – bilateral MCDK is usually incompatible with life</td>
</tr>
<tr>
<td>MCDK with other findings</td>
<td>Manage as indicated above for the other findings</td>
<td>Refer to urologist and nephrologist and inform tertiary renal centre</td>
</tr>
</tbody>
</table>
### Section 3: Other antenatal abnormalities

Antibiotic prophylaxis is not normally indicated for any of the following renal abnormalities provided there is no antenatal renal pelvis dilatation.

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<tr>
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<tr>
<td>Crossed fused ectopia</td>
<td>U/S at 2/52</td>
<td>Observe to 1 year; US at 3/12 intervals. Neonatal renal follow up clinic</td>
</tr>
<tr>
<td></td>
<td>A/P diameter &lt; 10mm</td>
<td>Refer to urologist</td>
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<tr>
<td></td>
<td>&gt; 10mm</td>
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<tr>
<td>Horseshoe kidney</td>
<td>U/S at 2/52</td>
<td>Neonatal renal follow up clinic. US at 3/12. Refer to urologist</td>
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<td></td>
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<tr>
<td></td>
<td>&gt; 10mm</td>
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</tr>
<tr>
<td>Pelvic kidney</td>
<td>U/S at 2/52</td>
<td>Neonatal renal follow up clinic. US at 3/12 and 1 year.</td>
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</tbody>
</table>

### Section 4: Family history of renal abnormalities

If the antenatal scan was normal then in the majority of cases a single US at 6 weeks of age and renal clinic review is all that is necessary. However when there is a history of vesicoureteric reflux then an MCUG should be arranged at 3 months.

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<tr>
<td>First degree relative with VUR</td>
<td>MCUG and U/S at 3 months with antibiotic cover</td>
<td>Neonatal renal/follow-up clinic to discuss results</td>
</tr>
<tr>
<td>Any other family history of renal abnormalities</td>
<td>U/S at 6 weeks</td>
<td>Neonatal renal/follow-up clinic to discuss results</td>
</tr>
</tbody>
</table>

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Commonest pathologies

1. Antenatal dilation of the renal pelves

Antenatally diagnosed renal tract abnormalities are a common antenatal finding, present in 1:100-1:300 pregnancies. The vast majority of abnormalities are unilateral and are benign in nature. More than 50% of antenatal abnormalities are transient and resolve on postnatal scanning. Good communication is particularly important and where relevant neonatologists, paediatric nephrologist and paediatric urologists should be able to speak to parents before delivery.

To parents the antenatal diagnosis of urinary tract abnormality is often a source of disproportionate anxiety during pregnancy. The vast majority of infants are outwardly normal and healthy at birth; only a small number develop serious problems requiring surgery or lifelong follow up.

A normal ‘anomaly’ scan at 20 weeks does not exclude a renal abnormality. High resolution two dimensional ultrasound scanning has led to a whole new spectrum of findings and in some cases the margins of normality and abnormality can be blurred. Any evidence of significant renal pelvic dilatation requires serial scans to assess progress. Where possible the detailed measurement of antenatal renal pelvis dilatation should be recorded rather than referral to severity or grade.

Abnormal renal tract dilatation is seen when the AP diameter of the renal pelvis is greater than 8mm or when calyceal involvement is seen. Although controversial, postnatal dilatation of the renal pelvis is considered significant if more than 7mm. Measurements >7mm and <10mm are classified as mild, >10mm <15 mm as moderate and >15 mm as marked. A thinned, echogenic or cystic cortex suggests dysplasia. A thick walled or dilated bladder in the presence of renal pelvis dilatation is also very significant. The ureter is not usually seen on US – when it is dilated then further investigations are necessary.

In general, the likelihood of post natal abnormality is proportional to the severity of the fetal abnormality. Mild degrees of renal pelvis dilatation tend to be associated with VUR while more severe degrees of dilatation are associated most commonly with PUJ. When there is a family history (first degree relative) then the risk in future pregnancies is variable and depends on the specific condition. In general when the condition is bilateral there is a greater risk of recurrence. However even for unilateral conditions with a low recurrence risk there is often considerable anxiety for future offspring and a normal postnatal ultrasound scan is very re-assuring to parents.

2. Bladder enlargement

A dilated bladder on antenatal scanning in the absence of any other abnormality is an unusual finding. It is particularly concerning in boys. Arrange a renal US on D3 and at two weeks. If abnormal refer to Urology, otherwise arrange renal clinic follow up.

3. Congenital nephrotic syndrome

Congenital nephrotic syndrome is very rare. The kidneys appear enlarged and echogenic. There is usually marked polyhydramnios. The outlook is poor. Discuss management urgently with nephrologist.

4. Crossed fused ectopia

This is an uncommon finding. One of the kidneys crosses the midline and fuses with the lower pole of the other. This may be difficult to distinguish between a single kidney on US and other investigations are necessary. There is a slight male predominance and crossing from LR occurs more frequently than from R-L. The point of fusion is usually between the upper pole of the crossed kidney and the lower pole of the normally positioned kidney. If there is antenatal dilatation then arrange an ultrasound scan and discuss with the urology team. Otherwise arrange a scan at 2 weeks and follow up in the neonatal renal clinic.

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5. Bright kidneys

The assessment of renal parenchymal texture is very much dependent on the settings of the machine and the experience of the operator. Specific causes of a bright kidney include polycystic kidney disease, cystic dysplasia, damage from obstructive uropathy, glomerulocystic disease and congenital nephrotic syndrome. Investigation and follow up should depend on the likelihood of any of these specific abnormalities. If the diagnosis is non specific arrange a renal US at 2 weeks. If this is normal then no follow up is necessary. If this is abnormal arrange follow up in the neonatal renal clinic.

6. Cystic dysplasia

Dysplasia takes numerous forms from renal agenesis to multicystic dysplasia. The multicystic dysplastic kidney is a large non functioning kidney where the whole of the renal substance is replaced with macrocysts of variable size. They are most commonly unilateral although they may be bilateral (fatal condition). Multicystic dysplastic kidneys can regress and disappear during pregnancy or grow to large sizes.

Cystic renal dysplasia is very variable in its presentation. Affected kidneys usually appear echobright and are usually small. There are sometimes co-existent abnormalities of the renal tract. Arrange a renal US at 2 weeks and a DMSA at 3 months and refer to nephrologists.

7. Duplex kidney

Renal duplication is a common anomaly. The collecting system is duplicated with two renal pelves and two ureters. The ureters may join into one ureter anywhere along their course or may enter the bladder through separate orifices. Most renal duplications are uncomplicated and asymptomatic. However they may be associated with both obstruction and VUR. The upper pole may be associated with an ectopic ureterocoele. Unilateral duplication is 5 times more common than bilateral.

Some degree of upper urinary tract dilatation may be seen in up to 0.8% of all patients and is seen more frequently when the duplication is bilateral. The vast majority of duplications are incomplete, with confluence of the ureters at some point above the ureteric orifice. US is the investigation of first choice. Arrange US by D3 if there is ureteric dilatation or at 4 weeks if there is not. If the renal pelvis AP diameter is less than or equal to 10 mm and there is no ureteric dilatation then follow up in renal clinic; otherwise refer to urology.

Duplex anomaly is transmitted as an autosomal dominant trait with incomplete penetration – among members of affected families the incidence is of the order of 8%. Arrange a renal US at 6 weeks. If normal no follow up is necessary. If abnormal discuss with radiologists and refer to neonatal renal clinic.

8. Enlarged kidney

Large kidneys can be caused by obstruction, abnormalities of parenchyma, rarely tumour or may represent organ hypertrophy as a hemi-hypertrophy syndrome. Follow up depends on whether any specific diagnosis is suspected. If there is no antenatal dilatation arrange a scan at 2 weeks and follow up in neonatal renal clinic.

9. Glomerulocystic disease

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Glomerulocystic kidney disease is a distinct histological entity that most commonly presents with very enlarged, diffusely echobright kidneys which are indistinguishable from polycystic kidneys. They may occur with other abnormalities and syndromes and chromosome analysis is usually indicated. Discuss management urgently with nephrologists.

10. Horseshoe kidney

Horseshoe kidneys are present in 1:400 - 800 and the anomaly is more common in boys. In the majority of cases the lower poles are joined by an isthmus of renal tissue. The fused portion may consist of fibrous tissue or normal renal parenchyma. The kidney is supported by multiple renal arteries arising from the lower abdominal aorta. These kidneys are at higher risk of reflux, infection and stones. There is also a strong association with horseshoe kidneys and certain chromosomal syndromes. If in doubt consider genetic review or chromosomal analysis. Because the ureters pass over the isthmus there is a high incidence (1 in 5) of pelviureteric abnormalities. Arrange a renal US at 2 weeks of age. If abnormal (suggesting dilatation or other abnormality) refer to urology. If normal follow up in neonatal renal clinic at 3 months.

11. Hydronephrosis

Hydronephrosis is a descriptive term denoting pathological dilatation of the renal pelvis and calyces. Hydronephrosis may be due to obstruction, VUR or developmental abnormalities of the upper urinary tract. Hydronephrosis is normally consistent with a dilated renal pelvis of >15 mm. When the renal pelvis AP diameter is greater than 10 mm refer to urology. MCU or MAG3 investigations may be necessary depending on progression of renal tract dilatation.

12. Pelvic kidney

Developmental renal anomalies occur early in embryonic stages when the renal tissue does not move with the normal relationship with other abdominal organs. An ectopically sited kidney may be located anywhere along the embryological path of ascent from the pelvis to the renal fossa. Pelvic kidneys are the commonest form of renal ectopia. In 90% of cases the condition is unilateral, with a left sided predominance. Abnormally placed kidneys may be very small and dysplastic or may have other anomalies such as obstruction of drainage.

Arrange an ultrasound at 2 weeks of age. Provided there is no renal pelvis or ureteric dilatation then follow up in renal clinic and repeat scan at 3/12 and 1 year to assess interval growth.

13. Pelvi-ureteric junction obstruction

There is no universally accepted definition of obstruction and sometimes the terms pelviureteric junction obstruction and hydronephrosis may be used synonymously. Pelviureteric junction obstruction (PUJ) is also a heterogeneous disorder with a number of causes and variations. The diagnostic pathway includes most of the standard investigations although the actual sequence depends on initial findings and early progression of abnormality. The calyces and pelvis are dilated to a varying degree with no ureteric dilatation. The renal pelvis may often be disproportionately more dilated than the calyces. A prominent or large renal pelvis does not necessarily mean that there is hydronephrosis or obstruction. There can be a prominent renal pelvis without calyceal dilatation. If the calyces are not dilated then there is not likely to be obstruction. The renal parenchyma may be thinned to a varying degree PUJ is the most common form of prenatally detected hydronephrosis. Mild to moderate obstruction does not give rise to detectable dilatation at the time of most routine ‘anomaly’ scans but more usually becomes apparent on later pregnancy scans. Before the
introduction of antenatal ultrasounds most infants presented with symptomatic urinary infections.

PUJ obstruction is more common in boys and it is sometimes associated with other renal abnormalities. Depending on the progression, surgery may be indicated. If the renal pelvis AP diameter is greater than 10mm then refer to urology otherwise follow up in neonatal renal clinic.

These conditions do not usually have a tendency to recur in other pregnancies. Arrange a routine renal US at 6 weeks in the presence of a family history of PUJ

14. Polycystic kidney

Inherited polycystic disease frequently presents antenatally. Both the infantile recessive form (ARPKD) and adult dominant form (ADPKD) can be detected on the anomaly scan. Although commonly perceived that ADPKD is an adult only condition, all forms of PKD can have manifestations in infants and children. Because simple cysts are extremely rare in childhood, the finding of even one cyst should alert the clinician to the possibility of ADPKD and may necessitate the scanning of parents or older siblings.

ARPKD typically presents as bilaterally enlarged echobright kidneys with oligohydramnios. There may be obstruction of labour due to the distended fetal abdomen. In these cases there is usually severe pulmonary disease and a poor outcome. However there may be less severe variations, with less marked oligohydramnios with a better outcome at birth; renal failure will eventually ensue at some point. Discuss management with nephrologist and if poor outcome is expected then a senior neonatologist should attend the delivery.

ADPKD is much more common and may present antenatally. When the kidneys are very enlarged and there is oligohydramnios then the prognosis is poor. However with moderate renal enlargement with a small number of cysts and normal liquor volume then the outcome is good. If neonatal disease is suspected then discuss management and follow up with Dr Moss.

Family history: On the whole families tend to have infants that follow a similar clinical pattern. Fetal diagnosis is available through chorionic villous biopsy. When the outcome is expected to be poor then there should be detailed consultation with senior staff. Whenever large echobright cystic kidneys are detected on antenatal scan, both parents should be offered renal US to look for evidence of ADPKD. However the majority of patients with ADPKD are usually diagnosed and become symptomatic in adulthood. If the neonatal presentation is unlikely on the basis of normal antenatal US and liquor volume then the parents should be informed to arrange a confirmatory US in early adult life for their child to exclude adult onset ADPKD. The family should be asked to arrange this when their child is older via their GP.

15. Single kidney

A single kidney may represent abnormal renal tract development or destruction following a fetal uropathy. Although this may be associated with other abnormalities in the majority of cases a single kidney is associated with a normal outcome and no long term follow up is necessary. If the single kidney was otherwise normal on antenatal scan, a 2 week follow up scan should be arranged and follow up through the neonatal renal clinic.

16. Ureteric dilatation

The normal ureter is not normally visible on antenatal US. Dilatation occurs in PUV or VUJ obstruction. Non obstructive causes include reflux +/- dysplasia. Diagnosis depends on whether the collecting system is obstructed (PUV, VUJ) or non obstructed and appropriate investigations should be arranged by following the AUTA flowchart.

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17. Ureterocele

A ureterocele is a cystic dilatation of the intravesical portion of the ureter, occurring in 1:500. They are usually associated with a duplex kidney and tend to occur in the ureter draining the upper pole. It is important to start the infant on prophylactic antibiotics when one is suspected on the basis of antenatal scans. Reflux may occur in 20-40% into the lower pole. Follow the AUTA guidelines and refer to urology if the diagnosis is confirmed on postnatal scans.

18. Urethral valves

The syndrome of posterior urethral valves is a serious condition that requires senior involvement. Diagnosis is often made antenatally and in some cases fetal drainage procedures may be necessary. In less severe cases the presentation may be delayed until early infancy with signs of sepsis, a palpable bladder or weak or intermittent urinary stream. The majority of cases are now detected antenatally and the family will normally have a chance to meet with the urologists. Arrange an urgent renal US and discuss with the urology consultant. An MCUG may need to be undertaken with a suprapubic catheter in place – discuss before arranging this.

19. Vesicoureteric junction obstruction

Vesicoureteric junction obstruction (VUJ) is a terminology which includes the major categories of ureteric dilatation (megaureter). These include obstructed megaureter, non refluxing non obstructed megaureter and refluxing megaureter. The distinction between these forms cannot be made on US alone and MCUG is always indicated to exclude VUR or in boys, posterior urethral valves. The ureter and renal pelvis is dilated down to the level of the bladder. The calyces may be mildly dilated with more marked dilatation of the distal ureter. VUJ may also be associated with obstruction of an upper pole renal duplication.

VUJ is seen in approximately 10% of prenatally detected uropathies. It is the second commonest cause of prenatally diagnosed hydronephrosis. Diagnosis is confirmed by MCUG (at 2 months) and MAG3 scanning (at 3 months). VUJ is not usually associated with other abnormalities. If the renal pelvis AP diameter is greater than 10mm then refer directly to urology, otherwise follow up in neonatal renal clinic.

For future pregnancies these conditions do not usually have a tendency to recur. Arrange a routine renal US at 6 weeks when there is a family history of this condition.

20. Vesicoureteric reflux

Vesicoureteric reflux (VUR) is a very important and complex condition which may lead to symptomatic infection and renal scarring. Advances in the understanding and natural history of VUR have led to a more conservative approach to management and the adoption of more selective indications for surgery. There is a gender difference in presentation. In girls the condition presents clinically in early to mid childhood and is usually of low grade. In boys the presentation is either clinically or prenatally and tends to present in early childhood. There are often moderate to high grades of VUR.

Genetic factors in VUR are now well established. VUR may be seen in 34% of siblings who presented with symptoms and in more than 50% of the children of women who have a history of VUR. Arrange an MCUG with antibiotic prophylaxis at 3 months of age and follow up in renal clinic.

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References

Review articles


Textbooks


Avery’s Diseases of the Newborn. Ed H William Taeusch. *Saunders 2004*