

# Urinary Tract Anomalies in Children - Diagnostic Dilemma.

Ajay Verma<sup>1</sup>, Rajul Rastogi<sup>2</sup>, Yuktika Gupta<sup>3</sup>, Neha<sup>4</sup>

<sup>1</sup>Assistant Professor, Department of Surgery, <sup>2</sup>Associate Professor, <sup>3</sup>Senior Resident, <sup>4</sup>PG Resident,  
Department of Radiodiagnosis, Teerthanker Mahaveer Medical College and Research Center,  
Moradabad, Uttar Pradesh, India

## Abstract

Urinary tract anomalies are a commonly encountered in the pediatric as well as pediatric surgery outpatient departments. As many of them are detected in antenatal fetal ultrasonographic examination, a proper counselling by an expert paediatric surgeon is required to alleviate the concerns and doubts of the parents. But in Indian scenario as an expert pediatric surgeon is often not available in most of the medium-sized cities and smaller towns, the radiologist and the treating paediatrician share the responsibility of early identification and prompt referral to higher centre for proper management. In this article, we will be discussing the clinical aspects of various congenital urinary tract anomalies in children in detail, however the embryology and development of these congenital anomalies is beyond the scope of this article.

## Introduction

Childhood urinary tract anomalies are broadly classified into congenital and acquired. Though many congenital urinary tract anomalies e.g. posterior urethral valve (PUV), pelviureteric junction obstruction (PUJO) are detected by antenatal ultrasonography (USG) yet some of them e.g. vesicoureteric reflux (VUR) are easily missed and picked up in postnatal life. Urinary tract calculus is an acquired disease but is often secondary to a pre-existing congenital urinary tract anomaly or repeated urinary tract infection (UTI).

## Classification

For descriptive purposes the anomalies are classified as per their site of origin. (Table 1).

### Kidney

The presence of supernumerary kidneys is very rare and less than 100 cases have been reported, when present, 60% of them are caudal to the ipsilateral kidney. This anomaly occurs more frequently on the left side. About half of them will join the bladder through separate ureter and half of them join the ipsilateral ureter. The extra kidney has its own renal capsule and blood supply. Most of these kidneys are asymptomatic and diagnosed at

### Address for correspondence

Dr. Rajul Rastogi, Associate Professor, Department of Radiodiagnosis,  
Teerthanker Mahaveer Medical College and Research Center, Moradabad,  
Uttar Pradesh - 244001, India E-mail: eesharastogi@gmail.com

Received: 26.09.2018

Accepted: 19.03.2019

Table 1 : Classification of urologic anomalies in children

### KIDNEY

- Supernumerary kidney
- Renal agenesis: Unilateral / Bilateral Malrotation
- Renal Ectopy
- Fusion anomaly: Horse shoe kidney
- Crossed renal ectopia
- Fetal lobulation
- Cystic disease of kidney
- Hydronephrosis
- Pelviureteric junction obstruction
- Tumours: Wilms tumour

### URETER

- Megaureter
- Duplication of ureter
- Ectopic ureter
- Ureterocele
- VUR

### BLADDER

- Exstrophy & Epispadias

### PUV

### URETHRA

- Hypospadias

autopsy; presence of any other associated renal anomaly should be looked for in these patients. Unilateral renal agenesis also goes unnoticed in most of the patients but patients of bilateral renal agenesis (potter's syndrome) succumb to early respiratory failure secondary to pulmonary hypoplasia.

Abnormal rotation, or malrotation, is most commonly associated with an ectopic or fused kidney, but may also occur in kidneys that undergo complete ascent. In the normal adult kidney, the renal pelvis is oriented medially and the calices point laterally. The fetal kidneys undergo a 90° rotation during the 6th–8th weeks of embryonic development to achieve normal orientation. The most common is an incomplete rotation, or non-rotation. The renal pelvis is anterior or between the fetal anterior and normal medial position in the adult. Other major types of anomalous rotation are reverse rotation and hyper-rotation (excessive rotation >180° & < 360°) in which the kidney faces laterally, but these are rare. In reverse rotation, the renal pelvis rotates laterally and the renal vessels cross the kidney anteriorly to reach the hilum. The anomalies of rotation are usually diagnosed incidentally during routine USG examination. Most of the patients are asymptomatic however some may complain of pain after developing hydronephrosis. It is important to establish the correct diagnosis and to exclude other pathologic conditions that can produce similar distortion of the kidney. Treatment of malrotation is reserved for alleviation of associated obstruction, calculi, or infection secondary to poor drainage.

Renal ectopy is described as kidney outside the renal fossa; the incidence of renal ectopy in post-mortem studies varies from 1 in 500 to 1 in 1290 [1, 2]. There is slight preponderance to left side, 10% cases are bilateral. The renal ectopy can be simple where it remains in the ipsilateral retroperitoneal space or crossed where it crosses midline and reaches the contra lateral side. Most common site for simple renal ectopy is pelvic region (opposite to sacrum below the aortic bifurcation). A kidney is intrathoracic when either a portion or all extends above the diaphragm. This accounts for less than 5% of renal ectopy, with an incidence of 1 in 13000 autopsies. Anomaly of rotation and fusion often accompany renal ectopy. Ptotic kidney is often confused with ectopic kidneys. In ectopic kidney, the length of ureter corresponds to the location but the ureter of ptotic kidney is redundant. Ptotic kidney is mobile and can be manipulated to its normal location. Most of the ectopic kidneys are diagnosed after UTI during USG examination.

The horseshoe kidney is the most common renal fusion anomaly, with the two renal masses joined at the lower poles in more than 90% of cases. The isthmus crossing

the midline joining the two kidneys consists of either renal parenchyma or fibrous tissue. The horseshoe kidney is usually positioned low in the abdomen, with the isthmus lying just below the junction of the inferior mesenteric artery and aorta. The reported incidence of horseshoe kidney varies from 1 in 400 to 1 in 1800 [3, 4]. In the majority of cases, there are multiple renal vessels. The blood supply to the isthmus is particularly variable, often supplied by a separate vessel. This may arise from the aorta, common iliac, or inferior mesenteric arteries. A report by Cascio et al found VUR in 13/40 patients with horse shoe kidney undergoing cystography [5]. Multicystic dysplasia [6] and autosomal dominant polycystic kidney disease have also been reported [7]. Hypospadias and undescended testes occur in 4% of the males, bicornuate uterus or septate vagina in 7% of females [8]. Retrocaval ureter has been found in association with a horseshoe kidney in six patients [9]. The diagnosis of a horseshoe kidney can be confirmed by a variety of imaging techniques, including USG, computerised tomography (CT), or magnetic resonance imaging (MRI).

Crossed renal ectopia is the second most common fusion anomaly. The ectopic kidney crosses the midline to lie on the opposite side from its ureteral insertion into the bladder. Figure 1 explains four common types of crossed renal ectopia. Crossed renal ectopia with fusion is most common type (85%). With all of these abnormalities, there is a slight male predominance and crossing from left to right occurs more frequently than right to left. McDonald and McClellan described six different varieties of crossed ectopia with fusion [10].

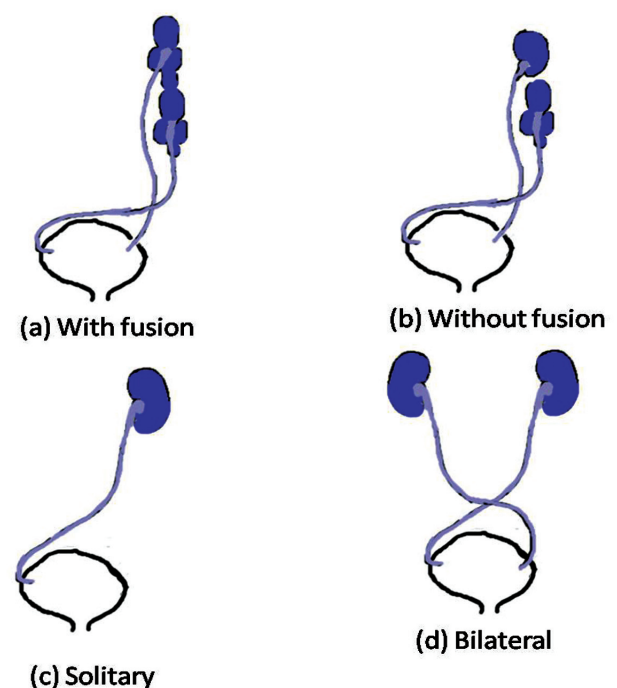


Figure 1: Types of crossed renal ectopia

Fetal lobulations are found at autopsy in 17.6% of children and in 3.9% of adults. This condition is of no clinical importance and should be recognized as a normal variant.

Less than 0.3% of children have simple renal cysts and they are usually not associated with subsequent problems. Cysts in children usually do not increase in size, and single cysts are commonly located in the right upper pole. Simple renal cysts do not impair renal function except in rare instances. They may play a role in causing infection or hypertension in some patients. Cysts in children occasionally cause pain. These cysts can be marsupialised by laparoscopy or open surgery. Autosomal dominant polycystic renal disease (ADPKD) and autosomal recessive polycystic renal disease (ARPKD) are genetic disease leading to multilocular renal cysts and early renal failure. The common features and differences are mentioned in Table 2. Multi cystic dysplastic kidney (MCDK) is the most common cystic disease diagnosed during childhood. Most cases are unilateral and asymptomatic; rare bilateral disease is usually fatal at birth as a result of Potter's syndrome. The incidence of MCDK is estimated at one in 2500 newborns [11] it is one of the most common fetal anomalies detected by prenatal ultrasound. It occurs slightly more commonly in the left kidney [12] and the affected child is more likely

to be male. MCDK is typically composed of cysts of varying size that do not appear to communicate and a small amount of abnormal - appearing renal parenchyma. The ureter from the affected kidney is atretic. On microscopic examination, the tissue between the cysts is dysplastic, with undifferentiated mesenchymal cells, often with cartilage and immature glomeruli and tubules. The renal cysts are also seen in patients of tuberous sclerosis, Meckel's Syndrome and Von-Hippel-Lindau disease.

Hydronephrosis is dilatation of renal collection system due to obstruction or retrograde flow of urine; it can be due to congenital PUJO or by any pathology leading to obstruction of flow of urine. Severe grades of VUR also cause hydronephrosis. Hydronephrosis is the most common abnormality detected on maternal - fetal ultrasound and accounts for nearly 50% of all prenatally detected lesions [13]. PUJ obstruction is twice as common in males as females, particularly in the neonatal period, with 66% occurring on the left side, as opposed to adults, in which there is a predilection for the right side [14]. The etiology of PUJO can be *intrinsic* (non peristaltic segment), *extrinsic* (crossing lower pole vessel) or *intraluminal* (calculus/polyp). The most common presentation is painless, palpable abdominal mass. It can also present with symptoms such as pain, fever and UTI.

Table 2 : Differences between ADPKD and ARPKD.

	<b>Autosomal dominant polycystic kidney disease (ADPKD)</b>	<b>Autosomal recessive polycystic kidney disease (ARPKD)</b>
<b>Genetics</b>	ADPKD has been associated with three different mutations: <i>PKD1</i> on chromosome 16, <i>PKD2</i> on chromosome, <i>PKD3</i> has not yet been characterised.	Transmitted by <i>PKHD1</i> , a gene located on chromosome 6.
<b>Clinical features</b>	By age 25 years, at least 85% of individuals will have cysts, but most will be asymptomatic. Microscopic and gross hematuria. Flank pain, hypertension, and renal colic secondary to either clots or stones. Cysts are macroscopic.	Most children with ARPKD die in utero, if they survive the first month of life, the chances of living for a year with proper supportive therapy are improved. Cysts are microscopic.
<b>Associated anomalies</b>	Colonic diverticula, hepatic cysts, and berry aneurysms. Hepatic cysts appear much more frequently in adults than in children and are more common in females.	Hepatic fibrosis, portal hypertension, esophageal varices, and hepatosplenomegaly.
<b>Prognosis</b>	Important to monitor these children for hypertension, because control of blood pressure delays the onset of renal failure.	Eventually, all infants with ARPKD develop renal failure.

Nephroblastoma, or Wilm’s tumor, is an embryonal tumor that develops from remnants of immature kidney. It is the most common renal tumor of childhood. Wilm’s tumor accounts for 6–7% of all childhood cancers. It typically affects young children (median age 3.5 years), although older children and occasionally even adults can be affected. Wilm’s tumor occurs at an earlier median age in children with bilateral tumors, 29.5 months for boys and 32.6 months for girls. Most children present as painless palpable abdominal mass, it has an overall good prognosis with 100% five year survival rate in stage 1 disease and more than 90% five year survival in stage 2 patients.

**Ureter & Bladder**

Megaureter describes a ureter that is abnormally enlarged. The ureter may be greatly elongated and tortuous as well. In general, the two principal pathologic types of megaureter are those that are obstructed and those with massive vesicoureteral reflux. A *duplex (duplicated) system* indicates a kidney with two pelvicaliceal systems. *Complete duplication* indicates a kidney with two ureters that drain separately into or below the bladder. *Incomplete duplication* refers to a kidney with two ureters that fuse into a single unit that drains into the bladder through a single orifice. A *bifid system* is a form of incomplete duplication. An *ectopic ureter* refers to a ureter that drains into an abnormal site. The ectopic ureter in male drains above the sphincter and hence is continent whereas in females it drains below sphincter making them incontinent. An *ureterocele* is a cystic dilatation of the lower end of the ureter where it joins the epithelium of the lower urinary tract. An *intravesical ureterocele* lies entirely within the bladder, whereas an *ectopic ureterocele* has a portion that lies below the bladder neck. Ureteroceles are called *single system* when associated with a single ureter and *duplex system* when associated with a double ureter.

Vesicoureteral Reflux (VUR), which is the abnormal flow of urine from the bladder to the kidney, allows bacteria from the bladder to gain access to the kidney. By itself, VUR does not cause UTI; bacteriuria must be present in the setting of VUR for UTI to develop. VUR is classified as in figure 2. The treatment of VUR can be medical, endoscopic or surgical based on grade of VUR [15].

In classic bladder exstrophy, the size of the exposed bladder also varies. The exposed bladder plate may be large, with wide diastasis of the pubis, or one or both sides of the bladder patch may be small and hypoplastic. Although the upper urinary tracts are usually normal at the time of birth, hydronephrosis does occur. After birth, the mucosa of the exposed bladder may, over time, undergo metaplasia associated with chronic inflammation, edema,

and fibrosis. This, in turn, may lead to chronic ureteral obstruction and hydronephrosis.

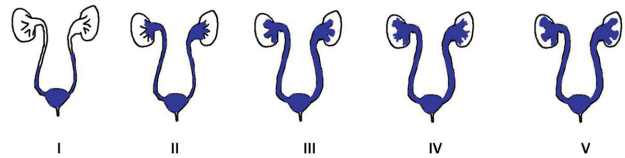


Figure 2: Classification of VUR. Based on voiding cystourethrogram. Grade I: reflux only to non dilated ureter and not reaching pelvis. Grade II: reflux reaching renal pelvis but ureter is not dilated. Grade III: reflux in dilated ureters but calices are not dilated. Grade IV: reflux is causing dilatation of renal calices but papillary impressions are maintained. Grade V: reflux causing loss of papillary impressions. Each ureter is graded separately.

**Urethra**

Posterior urethral valves (PUV) are the most common obstructive anomaly of the urethra. The incidence is between 1 in 5000 and 1 in 8000 male births. Most cases of PUV are detected prenatally by ultrasound showing hydronephrosis and/ or a distended thick-walled bladder. Postnatally, USG will show a thickened bladder wall and classically a dilated and elongated posterior urethra. Hydronephrosis will vary in degree and may be unilateral or bilateral.

Hypospadias is the most common congenital urologic anomaly. It occurs in 1 in 125 newborns. Hypospadias can be defined as an arrest in normal development of the urethra, foreskin, and ventral aspect of the penis resulting in various anomalies such as chordee or excessive ventral curvature, spatula like glans penis and dorsally hooded prepuce [11]. Hypospadias is classified as per location of meatus (Figure 3). Surgical aim is to achieve straight penis with meatus at the tip.

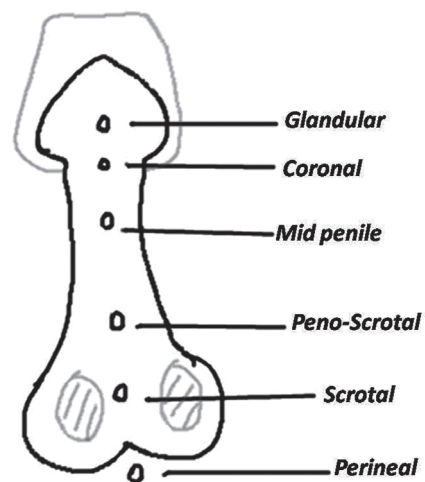


Figure 3: Classification of hypospadias based on location of meatus.

The identification of a urologic anomaly in children is a challenging issue; most of these have varied presentation and presence of associated anomalies makes their identification and management difficult. Whenever these anomalies are encountered by a radiologist or paediatrician it should be promptly referred to a pediatric surgeon for evaluation and management.

## Conclusion

To summarize, variety of congenital and acquired urinary tract anomalies occur in children with some of them escaping detection in antenatal period and presenting in childhood. A combined approach of a pediatric surgeon and radiologist is often required not only for early diagnosis but for optimal and early management.

<b>Conflict of interest:</b>	All authors declare no COI
<b>Ethics:</b>	There is no ethical violation as it is based on voluntary anonymous interviews
<b>Funding:</b>	No external funding
<b>Guarantor:</b>	Dr Rajul Rastogi will act as guarantor of this article on behalf of all co-authors.

## References

- Campbell MF. Renal ectopy. *J Urol* 1930; 24:187–98.
- Thompson GJ, Pace JM. Ectopic kidney: a review of 97 cases. *Surg Gynecol Obstet* 1937; 64:935–43.
- Campbell MF. Anomalies of the kidney. In: Campbell MF, Harrison JH, eds. *Urology*, Vol 2, 3rd edn. Philadelphia: WB Saunders, 1970: 1416–86.
- Dajani AM. Horseshoe kidney: a review of twenty-nine cases. *Br J Urol* 1966; 38:388–402.
- Cascio S, Sweeney B, Granata C et al. Vesicoureteral reflux and ureteropelvic junction obstruction in children with horseshoe kidney: treatment and outcome. *J Urol* 2002; 167:2566–8.
- Novak ME, Baum NH, Gonzales ET Jr. Horseshoe kidney with multicystic dysplasia associated with ureterocele. *Urology* 1977; 10: 456–8.
- Pitts WR Jr, Muecke EC. Horseshoe kidneys: a 40-year experience. *J Urol* 1975; 113:743–6.
- Boatman DL, Kolln CP, Flocks RH. Congenital anomalies associated with horseshoe kidney. *J Urol* 1972;
- Fernandes M, Scheuch J, Seebode JJ. Horseshoe kidney with retrocaval ureter: a case report. *J Urol* 1988; 140:362–4.
- McDonald JH, McClellan DS. Crossed renal ectopia. *Am J Surg* 1957; 93: 995–1002.
- Dillon E, Ryall A. A 10 year audit of antenatal ultrasound detection of renal disease. *Br J Radiol* 1998; 71:497–500.
- Rabelo EA, Oliveira EA, Diniz JS et al. Natural history of multicystic kidney conservatively managed: a retrospective study. *Pediatr Nephrol* 2004; 19:1102–7.
- Lettgen B, Meyer-Schwickerath M, Bedow W. Prenatal ultrasound diagnosis of the kidneys and efferent urinary tract: Possibilities, applications and dangers. *Monatsschrift Kinderheilkunde* 1993; 141: 462.
- Clark WR, Malek RS: Ureteropelvic junction obstruction. Observations on the classic type in adults. *Urol* 1987; 138:276.
- Bajpai M, Verma A, Panda SS. Endoscopic treatment of vesico-ureteral reflux: Experience of 99 ureteric moieties. *J Indian Assoc Pediatr Surg* 2013; 18: 133-5.
- Recent Advances in Hypospadias. Goel P, Bajpai M, Verma A. *JIMSA* 2014; 27(2): 95-100.

