

Congenital Anomalies of Kidney and Ureter

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Abstract

The kidney is a common site for congenital anomalies which may be responsible for considerable morbidity among young patients. Radiological investigations play a central role in diagnosing these anomalies with the screening ultrasonography being commonly used as a preliminary diagnostic study. Intravenous urography can be used to specifically identify an area of obstruction and to determine the presence of duplex collecting systems and a ureterocele. Computed tomography and magnetic resonance (MR) imaging are unsuitable for general screening but provide superb anatomic detail and added diagnostic specificity. A sound knowledge of the anatomical details and familiarity with these anomalies is essential for correct diagnosis and appropriate management so as to avoid the high rate of morbidity associated with these malformations.

Keywords: Kidney; Ureter; Intravenous urography; Duplex

Introduction

Congenital anomalies of the kidney and ureter are a significant cause of morbidity in foetus, neonate, infancy and adulthood. Radiological investigations provide a foundation for diagnosing these conditions giving vital clinical information. Various imaging modalities used to diagnose these anomalies are intravenous urography (IVU), voiding cystourethrogram (VCUG), Computed tomography (CT), Magnetic Resonance (MR) and nuclear imaging. Radiologist should be aware the urinary congenital anomalies and their imaging correlate in order to facilitate diagnosis and management.

There are innumerable congenital anomalies involving the kidney and ureter and the authors have attempted to give a pictorial overview of the imaging findings and embryology of various congenital anomalies of the kidney and ureter.

Congenital Anomalies of Kidneys

Anomalies in number

Anomalies in number can be in the form of supernumerary kidneys or renal agenesis

Supernumerary kidneys: Supernumerary kidney is an additional kidney to the number normally present (Figure 1). This can refer to more than two separate kidneys or an extra kidney coexisting with a horseshoe kidney. Upto five supernumerary kidneys have been reported in the literature. Supernumerary kidney is explained by secondary outpouching of the Wolffian duct or branching from initial ureteral bud; both induce metanephric anlage though they have separated entirely [1].

Supernumerary kidney needs to be differentiated from a Duplex kidney. Supernumerary kidney has a completely separate renal parenchyma and a renal capsule with a greater number of calyces as compared to Duplex kidney in which both poles are attached with a single renal capsule and the number of calyces does not exceed from that of the opposite kidney [2,3].

Two types of supernumerary kidney exist: (i) drained by a bifid ureter (ii) drained by a separate ureter. When a bifid system is present, supernumerary kidney lies caudally (Figure 2) and when

a separate ureter is seen then the supernumerary kidney is located cranially in relation to the normal kidney. In such a case the ureter enters the bladder ectopically and according to the Weigert-R Meyer rule the ureter may insert medially and inferiorly into the bladder [2]. If both horseshoe kidney and a supernumerary kidney coexist then the half of the horseshoe kidney on the side of the body containing the supernumerary kidney is always small. Complications include renal malignancy, stones and hydronephrosis. For the diagnosis of supernumerary kidney, intravenous pyelography, ultrasonography, nuclear scintigraphy, CT, and MRI can be used. On imaging, an

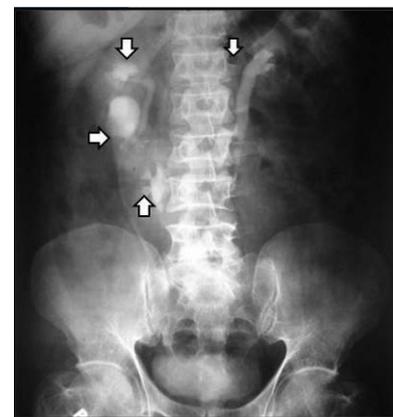


Figure 1: Intravenous Urogram film showing supernumerary kidneys (arrowheads) - four kidneys (three on right side and one on left side).

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Figure 2: Intravenous urogram (IVU) in a case of supernumerary kidney showing a normal left kidney and two right kidneys (arrows) with presence of a bifid ureter (arrowhead).

additional kidney is seen which is nearly always smaller than the normal kidneys with normal or impaired renal function.

Renal agenesis: Absence of development of kidney may be due to the absence of the metanephric blastema, maldevelopment of the ureteral bud or lack of induction of the metanephric blastema by the ureteral bud. Occasionally, post-natal involution of multicystic dysplastic kidney and hydronephrotic kidney can result in a solitary kidney [3]. It is important to look for additional associated ipsilateral urogenital anomalies which may include absence of the vas deferens, unicornuate uterus, absence or cysts of the seminal vesicle, skeletal abnormalities, anorectal malformations, cryptorchidism, cardiovascular abnormalities, VACTERL and MURCS anomalies. Renal genesis can be unilateral or bilateral. Bilateral renal agenesis is incompatible with postnatal life and is usually diagnosed antenatally because it causes maternal oligohydramnios. On imaging, unilateral renal agenesis can be diagnosed on plain film by the absence of a renal outline and medial displacement of the colonic flexure into the renal bed (Figure 3). The contralateral kidney, when normal, shows compensatory hypertrophy and this may be visible on the plain film.

Anomalies in position

Anomalies in position include anomalies of rotation and location.

Anomalies of rotation: Normally, the kidneys lie at an angle of 30 degrees from the horizontal making the renal pelvis to face medially and slightly anteriorly [4,5].

Five types of rotational anomalies have been described [6,7]

(i) Nonrotation – renal pelvis lies directly anterior to the kidney (kidney lies at angle of 90 degrees) (Figure 4)

(ii) Incomplete rotation – renal pelvis lies between 30-90 degree from horizontal (Figure 5)

(iii) Reverse rotation – renal pelvis lies laterally and renal vessels twisted anteriorly (Figure 6).

(iv) Excessive rotation – renal pelvis faces posteriorly or posteromedially and renal vessels twisted posteriorly.

(v) Transverse rotation – calyces pointing superiorly/inferiorly.

Although a number of theories have been proposed to explain the

anomalies of rotation, at the moment they are purely speculative. The embryological basis of rotational anomalies is still unknown [5,6].

Rotational anomaly though rare, has important implications from surgical as well as radiological point of view. On IVP, it may be mistaken for some more serious condition and may erroneously be attributed to displacement by paravertebral or lower pole mass due to displacement of ureters. It assumes a great importance in the context of present day surgical procedures like percutaneous nephrectomy and diagnostic evaluation of the kidney donors, etc. [6].

Anomalies of renal location: renal ectopia: Congenital abnormal location of a kidney is described as renal ectopia. It is important to differentiate this entity from renal ptosis which is defined as renal descent by 5 cm or more (2 lumbar vertebrae) in upright position. In renal ectopia, the arterial blood supply arises ectopically and the length of the ureter may be short or long depending on the location of the ectopic kidney whereas in ptotic kidney the ureter is of normal length (may be redundant when the patient stands) and the renal arteries arise from the normal sites.

Classification

Ipsilateral ectopia: Kidney is on the same side of the body as the



Figure 3: Intravenous urography film showing non-visualization of right renal outline suggestive of right renal agenesis and normal excreting left kidney (arrow).



Figure 4: Intravenous urography film showing non rotation (black arrow) of left kidney and normal rotation (white arrow) of right kidney.

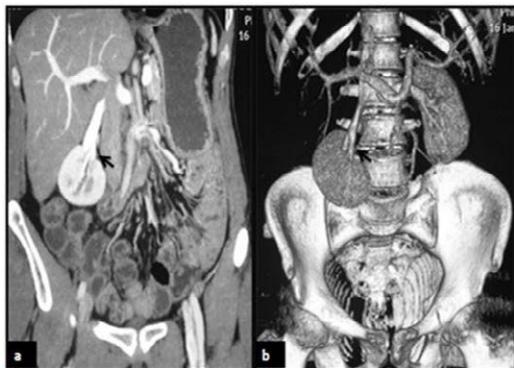


Figure 5: (a) Contrast-enhanced CT coronal MIP images showing incomplete rotation (arrow) of right kidney (b) Coronal CT volume-rendered images depicting (arrow) the finding better.



Figure 6: (a) Contrast-enhanced CT coronal MIP images showing reverse rotation (black arrow) of right kidney and non rotation of left kidney (white arrow) (b) Coronal CT volume-rendered images confirming the findings.

orifice of its attendant ureter. It is further divided into cranial or caudal ectopia in reference to relationship with its normal position. Caudal ectopia are further divided into – *Abdominal* (above the iliac crest but below L2 vertebra), *Iliac* (at the level of iliac crest) (Figure 7) and *Pelvic* (located in true pelvis) (Figure 8, 9).

Crossed renal ectopia: Kidney is located on the side of the body opposite the orifice of its attendant ureter. McDonald and McClellan classified renal crossed ectopia into four types: crossed renal ectopia with fusion (85%), crossed renal ectopia without fusion (10%), solitary crossed renal ectopia, and bilaterally crossed renal ectopia [8- 14].

Anomalies of renal fusion: Renal fusion anomalies are classified into horseshoe, disc and unilateral fused kidney or crossed fused renal ectopia. Embryological basis of renal fusion anomalies is related to their respective nephrogenic blastemas which squeeze together between the umbilical arteries at the beginning of the cranial migration of the ureteral buds leading to their fusion. Fused kidneys are usually ectopic in position. In all fused kidneys, the arterial supply and venous drainage are grossly abnormal [9].

Horseshoe kidney: Horseshoe kidney is the most common renal fusion anomaly, with an incidence of approximately 0.25% in the general population. It usually lies in a lower position than the normal kidneys because the isthmus does not permit ascent beyond the inferior mesenteric artery (IMA). Most cases are fused at the lower poles by an isthmus. Isthmus is comprised of either functioning renal parenchyma or fibrous tissue that crosses the midline of the body. The isthmus

usually lies anterior to the great vessels at the level of the L3-L5 vertebra, just below the origin of the IMA from the aorta [5,10].

Various genitourinary anomalies are associated with horseshoe



Figure 7: Coronal contrast-enhanced MIP images showing right caudal ectopic kidney (arrow) located at the level of iliac crest.



Figure 8: Intravenous urography film showing caudal ectopic right kidney (black arrow) in pelvis and normally located left kidney.



Figure 9: Intravenous urography film showing caudal ectopic pelvic kidney (black arrow) on left side and normally located right kidney (white arrow).

kidneys namely Vesicoureteral reflux, Ureteral duplication, ectopic ureterocele, retrocaval ureter, multicystic dysplasia, adult polycystic kidney disease, hypospadias, undescended testes, bicornuate uterus, and septate vagina [9]. Other congenital anomalies outside the genitourinary system are musculoskeletal (hemivertebrae, scoliosis, rib defects, clubfoot, congenital hip dislocation), cardiovascular (ventriculoseptal defects), gastrointestinal (anorectal malformation, malrotation and Meckel diverticulum), and central nervous systems (neural tube defects) anomalies. Complications of horseshoe kidneys are – more prone to injury (Figure 10), ureteropelvic junction obstruction, stones and tumors (Wilm’s tumor, carcinoid, transitional and renal cell carcinoma) (Figure 11).

Two types of horseshoe kidney are identified [5,11] depending upon whether midline or lateral fusion has occurred: (a) U-shaped and (b) L-shaped.

On imaging, horseshoe kidneys may be identified on abdominal radiographs. Four kinds of abnormal renal outlines [10] suggestive of horseshoe kidney are - lower than normal, too close to the spine, vertical long axis and visible isthmus (Figure 12). On excretory urography, ‘hand-holding calyces’ and ‘flower-vase’ appearance is seen (Figure 13).

Disc kidney: It is also known as cake or lump kidney. First identified and defined by Glenn, disc kidney is an anomaly in which “the entire renal substance is fused into one mass, lying in the pelvis, and giving rise to two separate ureters which enter the bladder in normal relationship” [12]. In rare instances, such kidneys possess one ureter. The fused kidney occupies prevertebral or presacral space [12] (Figure 14).

Unilateral fused kidney or crossed fused renal ectopia: Crossed fused renal ectopia is the second most common fusion abnormality of the kidney, with an estimated incidence of approximately 1:1300–1:7500 [13]. In crossed fused ectopia, one kidney crosses over to opposite side, and the parenchyma of the two kidneys fuse. Usually, the upper pole of the inferiorly positioned crossed ectopic kidney is fused to the lower pole of the superior, normally positioned kidney. The ureter of the ectopic kidney crosses the midline and enters the bladder on the opposite side [13].

Types of crossed fused ectopia [5,8]:

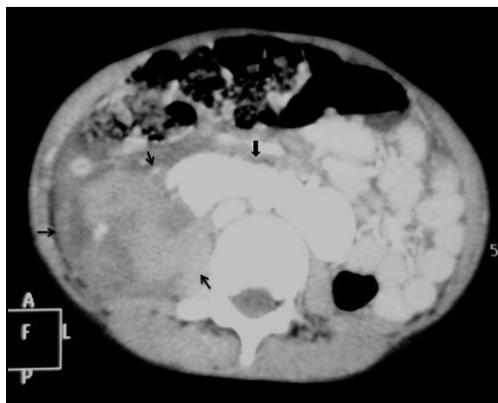


Figure 10: Axial contrast-enhanced CT scan a case of trauma in a horseshoe kidney showing isthmus (arrowhead) of horseshoe kidney and hematoma (arrows) formation on the right side due to trauma of right pole of horseshoe kidney.



Figure 11: Axial contrast-enhanced CT scan showing isthmus (white arrow) of horseshoe kidney with malignant mass (black arrow) in right half of horseshoe kidney.



Figure 12: Plain X-ray abdomen showing lower poles of kidneys (white arrows) lying medially and closer to the spine in a case of horseshoe kidney and bilateral symmetrical calculi (black arrows) in pelvis in lower ureters in a case of bilateral ureterocele.



Figure 13: Intravenous urography film in a case of horseshoe kidney (arrows) showing lower poles closer to the spine and lower calyces closer to each other giving the appearance of ‘hand holding calyces’.

- (i) Superior Ectopia – ectopic kidney crosses the midline and lies superior to the orthotopic kidney with fusion of the poles of kidney.
- (ii) Sigmoid or S-shaped Ectopia – ectopic kidney lies inferiorly



Figure 14: Axial contrast-enhanced CT scan showing disc like enhancing structure (arrows) in pelvis suggestive of a disc kidney.

with pelvis directed laterally and the pelvis of orthotopic kidney facing medially.

(iii) Unilateral Lump Kidney – two kidneys completely fused forming lump on one side.

(iv) L-shaped Kidney – ectopic kidney lies inferiorly and transversely and

(v) Unilateral disc kidney – kidneys fused along the medial concave border

(vi) Inferior Ectopia - ectopic kidney crosses the midline and lies inferior to the orthotopic kidney with fusion of the poles.

Blood supply to the ectopic kidney most frequently arises from the vessels on the ipsilateral side but occasionally arises from the contralateral side [9,15]. Cross fused renal ectopia is typically Asymptomatic and is diagnosed as an incidental finding when the patient is examined for other medical diseases. Complications are nephrolithiasis, ureteropelvic junction obstruction, hydronephrosis, reflux, ectopic ureteroceles and tumors [9-17].

On imaging, diagnosis of crossed renal ectopia may be suggested from abdominal radiograph if one renal outline is not visualised and the opposite renal outline is enlarged or when stones are seen at unusual positions. Excretory urography shows the absence of a kidney in its normal position with evidence of two kidneys on the same side of the abdomen vertically oriented one above the other. Ultrasound can determine if the kidney is in its normal renal fossa. The presence of the two kidneys on one side and the absence of a kidney in the contralateral side are suggestive of crossed ectopia (Figures 15- 20).

Anomalies in size

Renal hypoplasia: Renal hypoplasia signifies a congenital renal parenchyma anomaly in which too few nephrons are present [5]. Four types of renal hypoplasia exist – hypoplastic kidney with an orthotopically draining ureter also known as Miniature/Dwarf Kidney, – hypoplastic kidney with an ectopically inserting ureter, oligomeganephronia (bilateral renal hypoplasia with enlarged nephrons) and Ask-Upmark Kidney. Ask-Upmark kidney consists of a segmental renal scar and is characterized by hypertension, generally seen in young females. It usually affects only one kidney, which tends to be small and excretes normally. The scar is in the midzone. The number of calyces is less than seven and the calyces under these scars are clubbed [5-33].

Renal Dysplasia – It is a congenital renal parenchyma malformation in which abnormal nephrons and mesenchymal stroma are found.

It is important to differentiate hypoplastic kidneys from dysplastic kidneys. On imaging, a hypoplastic kidney is small (Figure 21) but otherwise normal whereas a dysplastic kidney is also small but it is poorly defined with presence of cortical cysts (Figure 22).

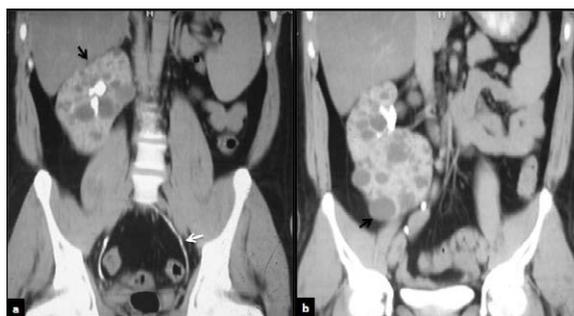


Figure 15: Contrast-enhanced CT coronal MIP images showing (a) crossed fused ectopia with polycystic kidney disease (black arrow) and opening of ectopic ureter (white arrow) in left side (b) crossed fused ectopic kidney with cysts (black arrow).



Figure 16: Axial T2W MRI of same patient depicting polycystic kidney disease (arrows) in case of right crossed fused ectopia.



Figure 17: Intravenous urography film in a case of ureteropelvic junction obstruction (PUJO) of right crossed fused ectopic kidney showing excretion of contrast in displaced pelvicalyceal system (black arrow) and ureter (white arrow) of orthotopic kidney with obscuration of right psoas shadow by PUJO of crossed fused ectopic kidney (black arrowhead) and non-visualised left kidney in left renal fossa.

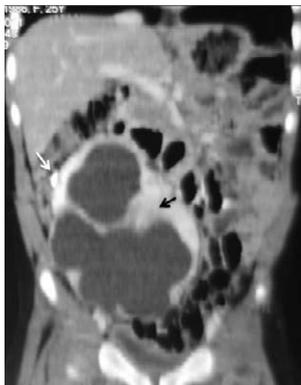


Figure 18: Coronal contrast-enhanced CT scan in same case of ureteropelvic junction obstruction (PUJO) of right crossed fused ectopic kidney showing excretion of contrast in displaced pelvicalyceal system (white arrow) and PUJO (black arrow) of crossed fused ectopic kidney.



Figure 19: Intravenous urography in a case of left crossed fused ectopia showing (a) orthotopic kidney (black arrowhead) with caudally placed crossed fused ectopic kidney (white arrowhead) and ureter (white arrow) of ectopic kidney opening on the right side (b) ureter (black arrow) of orthotopic kidney opening on left side.



Figure 20: Intravenous urography film in a case of left crossed fused ectopia showing ureteropelvic junction obstruction (black arrow) of the orthotopic kidney and crossed fused ectopic kidney (white arrow) located caudally and its ureter (curved arrow) crossing and opening in its normal position.

Differentials of unilateral renal dysgenesis are postobstructive atrophy, renal vascular anomalies, post-inflammatory atrophy, VUR, post-radiation therapy and heminephrectomy.

Congenital Anomalies of Ureter

Megaureter

Megaureter is defined as presence of an enlarged ureter with or without concomitant dilatation of the upper collecting system. A ureteric diameter of 7 mm or more should be considered a megaureter [18].

Types:

Primary: *Primary megaureter* includes all cases of megaureter due to an idiopathic congenital alteration at the Vesicoureteral junction (Figure 23). There are three subtypes - obstructed primary megaureter, refluxing primary megaureter, and nonrefluxing unobstructed primary megaureter.

In obstructed primary megaureter, there is dilatation above a short (0.5 - 4 cm), aperistaltic, normal-caliber juxtavesical section of a normally inserted ureter. Refluxing primary megaureter is caused by a short or absent intravesical ureter, congenital paraureteric diverticulum, or other derangement of the Vesicoureteral junction.

Secondary: *Secondary megaureter* occurs as a result of some abnormality involving the bladder or urethra (eg, urethral valves, neuropathic bladder dysfunction, urethral strictures, ureteroceles, and acquired causes of obstruction).

On imaging, first voiding cystourethrography (VCUG) is



Figure 21: Coronal contrast-enhanced CT scan showing small hypoplastic right kidney (arrow) which shows normal outline and nephrographic enhancement.



Figure 22: Coronal contrast-enhanced CT scan showing small left kidney with presence of multiple cysts (arrow) suggestive of dysplastic left kidney.



Figure 23: Intravenous urography film showing a case of dilated tortuous megaureter (white arrow) on the left side and a normal caliber ureter (black arrow) on the right side.



Figure 24: Intravenous urography film demonstrating bilateral ureterocele (arrow) larger in size on the right side.



Figure 25: Axial contrast-enhanced CT scan showing right sided ureterocele (white arrow) with calculi within the ureterocele (black arrow).

performed to exclude Vesicoureteral Reflux (VUR). In obstructed primary megaureter, VCUG demonstrates no VUR. US shows hydronephrosis and ureteral dilatation above the persistently narrowed distal aperistaltic segment. Real-time US reveals active peristaltic waves passing to and fro in the dilated ureter above the narrowed segment and disproportionate dilatation of the lower ureter relative to the upper ureter and renal pelvis [22].

Ureterocele

Ureterocele is ballooning of the distal end of the ureter. Ureteroceles may be associated with either a single or a duplex ureter.

Types:

(a) Simple or intravesical ureterocele- orifice of the ureter and the ureterocele are intravesical.

(b) Ectopic ureterocele – ureterocele lies in the submucosal of the bladder and may extend into the bladder neck or urethra.

On US, simple ureterocele is seen as a cystic intravesical mass. With real-time US, partial or complete collapse of a simple ureterocele secondary to ureteric peristalsis can be demonstrated. At VCUG, a collapsed simple ureterocele usually manifests as a rounded filling defect within the bladder. IVU shows the classic “cobra head” appearance consisting of a round or oval area of increased opacity surrounded by the radiolucent halo of the wall of the ureterocele (Figures 24 and 25). Ectopic ureterocele is almost invariably associated with a duplex collecting system and represents the distal portion of the ureter of the upper renal moiety. An ectopic ureterocele is more inferiorly located than a simple ureterocele.

Pseudoureteroceles [5] are rare and consist of two types – (i) coiled uniformly dilated distal segment of the ureter forming a submucosal mass (ii) ectopic ureter draining into a mesonephric duct cyst.

Duplex systems

Duplex system – a renal unit in which two pyelocalyceal systems are present and is associated with a single or bifid ureter (incomplete or complete duplication).

Nubbin Sign – refers to urographic appearance of a nonfunctioning or poorly functioning lower pole of a duplex kidney (Figure 26).

Bifid, Trifid and Multifid renal pelvis – refers to two, three or more renal pelvis that unite distal to the normally expected position of the ureteropelvic junction. The lower pole is generally larger than the other pelvis or pelvis and drains a larger number of calyces.

Bifid ureter – In the upper part there are two ureters while lower down the ureters join to form a single ureter. This joining may be extravascular (Y- junction) or intravesical (V-junction) (Figures 27 and 28). Double ureters – ureters remain completely separate to the point where they insert in the bladder or beyond. According to Weigert-R

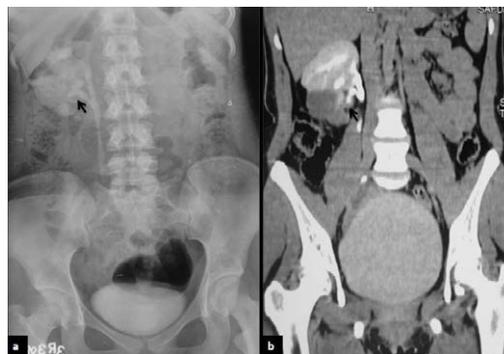


Figure 26: (a) Intravenous urography showing duplex right kidney with hydronephrosis (arrow) of inferior moiety and non-visualised left renal outline suggestive of left renal agenesis (b) Coronal contrast-enhanced CT scan of the same patient showing hydronephrosis (arrow) of inferior moiety.



Figure 27: Coronal contrast-enhanced MIP images showing incomplete left duplex system (black arrow) and hydronephrosis of upper moiety (white arrow) mimicking a cystic mass.



Figure 28: Intravenous urography film showing incomplete duplication (white arrow) on right side and complete duplication (black arrow) left side with ureterocele (curved arrow) of upper moiety ureter.

Meyer rule, the upper pole ureter opens below and medial to the lower-pole ureter (Figures 29 and 30).

Retrocaval ureter

Retrocaval or circumcaval ureter results from anomalous development of the infra-renal portion of inferior vena cava (IVC) and not from anomalous development of the ureter. There is anomalous development of the infrarenal IVC from the right posterior cardinal vein that is embryologically more medial. Classically, the ureter crosses posterior to the IVC and then travel medially and anteriorly to partially circumscribe the IVC [23].

Types: Type 1, the more common form, has severe or moderate hydronephrosis with extreme medial deviation of the middle ureteral segment, usually medial to the pedicle or across the midline at the L3 level resulting in “S” or “fish hook” deformity at the point of obstruction. In type 2 there is mild hydronephrosis and less medial deviation of the ureter. The ureter is noted to be sickle shaped at the level of obstruction [24]. On imaging, IVU shows characteristic reverse - J appearance due to medial deviation of the ureters with moderate proximal hydroureteronephrosis (Figure 31).

Vesicoureteral reflux

Vesicoureteral reflux (VUR) is the abnormal flow of urine from

the bladder into the upper urinary tract [25]. The pathophysiology behind this anomaly is either primary maturation abnormality of the vesicoureteral junction or a shorter intramural-submucosal segment of ureter. The primary diagnostic procedure for evaluation of VUR is voiding cystourethrography (VCUG), which should be performed after the first well-documented urinary tract infection [26]. The grade of reflux and whether reflux occurs during micturition or during bladder filling should be mentioned in the report [27-29] (Figure 32).



Figure 29: Intravenous urography film showing complete duplication right side with upper moiety (black arrow) with its ureter (black arrowhead) opening inferomedially and lower moiety (white arrow) with its ureter (white arrowhead) opening in normal location. Normal left pelvicalyceal system (curved arrow) is seen.

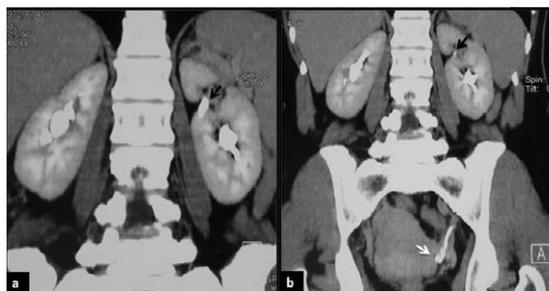


Figure 30: Contrast-enhanced CT coronal MIP images of a case of left sided complete duplication showing (a) two pelvicalyceal system (black arrow) and (b) two ureters (white arrow).



Figure 31: (a) Intravenous urography film showing reverse-J shape of upper right ureter (arrow) and moderate proximal hydroureteronephrosis suggestive of retrocaval ureter (b) Coronal contrast-enhanced MIP images confirming the same findings (arrow).

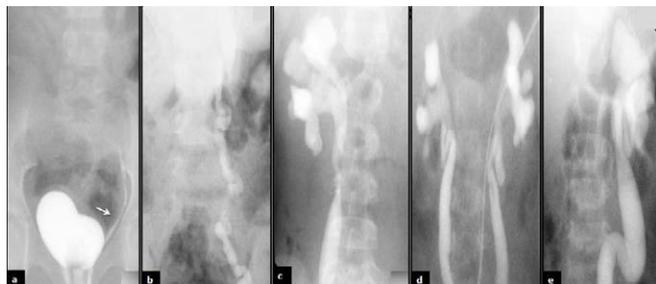


Figure 32: Voiding cystourethrography (VCUG) images show International Grading System of vesicoureteral reflux (VUR)—(a) grade I, (b) grade II, (c) grade III, (d) grade IV (e) and grade V. In grade I (a), contrast material reaches distal ureter (arrow). Contrast reaches renal pelvis in grades II to V, with increasingly severe pelvicaliceal and ureteral dilatation.

Grading of VUR: According to International Reflux Committee Study [30];

Grade I: Ureter only

Grade II: Ureter, pelvis, and calices; no dilatation; normal caliceal fornices.

Grade III: Mild or moderate dilatation or tortuosity of the ureter and moderate dilatation of the renal pelvis; no or slight blunting of the fornices.

Grade IV: Moderate dilatation or tortuosity of the ureter and moderate dilatation of the renal pelvis and calices; complete obliteration of the sharp angle of the fornices but maintenance of the papillary impressions in the majority of calices.

Grade V: Gross dilatation and tortuosity of the ureter; gross dilatation of the renal pelvis and calices; papillary impressions are no longer visible in the majority of the calices.

Ectopic ureter

Ectopic insertion of the ureter results from abnormal ureteral bud migration and usually results in caudal ectopia [31]. The opening of ectopic ureter in females is in the lower bladder, urethra, vestibule, and vagina and rarely into the uterus or a Gartner cyst. In males, the opening lies in the lower bladder, posterior urethra, seminal vesicle, vas deferens, ejaculatory duct or rarely into the rectum [32,33] (Figure 32). The fundamental difference between ureteral ectopia in females and in males is that in females, ectopic ureters can terminate at a level distal to the continence mechanisms of the bladder neck and external sphincter and thus may be associated with incontinence. In complete ureteral duplication with each segment having its own ureteral orifice in the bladder, the Weigert-Meyer rule applies. In these cases, the ureter draining the upper pole moiety frequently ends in an ureterocele, whereas reflux into the lower moiety typically occurs.

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