7.1 Part I

7.1.1 Urinary Tract by Ultrasound: The BUK Approach

A systematic approach to the urinary tract by ultrasound: the BUK approach.

7.1.1.1 Introduction

In the past, most imaging evaluations of the infant or child with signs or symptoms of a urinary tract abnormality began with a plain X-ray of the abdomen, a so-called KUB, with the letters standing for, from “top to bottom,” kidneys, ureters, and bladder. Today, however, imaging usually begins with an ultrasound of the fetus, and the standard approach is to survey the urinary tract of the infant and child from the “bottom to top” that is, to begin with the bladder and then to look at the ureters and finally the kidneys, in other words, a BUK instead of a KUB approach.

Radiologists often assume that the bladder is normal or that the fluid-filled structure in the pelvis is automatically the bladder. This leads to mistakes. For example, bladder exstrophy, a rare condition in which the bladder forms as a plate on the anterior pelvic wall, is diagnosed by noticing the absence of a normal bladder on fetal ultrasound. However, 50% of children born with bladder exstrophy are diagnosed postnatally because people fail to check if a normal bladder is present.

Other fluid-filled structures in the pelvis can be mistaken for the bladder. These include large ureteroceles, ovarian cysts or masses, and the dilated (obstructed) vagina.

Learning Objectives

- To describe the normal findings of the pediatric urogenital tract
- To list the potential causes of urinary tract dilatation
- To discuss the subtypes of congenital female tract anomalies
The ureters start at the renal pelvis, course down on the psoas muscles, cross the internal iliac vessels, and then enter the bladder posterolaterally. The fetal ureters are typically invisible unless they are dilated. The more dilated the ureter, the more tortuous it becomes because dilation increases width and also length. Extremely dilated ureters can be mistaken for dilated loops of bowel (Fig. 7.2).

In children, dilated ureters are often first noticed as tubular anechoic structures located posterolaterally while imaging the bladder. Peristalsing boluses of urine or “ureteral jets” are seen entering the bladder by color Doppler. There are no established values for normal or abnormal ureteral widths by ultrasound. However, a normal ureter in a child is 3–4 mm in diameter. Normal fluid-filled ureters can be seen peristalsing. Visualizing the ureter does not mean that it is abnormal.

Ureteral dilation is due to vesicoureteral reflux (VUR), obstruction, or both reflux and obstruction. Vesicoureteral reflux is the reverse flow of urine from the bladder to the kidney. Reflux of infected urine leads to pyelonephritis. Dilated ureters continue to peristalse and the changing diameter is commonly seen by ultrasound. The most common test to distinguish obstruction from reflux is a voiding cystourethrogram (VCUG), also called a micturating cystourethrogram (MCUG). If the VCUG shows reflux, the dilatation of the ureters and kidneys is due to reflux. If the refluxed material does not drain well at the end of the test, then either reflux coexists with obstruction. Drainage films are important to make this distinction [1]. If there is no reflux, then the dilation is due to obstruction.

The most common congenital cause of ureteral obstruction is primary megaureter. This is due to a functional obstruction at the orthotopic ureterovesical junction. Seventy
percent of patients with primary megaureter improve with time. The width of the diameter of the ureter can help to predict resolution [2]. Obstructed ureters may also be ectopic, as what occurs with the upper pole of a duplex kidney or, more rarely, a single ectopic ureter.

When the reflux is severe, the megacystis-megaureter association may lead to enlargement of the bladder (megacystis) and dilation of the ureter or ureters (megaureter) [3]. The bladder enlarges because there is recycling of urine so that the bladder never fully empties and is constantly refilling with refluxed urine from the ureters, and it eventually dilates. Severe megacystis-megaureter can lead to urinary retention.

7.1.1.4 Kidneys

Kidneys normally migrate from the fetal pelvis to the normal renal fossae at 4–6 weeks gestation. The renal fossae are paraspinous, centered at the lumbar vertebral level of L1–L2. Kidneys are first seen in utero at 10 weeks gestation. Nearby landmarks include the echogenic spine, and the suprarenal hypoechoic boomerang-shaped adrenal glands, and the liver and spleen. The adrenal cortex is hypoechoic and the medulla echogenic. The normal suprarenal or adrenal gland can be up to one-third the size of the kidney in the second trimester.

When the kidney does not migrate to the normal location, it is ectopic. Since kidneys normally ascend from the fetal pelvis to the renal fossae, ectopic kidneys are typically somewhere in-between. Occasionally, when the liver or diaphragm has not formed normally, ectopic kidneys may be present in the chest. Kidneys may also fuse while ascending, forming cross-fused ectopic kidneys, horseshoe kidneys, or lump kidneys.

When a kidney is absent from the renal fossa in utero, 50% are ectopic and 50% are absent [4]. When there is a solitary kidney, 23% are associated with other congenital anomalies, including those in the VACTERL association. Thirty percent of these patients have vesicoureteral reflux (personal unpublished data).

The normal echogenicity of a kidney in an infant or child is more variable than in an adult. In adults, normal kidneys are hypoechoic compared to the liver. In infants and children, normal kidneys may be relatively isoechogenic or hypoechoic. In newborns and premature infants, relatively echogenic kidneys may still be normal. In addition, kidneys are separated into lobes, with a central hypoechoic medulla and a peripheral relatively hypoechoic cortex. The adjacent lobes create an undulating cortex which is described as normal fetal lobation. The normal indentations between renal lobes are distinguished from scarring as the indentation is between the pyramids, versus centered at the tip of the pyramids in scarring. The hypoechoic pyramids become isoechogenic to adjacent renal parenchyma over time and are occasionally confused for masses or cysts (Fig. 7.3).

Compared to adults, renal masses are relatively uncommon in children. Renal tumors tend to be very large. The most common solid tumor in an infant is a mesoblastic nephroma. Despite their large size, they tend to be benign. The most common childhood renal tumor is Wilms’ tumor.

In the past three decades, the survival rate of children with these tumors approaches 90% [5]. Simple cysts are relatively uncommon compared to adults. If more than one simple cyst is identified, cystic renal disease should be considered, including autosomal dominant polycystic disease. Cystic tumors, including cystic nephroma or focal cystic dysplasia, are uncommon. When the whole kidney develops as abnormal, cysts with no normal renal parenchyma, a multicystic dysplastic kidney is formed.

The collecting tubules in the medullary pyramids drain into the minor calyces which drain into the major calyces and into the renal pelvis. When there is urinary tract dilation, there is progressive dilation of the pelvis and calyces. This is described by many terms, including pyelectasis in utero and hydronephrosis in fetuses and children.

There are many ways of describing the dilated urinary tract. The only system which unifies prenatal and postnatal imaging into one system is the Urinary Tract Dilation System (UTD) which was created by radiologists, urologists, and nephrologists. The six common descriptions are (1) anterior posterior renal pelvic diameter, (2) calyceal dilation, (3) ureteral dilation, (4) renal parenchymal thickness, (5) renal parenchymal echogenicity, and (6) bladder appearance. In fetuses, oligohydramnios which may be due to urinary tract abnormalities is also described [6–8].

Prenatal urinary tract dilation or hydronephrosis is common, present in 1–4% of pregnancies. Most prenatal hydronephrosis is mild and resolves [9, 10]. The most common pathology to cause pelvic and calyceal dilation without ure-
teral dilation is ureteropelvic junction obstruction. This is due to an intrinsic obstruction at the ureteropelvic junction or occasionally is due to a crossing anomalous renal vessel. The vessel causes obstruction when there is increased urine production and the vessel kinks the ureteropelvic junction. These obstructions, unlike intrinsic ureteropelvic junction obstructions, are intermittent and cause pain only during obstruction.

The greater the degree of prenatal or postnatal hydronephrosis, the greater the likelihood of ureteropelvic junction obstruction. The likelihood of vesicoureteral reflux is the same regardless of the degree of dilation [11]. MAG-3 and MRU examinations are both useful for quantifying the degree of obstruction and relative renal function [12].

There are certain entities that tend to affect the entire urinary tract: the bladder, ureters, and kidneys. The three most common entities are duplex kidneys with complete ureteral duplication, posterior urethral valves, and prune belly syndrome.

When there is complete ureteral duplication, the upper pole ureteral orifice is ectopic (the Weigert-Meyer rule). The ectopic upper pole ureter terminates anywhere from the trigone of the bladder, inferiorly and medially to the base of the bladder (the so-called ectopic pathway). In boys, ectopic ureters terminate above the urethral sphincter and Wolffian duct remnants including the ejaculatory ducts, vasa, and seminal vesicles. In females, ectopic ureters can extend below the urethral sphincter or into the vagina and can cause daytime and nighttime wetting. When the ectopic ureter is obstructed or ends in a ureterocele, it dilates. A ureterocele is cystic dilation of the intramural portion of the ureter, which appears as a thin-walled cyst in the bladder and is continuous with the ureter. The more ectopic the ureter, the more dysplastic the associated upper pole parenchyma. The lower pole is the analog of the single system ureter. There may be coexisting lower pole reflux or ureteropelvic junction obstruction.

Posterior urethral valves are thin leaflets which cause obstruction in the posterior urethra, at the base of the verumontanum, in boys. The valves cause a variable degree of bladder outlet obstruction, bladder wall thickening, and variable hydroureteronephrosis. Often the degree of left and right urinary tract dilation is asymmetric. The hydroureteronephrosis may be due to reflux, obstruction, or both. VCUG/MCUG diagnoses both the posterior urethral valves and reflux. Although this diagnosis can be suspected in utero, milder cases present in childhood with infection or milder hydroureteronephrosis. In severe cases, the kidneys may be dysplastic and high pressures may lead to fornical rupture, urinomas, and urine ascites (Fig. 7.4).

Prune belly syndrome can mimic posterior urethral valves. The triad of cryptorchism, dysplastic kidneys, and severe reflux can appear radiographically similar to posterior urethral valves. The bladder is dilated but thinner than in boys with valves, the ureters are severely dilated, and the kidneys are dysplastic. However, children with prune belly syndrome have lax anterior abdominal wall musculature and no posterior urethral valves despite a dilated posterior urethra. The anterior urethra may also be dilated, the so-called megalourethra.

7.1.1.5 Summary

If one systematically looks at the bladder, ureters, and kidneys (BUK), the urinary tract will be evaluated completely. The most common mistake is that people assume that the bladder is normal. Ureteral dilation should be evaluated carefully, as dilation distinguishes hydronephrosis (typically due to ureteropelvic junction obstruction) from hydroureteronephrosis (typically caused by primary megaloureter or reflux). An MCUG/VCUG distinguishes reflux from obstruction. When evaluating the kidney, carefully assess its

![Fig. 7.4](a–c) Posterior urethral valves, depending on severity, can affect the appearance of the bladder, ureters, and kidney. In this example of severe posterior urethral valves, (a) the bladder wall is thick and irregular; (b) there is severe hydronephrosis (H) characterized by pelvic and peripheral calyceal dilation, thinning and cystic dysplasia of the renal parenchyma, and ureteral dilation (U); and (c) A VCUG demonstrates posterior urethral valves (arrowhead) with severe dilation of the posterior urethra
presence, echogenicity, and for the presence and degree of urinary tract dilation. The common entities which affect the bladder, kidney, and ureters are duplex kidneys with complete ureteral duplication, posterior urethral valves, and prune belly syndrome.

7.2 Part II

7.2.1 Female Genital Tract

Ultrasound is the most important imaging technique in childhood pelvic imaging. MRI is only used in complex cases.

7.2.1.1 Normal Development

The paramesonephric (Müllerian) ducts are paired structures that undergo fusion and resorption that form the uterus, proximal fallopian tubes, and upper vagina. The distal segments of the Müllerian ducts fuse in the midline forming the uterovaginal canal. The development of the uterus is complete by 12 weeks' gestation. Interruption of normal fusion and resorption of the Müllerian ducts give rise to a broad spectrum of Müllerian duct anomalies. Lower part of the vagina and ovaries arises from the urogenital sinus and the primitive yolk sac, respectively [13].

7.2.1.2 Uterus

In the newborn girl, the female internal genitalia are prominent due to the maternal and placental hormone exposure in utero. The endometrium is normally clearly seen as an echogenic stripe at the center of the uterus. After 2–3 months until puberty, the uterus is relatively small and tubular in configuration (2–4 cm) with the cervix being as large and as thick as the fundus (<1 cm). The endometrial lining is relatively inconspicuous. During puberty the fundus of the uterus enlarges and becomes bulbous in contour. The postpubertal uterus is around 5–8 cm in length and has a pear-shaped appearance. The endometrial lining undergoes cyclic changes associated with the menstrual cycle [14] (Fig. 7.5).

7.2.1.3 Congenital Abnormalities

Female genital tract anomalies may result from agenesis, hypoplasia, or abnormalities related to disorders in lateral fusion, vertical fusion, or resorption. Congenital Müllerian anomalies occur in around 1.5% of females and are strongly associated with renal anomalies (in 30–50%) [15]; however congenital Müllerian anomalies are not associated with ovarian anomalies.

Ultrasound examination of the uterus should be performed in all female neonates with MCDK, unilateral renal dysplasia, single kidney, or adrenal hyperplasia due to the high risk of associated female tract anomalies. Additional MRI can be valuable as MRI provides great anatomical detail of both the endometrium and outer contour of the uterus [14].

Imperforate hymen is the most common obstructive anomaly of the female pelvis [16]. Usually there are no other associated abnormalities. Ultrasound shows fluid distention of the vagina with a lesser degree of cervical/uterine distention. Echogenic debris within the fluid is due to mucous secretions in neonates and blood in postmenarcheal girls. This fluid-debris level helps to distinguish a dilated vagina from the bladder or other cystic pelvic masses.

Partial or complete failure of resorption of the septum between the two Müllerian ducts results in a septate uterus. The external surface has a normal configuration and there are two endometrial cavities. The septum is complete if it extends to the cervical os. There is a single cervix.

Bicornuate uterus results from partial non-fusion of the Müllerian ducts. The external surface is deeply indented. A common challenge is to differentiate between a bicornuate uterus and a septate uterus. This distinction is clinically important because the septate uterus carries a high risk of miscarriage. The absence of the indentation of the external contour (<1 cm cleft) with the presence of duplicated endometrial cavity is the key feature to diagnose a septate uterus rather than a bicornuate uterus [17].

Complete non-fusion leads to a didelphys uterus. Two cervices are inevitably present. Unicornuate uterus occurs when there is a complete or near-complete arrested development of one Müllerian duct.

Fig. 7.5 (a–c) Sagittal image of a neonate (a), 4-year-old (b) and 18-year-old (c) girl, illustrates the relative large uterus in the newborn, the more tubular appearance in a child and the pear-shape appearance in the adolescent girl.
development of one of the Müllerian ducts resulting in a single-horned uterus with a single round ligament and fallopian tube [16]. Presence of endometrium in a rudimentary horn is a clinically important finding because there is an increased prevalence of endometriosis, pelvic pain and higher risk of problems during pregnancy (miscarriage, ectopic pregnancy, preterm labor, and uterine rupture).

Complete arrested development of both Müllerian ducts is also known as the Mayer-Rokitansky-Kuster-Hauser syndrome. In this syndrome there is a congenital absence of the proximal vagina, cervix, and uterus with normal external genitalia [17]. Primary amenorrhea is the most common clinical presentation.

7.2.1.4 Ovaries
Ovaries can be distinguished from other pelvic structures by identifying small follicle cysts within the ovaries that can be seen from birth. The size of the follicles decreases during a hormonally quiescent period. The conspicuity and number of follicle cysts increases after adrenarche (around 10–11 years of age) and puberty [18].

7.2.1.5 Ovarian Cysts and Other Lesions
In the fetus and in the neonatal period, large ovarian cysts are occasionally detected due to maternal and placental hormone exposure. Most neonatal cysts are asymptomatic and resolve within the first year of life. Complications from larger ovarian cysts include torsion, bleeding, and strangulation in an inguinal hernia [14].

7.2.1.6 Ovarian Torsion
Torsion is caused by partial or complete rotation of the ovary and/or fallopian tube around the infundibulopelvic ligament. In the neonate ovarian torsion occurs due to a cyst. During childhood torsion is most often caused by an ovarian mass (cystic teratoma). The sonographic features of ovarian torsion include enlarged ovary with peripherally located follicles. Often the ovary is located in the midline of the lower abdomen with the uterus deviated toward the affected side. Sometimes the whirlpool sign of twisted vascular pedicle can be seen. Doppler findings can be widely variable related to the dual blood supply of the ovaries (uterine and ovary artery) [14].

7.2.1.7 Ovarian Neoplasm
Ovarian neoplasms are extremely rare in the neonate and infant. The most common benign ovarian tumor in childhood is a cystic teratoma, accounting more than 90% of all benign ovarian tumors. Most cystic teratomas have a complex appearance on sonographic imaging that depends on the relative amount of fat, calcium, serous fluid, and hair within the lesion [19].

7.3 Part III
7.3.1 Male Genital Tract
7.3.1.1 Normal Development
The descent of the testis occurs at two stages. The intra-abdominal descent takes place at 10–15 weeks’ and the inguinoscrotal descent occurs at 26–40 weeks’ gestation [13]. At ultrasound, the testis is a homogenous bean-shaped structure, which has an echogenic mediastinum as one of its landmarks. Blood flow is normally homogeneous.

7.3.1.2 Congenital Abnormalities
Ultrasound is often used to identify the location of the undescended testis (also called cryptorchidism) with variable success. Some people advocate the use of MRI. However, the appearance of a smaller, ectopic testis is often similar to lymph node with either ultrasound or MRI. The echogenic mediastinum and the homogeneous blood flow can be useful ultrasound characteristics to distinguish the testis from an inguinal node.

7.3.1.3 Testicular Torsion
Testicular torsion can occur in newborns and infants. They will present with an enlarged, discolored scrotum. The testis is often unsalvageable because the torsion is discovered too late. Extravaginal torsions are more common in infancy, compared to intravaginal torsions, which occurs during puberty. Ultrasound findings of testicular torsion are complete absence of blood flow in the testis in comparison to the other side. With prolonged torsion the affected testis becomes hypoechoic. Most often the testis lies more cranially within the scrotal sac and the twisted spermatic cord can be seen. The most important part of the examination is to compare the findings with the normal side (Fig. 7.6) [20].

Fig. 7.6 Sagittal ultrasound image shows an inguinal hernia in a female infant that contains uterus and both ovaries.
with the herniated bowel loops suggests viability. The bowel, ovaries, and fallopian tubes are the organs that are most commonly incarcerated. Ultrasound is an accurate method to determine the content of inguinal hernia. Peristalsis with the herniated bowel loops suggests viability.

7.3.1.4 Inguinal Hernia

Indirect, sliding hernia inguinalis is an uncommon finding in male and female neonates, especially in premature infants (up to 30%) due to a patent processus vaginalis (Fig. 7.7). The bowel, ovaries, and fallopian tubes are the organs that are most commonly incarcerated. Ultrasound is an accurate method to determine the content of inguinal hernia. Peristalsis with the herniated bowel loops suggests viability.

Key Points
- Ultrasound is an essential tool in the evaluation of the urogenital tract in infants and children.
- Look systematically at the bladder, ureters, and kidneys to evaluate congenital urinary tract anomalies.
- The finding of dilated ureter distinguishes UPJ obstruction from reflux/primary megaureter.
- In the newborn girl, the female internal genitalia are prominent due to the maternal and placental hormone exposure in utero.
- Female genital tract anomalies are strongly associated with renal anomalies.
- Indirect, sliding hernia inguinalis is a common finding in male and female neonates, especially in premature infants (up to 30%) due to a patent processus vaginalis.

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