Nephrourological malformations as a radiological challenge

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Authors: M. VICENTE QUÍLEZ\textsuperscript{1}, C. Mejia Gomez\textsuperscript{2}, G. Anes\textsuperscript{1}, S. González Sánchez\textsuperscript{1}, A. L. Salgado Bernal\textsuperscript{1}, S. Sánchez\textsuperscript{1}, S. Gálvez García\textsuperscript{1}, M. Morán-Hevia\textsuperscript{1}, \textsuperscript{1}Oviedo/ES, \textsuperscript{2}Oviedo, oviedo/ES

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Learning objectives

- To revise and discuss the radiographic findings of congenital nephrourological malformations in children.

- To remark the importance of early diagnosis for an early treatment and the imaging methods most commonly used.

Background

The urinary tract is the system that is third most commonly affected by congenital malformations, preceded by the central nervous system and the cardiovascular system. Congenital abnormalities of the urinary tract are found relatively frequently, in around 0.5% of all pregnancies. The detection rate for urinary tract abnormalities in routine prenatal ultrasonography examinations is around 20 to 30% of the total number of malformations.

Congenital anomalies of the lower urinary tract are a significant cause of morbidity in infancy. Radiologic investigation is an important source of clinical information in lower urinary tract disorders but should not inconvenience the patient, expose the patient to unnecessary radiation, or delay surgical correction.

In pediatric patients with suspected underlying urologic structural anomalies, screening ultrasound is commonly the initial diagnostic study. If dilatation of the urinary tract is confirmed, voiding cystourethrography (VCUG) is performed to determine the presence of vesicoureteral reflux (VUR) and other causes of upper tract dilatation. If VUR is confirmed, follow-up with nuclear cystography or echo-enhanced cystosonography may be performed. If VUR is excluded, nuclear diuresis renography is the primary test for differentiating between obstructed and nonobstructed megaureter. Intravenous urography can be used to specifically identify an area of obstruction and to determine the presence of duplex collecting systems and an ureterocele. Computed tomography and magnetic resonance (MR) imaging are unsuitable for general screening but provide superb anatomic detail and added diagnostic specificity. MR imaging is mandatory in the evaluation of associated spinal anomalies. MR urography can demonstrate ectopic extravesical ureteric insertions, thereby providing a global view of the malformation.
Familiarity with anomalies of the lower urinary tract is essential for correct diagnosis and appropriate management.

Findings and procedure details

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3) RENAL HYPOPLASIA
4) RENAL DYSPLASIA
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1) NEPHROBLASTOMATOSIS

G) CONGENITAL TUMOURS

1) MESOBLASTIC NEPHROMA
A) RENAL ANOMALIES

1) SUPERNUMERARY KIDNEYS

Supernumerary kidney is a rare congenital anomaly. Bilateral supernumerary kidney is an even rarer anomaly.

Embryologically, supernumerary kidneys are formed by aberrant division of the nephrogenic cord into two metanephric blastemas with bifurcation of one bud. The supernumerary kidney may be either totally separate from the normal kidney or connected to it by loose areolar tissue.

Supernumerary kidneys have been reported to be associated with various congenital anomalies: ectopic ureteric opening, horseshoe and pseudo-horseshoe kidney, coarctation of the aorta, ureteral and vaginal atresia, complete duplication of the urethra and megaureter. Because of the wide range of combined congenital anomalies and the relative rarity of such cases, it is difficult to standardize a protocol for diagnosis and, thus, overdiagnosis with many unnecessary imaging tests is done. Therefore, diagnosis of patients with supernumerary kidneys represents a challenge.

In the literature, majority of these cases were diagnosed using many kinds of radiological imaging techniques, including ultrasound, CT scan, IVP, magnetic resonance imaging (MRI), CT angiography and dimercapto succinic acid (DMSA) and diethylene triamine pentacetate (DTPA) scans.

We believe that ultrasound, IVP and CT seem to be enough for the diagnosis in majority of the cases.

2) RENAL AGENESIS

If no vestige of renal tissue is formed, the condition is called agesenis.

Renal agenesis can be unilateral or bilateral.

Unilateral renal agenesis occurs about once in 1,000 individuals. It is sometimes associated with anomalies elsewhere in the genitourinary tract (e.g. uterine or vaginal duplication), in the gastrointestinal tract or in the cardiopulmonary system (VACTERL association).
Bilateral renal agenesis (Potter syndrome) occurs once in 4,500 -5,000 pregnancies. In this condition, oligohydramnios is uniformly present after about 20 weeks gestation and gives rise to the typical appearance of the face and extremities. The diagnosis of this condition is based primarily on the absence of renal outlines and the absence of the bladder.

Another feature we have found useful in diagnosing bilateral renal agenesis is the appearance of the fetal adrenal glands. Potter has noted flat discoid adrenals in the posterior abdomen in 50 cases of bilateral renal agenesis. They lie in the posterior abdomen and are characterized by a hypoechoic periphery and a thin central echogenic line.

Diagnosis of bilateral renal agenesis is suspected on prenatal ultrasound with severe oligohydramnios and absence of kidneys. It has very poor prognosis, usually stillborn or die shortly after birth.

Prognosis of a patient with unilateral renal agenesis depends on the state of the other kidney. A unique kidney usually develops normally and has a compensatory hypertrophy. In patients with unilateral renal agenesis, malformations of the lower urinary tract, genitals and other organs are often found.

3) RENAL HYPOPLASIA

Small congenital kidney buy otherwise similar to a normal organ, which has no pathological evidence of dysplasia.

Simple hypoplasia: Congenital small kidney resembling a normal one but smaller.

Segmental hypoplasia: Special type of unilateral renal hypoplasia. When associated with hypertension it is called Ask-Upmark kidney. The kidney is small and has a stenotic artery and fewer renal lobes.

Radiologically a small kidney is observed with a transverse notch on the kidney surface, adjacent to an elongated expansion and dilated renal pelvis. Differential diagnosis of chronic pyelonephritis image is very difficult.

4) RENAL DYSPLASIA
It is defined as abnormal metanephric differentiation. The kidney is represented by a nodule of tissue which has no morphologic or histologic resemblance to normal parenchyma.

Dysplastic kidneys are always associated with other congenital malformations of ureter or lower urinary tract.

They are classified into several types: hypoplastic dysplasia, multicystic dysplasia, aplasia, segmental dysplasia of the superior pole of a double collecting system and dysplasia associated with bilateral hydronephrosis.

Overall, renal dysplasia is the leading cause of end-stage renal disease in children.

4.1. MULTICYSTIC DYSPLASTIC KIDNEY

It is a form of renal dysplasia characterized by the presence of multiple macroscopic and noncommunicating cysts, usually peripheral, separated by dysplastic parenchyma and the absence of a normal pyelocaliceal system.

Cystic changes in multicystic kidneys are not visible until after about 20 weeks' gestation, although earlier, the kidneys may be more echogenic than usual.

Furthermore, the cystic dysplastic changes are not static postnatally, and the cysts may begin to disappear in infancy. Bilateral multicystic dysplastic kidneys are usually fatal in infancy.

The lesion is usually unilateral but it is bilateral in 20% of affected individuals.

The condition is associated with ureteral or ureteropelvic atresia, and the affected kidney is nonfunctional. Other terms used to describe this condition include multicystic kidney and multicystic renal dysplasia.

The most common cause of formation of a multicystic dysplastic kidney is the severe obstruction during embryogenesis due to an uretero-pyelic atresia in the metanephric phase in the development of the kidney.
The place of atresia determines the type of multicystic dysplastic kidney. The most common type is the one in which both the proximal ureter and renal pelvis and infundibula are atretic.

Multicystic dysplastic kidney is the most common cause of an abdominal mass in the newborn period and is the most common cystic malformation of the kidney in infancy.

5) INFANTILE POLYCYSTIC KIDNEY DISEASE

Most cases of infantile polycystic kidney disease are autosomal recessive, but the autosomal dominant form may be manifested in infancy as well. The disease is associated with a spectrum of pathologic changes involving both the kidneys and liver, including bilateral renal enlargement. The kidneys contain innumerable microscopic cortical and medullary cysts representing dilated distal tubules and collecting ducts.

Cysts may also be seen in the liver, spleen, and pancreas, usually in patients with the dominant form of the disease. The classic liver abnormality of the recessive form is periportal fibrosis.

The cysts characteristic of this disease provide acoustical interfaces that cause increased echogenicity and poor corticomedullary differentiation on US images. Sometimes a peripheral hypoechoic cortical rim is also evident. Macroscopic cysts are not usually present, but may be. Milder forms of recessive polycystic kidney disease are not detectable in utero.

When present in young children, the adult type of polycystic kidney disease can be similar in appearance to the infantile variety.

6) RENAL ECTOPIA: ABNORMALITIES OF MIGRATION AND FUSION

Renal ectopia and abnormalities of migration and fusion of the kidneys are studied together due to their common characteristics. They are common, appearing in 1/500 children.

6.1. RENAL ECTOPIA
Renal ectopia describes kidneys which are congenitally located in abnormal anatomic positions.

Renal ectopia is classified as follows:

Pelvic: kidney located in the true pelvis.

Iliac: kidney located either in the iliac fossa or opposite the crest of ilium.

Abdominal: the kidney is "fixed" above the crest of the ilium or below the level of L2 and L3.

6.2. CROSSED RENAL ECTOPIA

Probably the best classification has been that devised by McDonald and McClellan in four varieties of crossed renal ectopia:

1. Crossed ectopy with fusion.
2. Crossed ectopy without fusion.
3. Crossed ectopy with solitary kidney.
4. Bilateral crossed ectopia.

Unilateral fused type (inferior ectopia), which is by far the most common, the crossed ectopic kidney is always inferior with its upper pole fused to the lower pole of the other normally situated kidney. Normally the lower kidney is malrotated, looking both renal pelvis toward the midline.

Differential diagnosis: Urographically this condition may be confused with crossed ectopia without fusion or with an extremely mobile kidney which lies close to the midline or barely crosses it. Surgical exploration is necessary in some cases to verify the fusion. An ectopic kidney with a completely duplicated pelvis and a functionless kidney in the other side also might be confused with crossed renal ectopia with fusion.

6.3. HORSESHOE KIDNEY
It is the most common type of renal fusion and it appears in 1/600 births. There is a fusion of the lower poles of both kidneys on the midline below the isthmus, which is usually located anterior to the aorta and inferior vena cava.

The hilum in horseshoe kidney is a complex structure, usually with multiple renal vessels leaving the aorta.

There is a slight increase in the incidence of Wilms tumor, renovascular hypertension and adenocarcinoma.

### 6.4. RENAL MALROTATION

Anomalies in position of the kidney are rather common. No doubt the condition most frequently seen is malrotation in respect to the vertical axis of the kidney. This can be due to either incomplete or excessive rotation. Because of the anterior or lateral position of the renal pelvis, the upper part of the ureter often appears to be displaced laterally.

Malrotacion can be unilateral or bilateral.

The incidence of malrotation in ectopic and fused kidneys is much higher than in normally situated kidneys.

### B) ANOMALIES OF THE URINARY SYSTEM

#### 1) URINARY TRACT DUPLICATIONS

Duplication of the upper urinary tract is the most common congenital malformation of the urinary tract.

It is one of the commonest congenital obstructive urological diseases, occurring in around 1% of live births. Females are more affected than males, and this condition occurs unilaterally in 83% to 90% of the cases.

Duplications usually follow a benign course and represent an ultrasound incidental finding.
1.1 DUPLEX COLLECTING SYSTEM

A) Incomplete duplication: It is twice more common than complete. The bifurcation can be high (bifid pelvis), anywhere in the ureter or next to the bladder (bifid distal ureter).

B) Complete duplication: Each segment has its own hole opening in the bladder, according to the rule Weigert-Meyert, the ureteral orifice of the superior system is inserted into the medial and lower bladder to its normal location at the mouth of the lower ureter. Children with complete duplication have higher incidence of urinary tract infections, VUR, cortical scarring and obstruction.

The ureter that drains the upper unit commonly presents relative obstruction, thus leading to dysplasia of the upper renal pole. On the other hand, the ureter that drains the lower unit is inserted topically in the vesical trigone, and may present ureteral-vesical reflux. Dilatation of the upper unit of the collecting system may simulate a large renal cyst at the upper pole.

The upper renal segment is involved in 85% of the cases and ureteroceles occur between 24 and 47% of the cases. Ureteroceles have been reported in up to 50% of renal duplications, with hydronephrosis of the upper pole.

The diagnosis of pyeloureteral duplication can be made prenatally by means of ultrasonography.

The findings that lead to suspicion of pyeloureteral duplication are identification of two separate renal poles, dilatation or cystic areas in an upper or lower pole, a dilated ureter and an anechoic cystic structure projecting into the bladder that is suggestive of ureteroceles.

2) CONGENITAL MEGACALYCOSIS

It consists in unobtrusive increased size of the chalices associated with hypoplasia of the medullary pyramids.

This feature is important because it can be confused radiographically with obstructive hydronephrosis or vesicoureteral reflex.
C) ANOMALIES OF THE URETER

1) URETEROPELVIC JUNCTION (UPJ) OBSTRUCTION

Obstruction to the passage of urine from the pelvis into the proximal ureter.

It is a common condition and it is often bilateral, although its severity is not symmetrical.

It results from a variety of causes. Intrinsic causes include:

I: partial replacement of the ureteropelvic junction muscle by collagen.

II: abnormal arrangement of the junction muscle bundles, causing
dysmotihity.

III: mucosal folds or polyps.

Extrinsic abnormalities, such as aberrant vessels and adventitial bands, are often seen at surgery, but are rarely the primary cause of obstruction.

The degree of pelvicahiectasis in ureteropelvic junction obstruction is variable and can be quite severe. In 30% of cases, it is present bilaterally.

It can be associated with other malformations of the urinary tract such as vesico-ureteral reflux and obstruction of the ureterovesical junction.

Imaging tests:

1. Ultrasound: Usually the first test done before the suspected diagnosis of UPJ obstruction.

Ultrasonography without the presence of other associated malformations shows the presence of hydronephrosis without signs of ureteral dilatation and normal bladder.

Hydronephrosis can be described as mild (renal pelvis dilatation without calicial dilation), moderate (renal pelvis and calicial dilation) or severe (renal pelvis and calicial dilation with thinning of the cortex).
The degree of parenchymal thinning is in relation to the severity and duration of the obstruction and the degree of atrophy.

If a compatible cystic mass with hydronephrosis was observed, we must perform a VCUG as the next diagnostic test. It is done to rule out reflux, urethral obstruction, or both.

Intravenous urography (IVU) also must be performed to confirm the cause and location of the obstruction and to determine the function of the kidneys.

2. **Simple RX:** If the stenosis of the ureteropelvic junction is severe, simple rx can show the presence of a soft tissue mass in the renal area, with displacement of intestinal gas.

3. **Voiding cystourethrogram (VCUG):** The ultrasound finding of hydronephrosis may be due to UPJ obstruction, vesicoureteral reflux or obstruction due to reflux, so VCUG is always done.

It is frequent the association between UPJ obstruction and reflux. If the ureter is tortuous, this suggests that the dilatation of the renal pelvis is not only due to reflux. If reflux and UPJ obstruction are present, surgical management changes: we must do first pyeloplasty, because reflux may disappear.

4. **Intravenous urography:** It determines qualitatively the renal function, the degree of obstruction and shows the anatomy.

**Natural history and prognosis:**

Some patients have a spontaneous improvement of obstruction, but most of them are invariable for many years, maintaining a dilated collecting system. Most pediatric urologists perform pyeloplasty in patients with severe UPJ obstruction, and follow-up to those with only mild or moderate stenosis, performing an ultrasound study periodically (every 6 months or a year).

Indications and timing of surgery for UPJ obstruction are continually under review.

**2) CONGENITAL OBSTRUCTION OF MIDDLE URETER**
Because of its rarity, middle ureter obstruction is often confused with a low stenosis of the UPJ obstruction or an aperistaltic long segment of a primary megaureter.

### 3) URETEROVESICAL JUNCTION (UVJ) OBSTRUCTION

Distal ureter obstruction or vesicoureteral junction obstruction can be congenital or acquired:

- **Congenital causes:** Primary megaureter, refluxing primary megaureter, primary megaureter with bladder sacculation, simple ureterocele, ectopic ureterocele and ectopic ureter.

- **Acquired causes:** Stenosis due to ureteral reimplantation, neurogenic bladder, complications of infection and stone formation.

### 4) PRIMARY MEGAURETER

Megaureter is a generic term indicating the presence of an enlarged ureter with or without concomitant dilatation of the upper collecting system. The normal ureter in children rarely exceeds 5 mm in diameter. In practice, a ureter with a diameter of 7 mm or more should be considered a megaureter.

Primary megaureter is an inherently compound term that includes all cases of megaureter due to an idiopathic congenital alteration at the vesicoureteral junction.

There are three major categories of primary megaureter:

1. Obstructed primary megaureter
2. Refluxing primary megaureter
3. Nonrefluxing unobstructed primary megaureter

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

Primary megaureter:
1. **OBSTRUCTED PRIMARY MEGAURETER**

There is dilatation above a short (0.5- 4cm), aperistaltic, normal-caliber juxtavesical section of a normally inserted ureter. The ureteral orifice and the submucosal tunnel are normal. The normal ureter proximal to the aperistaltic segment dilates because of relative obstruction. This phenomenon is somewhat similar to achalasia and Hirschsprung disease. The cause of distal ureteral aperistalsis is not known. It may be due to a paucity of ganglion cells in the connective tissue that surrounds the distal ureter; however, aganglionosis has not been proved to be the problem. Tokunaka et al reported a muscular derangement and increased interstitial connective tissue in the nondilated terminal ureteral segment. All investigations with electron microscopy demonstrated an excess of collagen between muscle cells.

2. **REFLUXING PRIMARY MEGAURETER**

It is caused by a short or absent intravesical ureter, congenital paraureteric diverticulum, or other derangement of the vesicoureteral junction. Lee et al assumed that the marked increase in collagen and significant decrease in smooth muscle could be major contributing factors in the pathogenesis of refluxing primary megaureter.

3. **NONREFLUXING UNOBSCTURED PRIMARY MEGAURETER**

There is neither reflux nor stenosis of the vesicoureteral junction, but the ureter is dilated beginning at a point just above the bladder. The cause of this phenomenon is unknown, but several theoretic explanations have been given based on transitional (prenatal to postnatal) renal physiologic and histoanatomic features of the developing ureter. Most primary megaureter in neonates falls into this category.

**Imaging tests:**

1. **Ultrasound:** hydronephrosis and ureteral dilatation is observed above the distal segment.

2. **Voiding Cysto-urethrogram (VCUG):** It is essential to perform Voiding Cysto-urethrogram (VCUG) to differentiate between reflux, ureterovesical junction obstruction or a combination of both.
In the vesicoureteral reflux all the ureter is dilated, however primary megaureter has a portion of normal caliber in the ureterovesical junction.

3. **Intravenous urography (IVU):** After performing ultrasound and VCUG, an IVU is often made to verify the amount of vesicoureteral junction stenosis and renal function.

**Natural history and treatment:**

Serial ultrasound studies (every 6-12 months) are made in non-operated patients in order to detect any increase of hydronephrosis.

Severe primary megaureter normally requires ureteral reimplantation.

5) **RETROCAVALURETER**

It is the only anomaly of genitourinary tract that is essentially limited to the right side.

The condition is explained embryologically as the result of the "anomalous" persistence of the embryonic right posterior cardinal vein as the adult vena cava. Instead of pursuing its normal paravertebral course, the ureter is directed toward the midline so that it overlies the vertebral column in the region of the third and fourth lumbar vertebrae. Lying against the vertebral column, it passes behind the vena cava, encircles it, and then resumes its normal position. In doing so, a sickle-shaped curve is imparted to the ureter.

It would be more correct to speak about pre-ureteral vena cava as it is the cava which has an abnormal location.

This condition has no pathognomonic signs or symptoms. It becomes a clinical problem only because the hydronephrosis may result from obstruction of the ureter as it passes behind and around the vena cava. Any patient who has pyelectasis on the right side and dilatation involving only the upper part of the ureter may be suspected of having retrocaval ureter.

Urographic diagnosis may be rather difficult, as it is often impossible to fill the ureter adequately with contrast medium.
An opaque catheter inserted into the ureter may establish the diagnosis, since it may be seen to pass toward the midline, so that it overlies the vertebral column in the region of the third and fourth lumbar vertebrae. In this manner a sickle-shaped curve is imparted to the catheter. A satisfactory ureterogram will disclose a gentle curving of the midportion of the ureter, which is close to the vena cava, and with the convexity directed superiorly and medially.

Surgery is only indicated when there is a significant obstruction.

6) URETEROCELE

Ureteroceles represent cystic dilatation of the intravesical segment of the ureter.

They may be associated with either a single or a duplex ureter. The congenital defect is the obstruction of the meatus, and the ureterocele is simply a hyperplastic response to this obstruction. The outer wall is composed of bladder epithelium and the inner wall of ureteral epithelium, with connective tissue and muscle fiber in between.

A ureterocele may be as small 1 cm, or it may fill the entire bladder and prolapse through the urethra. Ureteral duplication is present in about 75% of patients with ureteroceles.

Ureteroceles may be either simple or ectopic.

5.1. In simple ureteroceles, stenosis of the distal end of the normally positioned ureteral orifice leads to ballooning of the segment immediately above it, forming the ureterocele. In young patients, the stenosis is presumed to be congenital, although in older children and adults it can be secondary to an inflammatory stricture.

5.2. An ectopic ureterocele is the cystlike protrusion into the bladder lumen of the dilated submucosal distal portion of an ectopic ureter. It 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. It is usually unilateral and is far more common in girls than in boys.

7) ECTOPIC URETER
Ectopic insertion of the ureter stems from abnormal ureteral bud migration and usually results in caudal ectopia. Normally, the primitive ureteral bud travels cephalad, whereas the wolffian duct, from which it originates, travels caudal. However, if the ureteral bud fails to separate from the wolffian duct, it may be carried into a more caudal position than normal. Consequently, the opening of the ureter becomes caudally ectopic and, in the female, inserts into the lower bladder, urethra, vestibule, or vagina.

In males, it empties into the lower bladder, posterior urethra, seminal vesicle, vas deferens, or ejaculatory duct. In very rare instances, it can empty into the rectum.

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. Approximately one-half of female patients with ectopic ureters present with a classic history of continuous dribbling incontinence despite what appears to be a normal voiding pattern.

An ectopic ureter can drain a single kidney, but about 70% are associated with complete ureteral duplication. In complete ureteral duplication with each segment having its own ureteral orifice in the bladder, the Weigert-Meyer rule applies. This rule states that the ureteral orifice of the upper pole moiety inserts into the bladder medial and inferior to both its normal location and the orifice of the ureter draining the lower renal segment. In these cases, the ureter draining the upper pole moiety frequently ends in an ureterocele, whereas reflux into the lower moiety typically occurs.

7) PRIMARY VESICOUРЕTERAL REFLUX

VUR is the abnormal flow of urine from the bladder into the upper urinary tract.

In the majority of cases, it occurs as a result of a primary maturation abnormality of the vesicoureteral junction or a short distal ureteric submucosal tunnel in the bladder that alters the function of the valve mechanism. VUR may be an isolated anomaly or associated with other congenital anomalies such as posterior urethral valves or complete duplication of the urinary tract.

Reflux predisposes to renal infection (pyelonephritis) because it carries bacteria from the bladder to the upper urinary tract. The majority of pediatric patients who develop renal scars after a urinary tract infection have VUR, and higher grades of reflux are associated with an increase in parenchymal scarring.
Detection of VUR in neonates and infants is particularly important because these patients are more predisposed to the formation of renal scars than are older children. Reflux nephropathy is a common cause of renal failure; therefore, it is important that this condition be detected as early as possible to allow prompt prophylactic antibiotic treatment and hopefully reduce the risk of scarring and reflux nephropathy. Reflux is also the most common cause of antenatal hydronephrosis, being responsible for 40% of intrauterine cases.

**Imaging tests:**

1. **Ultrasound:** It is used to assess the renal parenchyma, collecting system and any other associated anomalies such as ectopic ureterocele. However, if it is normal it does not exclude reflux.

2. **VCUG:** We should always perform a cystography, to document the presence and degree of VUR.

In VCUG we appreciate retrograde course of the contrast from the bladder to the ureter (unilateral or bilateral). It can be classified according to the time when reflux occurs in: active (only during urination) and passive (occurs during the filling phase of the bladder during cystography); and according to the level reached and the degree of dilation of the collecting system in five degrees (International Classification of VUR):

**GRADES OF VESICoureTERAL REFLUX**

- **grade 1:** reflux limited to ureter
- **grade 2:** reflux into renal pelvis
- **grade 3:** mild dilatation of ureter and pelvicalyceal system.
- **grade 4:**
  - tortuous ureter with moderate dilatation
  - blunting of fornices but preserved papillary impressions
- **grade 5:**
tortuous ureter with severe dilatation of ureter and pelvicalyceal system loss of fornices and papillary impressions

It is important to note that each side may have a different grade of reflux.

Spontaneous resolution usually occurs at 5-6 years of age in girls and little earlier in children.

**Treatment:** We have to consider surgical reimplantation of the ureter if it has not disappeared after a reasonable time, if infections occur frequently or it is difficult the fulfillment of prophylactic antibiotic treatment.

**D) ANOMALIES OF THE BLADDER**

1) **BLADDER AGENESIS**

Agenesis of the bladder is an extremely rare congenital anomaly. Most infants with the anomaly are stillborn and have other urogenital tract anomalies as well as neurologic and orthopedic disorders.

The cause of agenesis of the bladder is uncertain. Because the hindgut is normal in these infants, it may be assumed that embryologic division of the cloaca into the urogenital sinus and anorectum proceeded normally. Bladder agenesis may be the result of secondary loss of the anterior division of the cloaca, perhaps owing to a lack of distention with urine caused by failure of incorporation of the mesonephric ducts and ureters into the trigone, thus preventing urine from accumulating in the bladder. In most cases in which the bladder is absent, the ureters have the same course as ureters with ectopic vestibular openings.

2) **BLADDER DUPLICATION**

Complete duplication of the bladder and urethra is a rare anomaly.

Duplication of the bladder may occur in the sagittal or coronal plane. The most common form is sagittal duplication, in which two bladders lie side by side and are separated by a fold of peritoneum and loose areolar tissue. Each bladder receives the ureter of the ipsilateral kidney and is drained by its own urethra, the ureter and urethra lying side by
side. In some cases, only one bladder communicates with a single urethra, leaving the opposite side obstructed with no outlet. Most obstructed systems are associated with renal dysplasia and nonfunction.

3) BLADDER DIVERTICULA

Congenital bladder diverticula that are not associated with posterior urethral valves or neuropathic bladder are unusual but not rare and occur almost exclusively in boys.

Bladder diverticula can be unilateral or bilateral and are caused by congenital bladder muscular anomalies. A diverticulum that occurs at the ureterovesical junction is usually called periureteric diverticulum, classically known as Hutch diverticulum, and it is often associated with VUR. This is because the presence of the diverticulum alters the normal slanted insertion of the ureter into the bladder.

**Imaging tests:**

Diverticula are easy to diagnose at Voiding Cystourethrography (VCUG). At US, diverticula appear as round or oval anechoic fluid collections that arise from the base of the bladder or around the ureteric orifice.

4) PRUNE-BELLY SYNDROME

The term "prune belly" refers to a lax, wrinkled abdominal wall, that results from the absence of rectus muscles, which is frequently associated with other anomalies.

It is a specific constellation of anomalies consisting of three major findings associated with a number of other respiratory, gastrointestinal, musculoskeletal, and cardiovascular anomalies. Bilateral, nonpalpable undescended testes are present, and there is an abnormal urinary tract characterized by tortuous, dilated ureters; a megalocystic, dilated prostatic urethra; and renal dysmorphism. This syndrome is also called Eagle-Barrett syndrome and Triad syndrome.

5) URACHAL ANOMALIES

The urachus develops from the superior portion of the urogenital sinus and connects the dome of the bladder to the allantoic duct during fetal life. The urachus is located
behind the abdominal wall and anterior to the peritoneum in the space of Retzius. Before birth, the urachus is obliterated and becomes a vestigial structure known as the medial umbilical ligament.

In the absence of complete obliteration, the urachus persists as either a patent urachus, urachal cyst, urachal sinus, or urachal diverticulum.

A persistent urachus frequently coexists with congenital lower urinary tract obstruction such as posterior urethral valves or Prune-Belly syndrome. It may also coexist with ventral abdominal wall defects such as omphalocele. Although adenocarcinoma of the urachus is rare, it has been reported in patients as young as 15 years of age.

A patent urachus manifests at longitudinal US as a tubular connection between the anterosuperior aspect of the bladder and the umbilicus.

A urachal cyst forms when both the umbilical and vesical ends of the urachal lumen close while an intervening portion remains patent and fluid filled. Urachal cysts usually remain obscure until complicated by infection or bleeding. An uncomplicated urachal cyst appears as a collection of simple fluid localized in the midline of the anterior abdominal wall, between the umbilicus and the pubis and often contiguous with the bladder dome.

**Imaging tests:**

Diagnostic evaluation should begin with US, which usually allows localization of the mass and delineation of its limits. A urachal cyst may become infected and demonstrate features of mixed echogenicity at US, attenuation and signal intensity that deviate upward from those of water at CT and MR imaging, respectively, and thickening of the urachal wall.

The differential diagnosis includes bladder diverticulum, vitelline cyst, mesenteric cyst, Meckel diverticulum, umbilical hernia, and even ovarian cyst.

**E) ANOMALIES OF THE URETHRA**

**1) POSTERIOR URETHRAL VALVES**
Posterior urethral valves are by far the most common congenital obstructive lesion of the urethra, occurring only in phenotypic boys. Young initially classified posterior urethral valves into three types, but it is now clear that there is only one type (formerly called type I). Posterior urethral valves result from the formation of a thick, valvelike membrane from tissue of wolffian duct origin that courses obliquely from the verumontanum to the most distal portion of the prostatic urethra. In essence, the valve is a diaphragm, but because it is more rigid along its line of fusion, progressive distention during voiding causes it to become bilobed or saillike. The Young type III valve is a congenital, disklike membrane that is oriented across the urethral lumen at the level of the membranous urethra. An orifice of variable size is typically located centrally.

**Imaging tests:**

VCUG is the best imaging technique for the diagnosis. Radiologic findings include dilatation and elongation of the posterior urethra and, occasionally, a linear radiolucent band corresponding to the Valve.

**F) CONGENITAL PRETUMOROUS CONDITIONS**

**1) NEPHROBLASTOMATOSIS**

Nephroblastomatosis refers to diffuse or multifocal involvement of the kidneys with nephrogenic rests.

Nephrogenic rests are foci of metanephric blastema that persist beyond 36 weeks gestation and have the potential for malignant transformation into Wilms tumour.

Nephrogenic rest are found incidentally in 1% of infants. It is currently believed that nephrogenic rests give rise to approximately 30-40% of Wilms tumours. Nephrogenic rests are found in up to 99% of bilateral Wilms tumours.

There are two pathologic subtypes of nephrogenic rest:

1. Perilobar rest (90%)
2. Intralobar rest (10%): more associated with Wilms tumour.

**Imaging tests:**
1. Ultrasound:

Ultrasound may demonstrate hypoechoic nodules but is less sensitive than MR imaging and CT. Diffuse nephroblastomatosis is usually seen as reniform enlargement with a thick peripheral rind of tissue that may show striated enhancement. On ultrasound, the enlarged kidney may have diffusely decreased echogenicity.

2. CT:

At CT, macroscopic nephrogenic rests appear as low-attenuation peripheral nodules with poor enhancement relative to that of adjacent normal renal parenchyma.

3. MRI:

Typical signal characteristics include:

- **T1:** the nodules demonstrate low-signal-intensity foci
- **T2:** the nodules demonstrate low-signal-intensity foci

Treatment and prognosis:

Treatment for nephrogenic rests is controversial. Some investigators recommend chemotherapy, whereas others maintain that close serial radiologic evaluation of enlarging masses is sufficient.

Differential diagnosis:

Renal lymphoma can mimic the appearance of nephroblastomatosis but is unusual in infants and young children.

G) CONGENITAL TUMOURS

1) MESOBLASTIC NEPHROMA

Mesoblastic nephroma, also sometimes known as a congenital mesoblastic nephroma (CMN) or fetal renal hamartoma, is in general a benign renal tumour. It typically occurs in utero or in infancy.

It is the commonest neonatal renal tumour.
Diagnosis is usually in the antenatal period or immediately after birth. The tumour can account for ~3-6% of all renal neoplasms in children. Approximately 50% occur during the neonatal period and 80% of cases are reported within the first month of life.

Most common clinical presentation is a palpable abdominal mass, with haematuria occurring less frequently.

**Subtypes:**

There are two main pathological variants:

1) Classic mesoblastic nephroma.

2) Cellular mesoblastic nephroma:

- more heterogeneous in appearance on imaging
- tends to be larger and presents later in infancy
- may exhibit aggressive behaviour including vascular encasement and metastasis

**Associations:**

- Polyhydramnios
- Fetal hypercalcaemia

**Imaging tests:**

Non specific and not an imaging modality of choice but if performed incidentally in a neonate, may demonstrate a soft tissue mass displacing bowel. Calcification is rare.

**1. Ultrasound:**

Sonographic appearance can vary depending on the pathological variant. In general it is a well-defined mass with low-level homogeneous echoes. The presence of concentric echogenic and hypoechoic rings can be a helpful diagnostic feature. A more complex pattern due to haemorrhage, cyst formation and necrosis can also be seen and tends to favour the cellular variant. Colour Doppler interrogation may show increased vascularity.

Antenatal ultrasound may also show evidence of associated polyhydramnios.

**2. CT:**
Usually not performed in an antenatal situation. Tends to be generally of low attenuation at CT. More content required.

3. MRI:

Best modality for cross sectional imaging antenatally and can better assess anatomical relationships.

Unless complicated necrosis and haemorrhage (both generally uncommon), general signal characteristics within the mass include:

**T1**: homogeneously hypo-intense

**T2**: homogeneously hypo-intense

**Treatment and prognosis:**

The majority are benign tumours and have a favourable outcome. The cellular variant can at times be aggressive. As a surgical option, a nephrectomy usually suffices as treatment.

**Images for this section:**
**Fig. 1:** Renal agenesis. Absence of kidney in right renal fossa. Left kidney presents a normal size, morphology and echogenicity, which shows a volume of 177 cm³, without dilatation of the urinary tract. Renal scintigraphy shows the existence of one functional kidney.
Fig. 2: Renal atrophy. Asymmetry of renal size secondary to right renal atrophy with the presence of a huge calculus occupying all the calycial groups and renal pelvis, conditioning dilatation of the upper calycial group. Renal cortex is thinned and measures 5 mm. It has an approximate volume of 53 cm³. Left kidney presents normal characteristics, with a volume of 90 cm³.
Fig. 3: Renal atrophy (same patient) Left kidney presents normal characteristics, with a volume of 90 cm³. Renal scintigraphy shows hypofunction of the right kidney.
**Fig. 4:** Renal hypoplasia. Hypoplasia of the left kidney, which has a volume of 7.5 cm³. Right kidney has a volume of 102 cc, it presents a normal morphology and echogenicity, with a good corticomedullary differentiation.
**Fig. 5:** Renal hypoplasia (same patient) Right kidney has a volume of 102 cc, it presents a normal morphology and echogenicity, with a good corticomedullary differentiation.
Fig. 6: Renal hypoplasia (same patient) Renal scintigraphy demonstrates less functionality of the left kidney and a complete function of the contralateral one.
**Fig. 7:** Renal hypoplasia. Asymmetry in the size of both kidneys, showing the right kidney a discrete alteration in morphology, probably because of the presence of scars on his cortical and measures 79x49 mm. Left kidney measures 110x55 mm and does not present any anomaly.
Fig. 8: Renal hypoplasia (same patient) Left kidney measures 110x55 mm and does not present any anomaly.
Fig. 9: Multicystic dysplastic kidney. Dysplastic right kidney which shows an increase in its size with the presence of multiple cysts (the largest measures 45 mm), without dilatation of the urinary tract.
**Fig. 10:** Multicystic dysplastic kidney. Large cystic mass in the right kidney cell that replaces the kidney. It presents at least three cysts whose size is over 40 mm and the largest measures 46 mm.
Fig. 11: Polycystic kidney disease. Enlarged kidneys with hyperechoic cortex and loss of corticomedullary differentiation. Presence of small cortical cysts.
**Fig. 12:** Crossed- fused renal ectopia. Absence of left kidney on the left renal fossa. On the right renal fossa, a large kidney (13 cm) with two renal hila is identified.
Fig. 13: Crossed- fused renal ectopia (same patient)
Fig. 14: Crossed- fused renal ectopia. Ectopic left kidney on the right renal fossa fused to the lower pole of the right kidney. The upper kidney shows a slight pyelocalyceal ectasia, with a pelvis whose anteroposterior (AP) axis is 10 mm. The lower kidney presents significant pyelocalyceal dilation (23 mm in the AP diameter), with thinning of the cortex. Dilatation of the renal pelvis suggests vesicoureteral reflux, identifying only one ureter dilated distally located on the left side of the bladder, with an important peristalsis and changes in its diameter during the study.
Fig. 15: Crossed- fused renal ectopia (same patient). Dilatation of the renal pelvis suggests vesicoureteral reflux, identifying only one ureter dilated distally located on the left side of the bladder, with an important peristalsis and changes in its diameter during the study.
Fig. 16: horseshoe kidney. Horseshoe kidney with atrophy of the right kidney and left pyelocalycial ectasia. It exists renal parenchyma that crosses the midline.
**Fig. 17**: Horseshoe kidney (same patient) Both kidneys crossing the midline. References: Fig 17a: Radiopaedia.org. Fig 17b: Hospital Universitario Central de Asturias, Oviedo, SPAIN.
Fig. 18: Horseshoe kidney with stenosis of the left ureteropelvic junction. Slight dilatation of the pelvis of the right kidney, measuring 15 mm in AP diameter. Important hydronephrosis affecting the left kidney with marked dilatation of the renal calyces which shows a loss of its normal morphology, associated with renal cortical thinning. Ureters do not present dilatation. CT urography with volumetric reconstruction corroborated echographic findings.
**Fig. 19:** Horseshoe kidney with stenosis of the left ureteropelvic junction (same patient). CT urography with volumetric reconstruction where echographic findings are corroborated.
**Fig. 20:** Kidney malrotation. Right malrotated kidney with the major axis in horizontal position, but it presents normal size and morphology, without dilatation of the urinary tract.
Fig. 21: Duplex collecting system with ureterocele. Left duplicated collecting system. Upper renal group shows a loss of cortico-medullary differentiation with small cysts, which suggests renal dysplasia. It is observed ureterohydronephrosis of the top group with distal ureterocele. The thick wall and the echogenic content in the whole urinary tract indicates pyonephrosis. References: Fig 21a, 21b(left): Hospital Universitario Central de Asturias, Oviedo, SPAIN. Fig 21c (right): Radiopaedia.org
**Fig. 22:** Duplex collecting system with ureterocele (same patient). Cystography: Filling defect on the left lower region of the bladder. Taking into account the previous ultrasound study, it is compatible with a left ureterocele. The rest of the bladder is smooth-walled.
**Fig. 23:** Duplex collecting system. Left duplicated collecting system with cortical thinning in the upper pole and dilatation of both calycial groups measuring 15 mm the top group and 5mm lower one.
Fig. 24: Duplex collecting system (same patient). Both ureters are visualized until the entry into the bladder.
Fig. 25: Bilateral ureteropelvic junction obstruction. Severe right hydroureter with an anteroposterior diameter of the renal pelvis of 47 mm. The cortex is significantly thinned. Moderate left pyelocalycial dilation with an AP diameter of the renal pelvis of 19 mm. Ureters are not dilated.
**Fig. 26:** Bilateral ureteropelvic junction obstruction (same patient). Ureters are not dilated.
Fig. 27: Bilateral ureteropelvic junction obstruction (same patient). Antegrade pyelography performed through a nephrostomy catheter: Contrast agent is inserted through a right nephrostomy catheter with distal end in the superior calycial group. Only the upper calycial group and renal pelvis are filled of contrast, which present a severe dilatation, and no contrast agent is present in the ureter. Delayed Rx of abdomen (90 minutes later), contrast is identified in bladder.
Fig. 28: Ureteropelvic junction obstruction (same patient). Coronal and axial CT and volumetric reconstruction after the administration of contrast medium. Important right pyelocalycial dilation, with a renal pelvis measuring 45 mm in AP diameter. It is not possible to identify the ureter and it has not been achieved an adequate filling of the urinary tract with contrast agent, since the renal elimination is very delayed due to the stenosis. Drainage catheter in the right renal fossa.
Fig. 29: Bilateral ureterovesical junction stenosis. Ultrasound and MR urography (T2 in coronal plane and reconstruction of the urinary tract). Bilateral hydronephrosis grade V, with loss of normal morphology of renal calyces and a thinned renal cortex. The AP diameter of the right renal pelvis is 13 mm and the left one 6 mm, with dilated ureters since their entry into the bladder, with maximum diameters of 21 mm on the right side and 13 mm on the left side.
**Fig. 30:** Bilateral ureterovesical junction stenosis (same patient). Dilated ureters since their entry into the bladder, with maximum diameters of 21 mm on the right side and 13 mm on the left side.
**Fig. 31:** Bilateral ureterovesical junction stenosis (same patient). MR urography: There is a retrograde dilatation of the entire urinary tract.
**Fig. 32:** Bilateral ureterovesical junction stenosis (same patient). Cystography: No vesicoureteral reflux was observed.
**Fig. 33:** Primary megaureter. Slight right pelvic ectasia with 8 mm which is associated with dilatation of the ureter throughout its course except in its distal area where it tapers gradually (star) not being visible at the entry into the bladder.
Fig. 34: Primary megaureter. Cystography of the same patient: no vesicoureteral reflux is identified.
Fig. 35: Circumcaval or retrocaval ureter. The hydronephrosis may result from obstruction of the ureter as it passes behind and around the vena cava.
Fig. 36: Ureterocele in a duplex collecting system. On the floor of the bladder, there is an oval image that follows from the distal portion of the ureter of the lower half of the right kidney, which has a size of approximately 7 x 5 mm.
Fig. 37: Ureterocele in a duplex collecting system (same patient). Cistography: Bladder has a normal morphology, identifying a filling defect consistent with the ureterocele described in ultrasonography. Vesicoureteral reflux in the lower half of the right kidney, grade IV. Intravenous urography: dilation of the distal ureter.
**Fig. 38:** Ectopic ureterocele. Distal ureter which does not ends in bladder and probably empties into the prostatic urethra.
Fig. 39: Grades of vesicoureteral reflux.
Fig. 40: Vesicoureteral reflux: Grade I. Cistography. Duplex collecting system with reflux of both left ureters identifying both ureters until the joint between its upper and middle third. Presence of trabeculated walls with pseudodiverticula.
Fig. 41: Vesicoureteral reflux: Grade II right and III left. Cistography.
Fig. 42: Vesicoureteral reflux: Grade III right and IV left. Cistography.
Fig. 43: Vesicoureteral reflux. Ultrasound: Left kidney shows moderate pyelocalyceal dilation, measuring 10 mm the AP pelvic diameter. The ureter is dilated (8 mm maximum size) and tortuous, visualized until its end where it is sharpened. The findings suggested vesicoureteral reflux as the first possibility, which was corroborated by the cistography.
Fig. 44: Vesicoureteral reflux: Grade IV left. Cistography of the same patient.
Fig. 45: Vesicoureteral reflux: Grade V. Cistography.
**Fig. 46:** Bladder diverticulum. Thin-walled bladder with a diverticulum in its right posterolateral wall.
Fig. 47: Bladder diverticulum. Cystography reveals the presence of a bladder diverticulum showing no VUR. Intravenous urography: Ureters with normal calibers, ending the left one in a bladder diverticulum.
Fig. 48: Hutch diverticulum. Great diverticulum in the left vesicoureteral junction that is associated with vesicoureteral reflux, grade II.
Fig. 49: Urachal cyst. At the dome of the bladder there is a small cystic formation where the urachus is formed and measures 5 x 10 mm.
**Fig. 50:** Posterior urethral valve. Urethrocystography: Bladder presents a trabeculated and thickened wall with small pseudodiverticula. There is also a significant dilatation of the posterior urethra secondary to a stenosis located between the prostatic and the membranous urethra. References: Fig 50a: © 1996-2014 The Children's Hospital of Philadelphia. Fig 50b: Hospital Universitario Central de Asturias, Oviedo, SPAIN.
Fig. 51: Mesoblastic Nephroma: Right renal mass that measures 45x96x45 mm. It is a well defined and hyperechoic lesion, which conditions dilation of the upper calyceal group.
**Fig. 52:** Mesoblastic Nephroma (same patient). CT scan with IV contrast reveals a right renal mass, it is moderately heterogeneous with hypodense areas but without necrosis or cystic changes. It occupies the central area of the kidney where renal hilum is affected, although renal vein is permeable. This lesion contacts the right hepatic lobe and the psoas muscle.
Conclusion

In pediatric patients with suspected underlying nephro-urologic structural anomalies, screening ultrasonography is commonly the initial diagnostic study, either in the prenatal or postnatal period.

The radiologist plays an important role in the diagnosis, therapeutic decision, prognosis and follow up of nephrourological anomalies. Simple radiology and ultrasound scan are really important tools for early diagnosis.

Personal information

Dra. María Vicente Quílez.
Hospital Universitario Central de Asturias.
Calle Carretera de Rubín, s/n, 33011.
Oviedo (Asturias, SPAIN).

Email: mariavquilez@gmail.com

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