Surgical management of renal tract problems

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Abstract

The majority of renal tract problems in childhood are congenital in origin with an overall incidence of 2–4 per 1000 live births. Some anomalies may occur in isolation e.g. posterior urethral valves; whilst others may be part of a more complex spectrum of anomalies. Prenatal screening has highlighted an increasing number of children with renal tract anomalies which may require paediatric specialist involvement in the immediate postnatal period. Most congenital renal tract problems are managed by paediatric urologists and paediatric surgeons with a special interest in urology. However, it is important that paediatricians have an understanding of optimal management. The aim of this review is to highlight the most common renal tract disorders of childhood and to discuss their aetiology, clinical presentation, investigation and surgical management.

Keywords duplex kidney; horseshoe kidney; multicystic dysplastic kidney; pelvi-ureteric junction obstruction; posterior urethral valves; vesico-ureteric obstruction; vesico-ureteric reflux

Topics covered in this review.

Investigation of renal tract problems
Pelvi-ureteric junction obstruction
Duplex anomalies
Multicystic dysplastic kidney
Horseshoe kidney
Vesico-ureteric reflux
Vesico-ureteric junction obstruction
Posterior urethral valves

Table 1

Investigation of renal tract problems

Most common paediatric renal tract problems (Table 1) can be investigated using four primary diagnostic studies.

Renal ultrasound scan

A renal tract ultrasound scan (USS) evaluates the following: renal size and growth, structural anomalies, significant parenchymal thinning or scars, hydronephrosis, bladder capacity and post-void residual volume. The anterior–posterior (AP) diameter of the renal pelvis represents the distance between the renal cortex in the transverse plane and is reported in the context of hydronephrosis.

Micturating cysto-urethrogram

A micturating cysto-urethrogram (MCUG) requires the insertion of a urethral catheter to allow the bladder to be filled with contrast. A MCUG demonstrates the size and shape of the bladder, presence of vesico-ureteric reflux (VUR), appearance of the bladder neck and must always include catheter-out voiding urethral views in boys.

DMSA scan

A di-mercapto-succinic acid scan (DMSA) is a nuclear medicine study which evaluates renal tubular extraction. The radio-isotope is given intravenously and images are taken 2 hours later. A DMSA demonstrates differential function, relative renal size and focal cortical thinning.

MAG3 renogram

A di-mercapto-acetyltriglycine renogram (MAG3) nuclear medicine study evaluates renal tubular secretion. The radio-isotope is given intravenously and images are taken immediately afterwards. A MAG3 demonstrates differential function and drainage of the kidney. Studies performed in the context of renal pelvis dilatation require an assessment of the effect of change in posture and micturition on drainage. In children who are toilet trained and will void on demand, a MAG-3 indirect cystogram can assess for VUR without the need for catheterisation.

Pelvi-ureteric junction obstruction

Obstruction at the level of the pelvi-ureteric junction (PUJ) may be due to intrinsic narrowing of the PUJ, high insertion of the PUJ into the renal pelvis or due to extrinsic compression from accessory lower pole vessels or a combination of the above. The obstruction causes a reduction in urine flow and progressive renal dilatation, culminating in irreversible loss of nephrons. The incidence is difficult to quantify but approximately 1:1000–1500 children will undergo an operation for this condition. The majority of patients are asymptomatic and are diagnosed after prenatal scanning has demonstrated hydronephrosis which is confirmed postnatally. Older children may present with intermittent loin pain, infection or haematuria. Renal USS and MAG-3 renogram aid the diagnosis and guide the definitive management.

For children without symptoms, with moderate hydronephrosis (<20 mm) and absent or minimal calyceal dilatation, and a differential function >40% on MAG-3 study, it is acceptable to manage these cases conservatively. A significant proportion will show improving hydronephrosis, will maintain good function and will not require surgical intervention.

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Indications for surgery for PUJ obstruction

In patients with a significant degree of hydronephrosis (>30 mm) or increasing hydronephrosis, where differential functional is <40% or in children with symptoms, then surgical intervention may be indicated.

Pyeloplasty for PUJ obstruction

The Anderson–Hynes pyeloplasty is the procedure of choice for children of all ages. The operation can be performed in an open manner, laparoscopically or robotically. The approach chosen has little effect on the success of the procedure.

In infants less than one year old, the open approach is preferred, as it can be performed with a short operative time and through a small incision. Laparoscopic pyeloplasty in infants is possible but requires very advanced laparoscopic skills and confers little overall advantage to the patient.

For older children, the approach depends on the operating surgeon's preference and experience. Laparoscopic and robotic techniques can result in the child having a shorter length of stay, less analgesia, and intraoperative blood loss, in comparison to the open technique. They are particularly advantageous in older children and adolescents where access to the renal pelvis is more difficult using an open approach. Robotic surgery is the most expensive option and very few hospitals have access to the necessary equipment. Therefore a laparoscopic or open approach is often more common in older children.

In all types of operation the stenotic PUJ is excised and the distal ureter spatulated to allow a wide anastomosis to be performed between the renal pelvis and proximal ureter. The use of a trans-anastomotic stent varies considerably amongst clinicians. Complications are rare and include bleeding, infection, anastomotic leak and anastomotic stenosis. The procedure has an overall success rate of 95%. Repeat renal USS after 3–6 months typically shows a reduction in the degree of hydronephrosis but it usually takes several years for this to fall within normal limits. A repeat MAG-3 is performed 12 months after surgery to demonstrate improved drainage and preserved function of the kidney (Figure 1).

Duplex anomalies

Duplex renal anomalies affect approximately 1% of the population and 30–40% of patients will have bilateral findings. Most are *incomplete* duplex kidneys; the ureteric bud bifurcates and therefore there are two ureters at the level of the kidney but only one ureteric orifice correctly sited within the bladder. *Complete* duplex anomalies occur less frequently and arise when two ureteric buds develop from the distal mesonephric duct and interact independently with the metanephros forming a kidney with an upper and a lower pole. The ureter relating to the lower pole has an abnormal lateral position within the bladder and is associated with vesico-ureteric reflux. The ureter of the upper pole may have an associated ureterocele (cystic dilatation of the intravesical ureter) which can cause obstructive symptoms. In some girls the upper pole ureter may be ectopic, inserting into either the urethra or vagina.

Most cases are detected prenatally with hydronephrosis resulting from the ureterocele or reflux. Older children may present with recurrent UTI or dribbling urinary incontinence. A MCUG is recommended in all cases where a ureterocele is detected in order to demonstrate the relationship of the ureterocele to the bladder neck and the presence of lower pole VUR. Ectopic ureters can be very difficult to identify cystoscopically and in such cases magnetic resonance urogram (MRU) may demonstrate the abnormality.

Surgical management of duplex anomalies

The majority of children with an incomplete duplex anomaly will be asymptomatic. However, for complete duplex anomalies the surgical strategy depends on the age of the child and whether the issues relate to the ureterocele, ectopic ureter or VUR.

In infants with a ureterocele causing significant dilatation of the upper pole, the first line of intervention can be cystoscopic puncture of the ureterocele. This is a relatively straightforward procedure but does carry a small risk of infection. Good decompression is confirmed by renal USS and if the child remains asymptomatic it may be that no further intervention is required, even if there is minimal or no function in the upper moiety. Children with recurrent UTIs and significant hydronephrosis will benefit from open or laparoscopic upper pole hemi-nephroureterectomy. This is also the procedure of choice for girls with dribbling incontinence due to an ectopic upper moiety ureter.

Recently it has been suggested that in children with incontinence due to an ectopic ureter, that simple laparoscopic ligation of the ureter alone can be carried out, without removal of the upper renal moiety. This novel technique has been successful in helping children to become continent. However a post-operative complication is hydronephrosis which does require follow-up imaging for monitoring.

In children in whom investigations have revealed significant VUR into a poorly functioning lower pole there are three possible strategies for management. If the child is asymptomatic they can be managed conservatively with antibiotic prophylaxis. A second option is to perform endoscopic injection of Deflux™ with an associated 50–60% success rate, significantly lower than that for single system VUR. The remaining option is to perform open or laparoscopic lower pole hemi-nephroureterectomy which is favoured in those cases with minimal function in the lower pole and recurrent UTIs.

With open or laparoscopic hemi-nephroureterectomy there is a small but definite risk of some loss of function of the remaining moiety. Approximately 5% of children having this procedure will have future symptoms relating to the distal ureteric stump and may require a further procedure to remove it.

Multicystic dysplastic kidney

A multicystic dysplastic kidney (MCDK) arises due to abnormal interaction of the ureteric bud with the developing metanephros resulting in failure to induce normal nephrogenesis. The incidence of MCDK is 1:2000–4000 live births. Most cases are sporadic and there may be associated contra-lateral VUR in up to 40% of patients.

A MCDK is usually diagnosed prenatally with the finding of multiple cysts in the area of the renal bed with thin, abnormal parenchyma. Older children may be diagnosed following investigation for UTI, abdominal pain or a palpable abdominal mass.