Congenital Anomalies of the Kidneys, Collecting System, Bladder, and Urethra

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Education Gap

Several congenital anomalies of the kidney and urinary tract are incidental findings. An understanding of when to suspect and how to diagnose, manage, and use timely and appropriate investigations and consults is necessary.

Objectives

After completing this article, readers should be able to:

1. Develop an awareness of various congenital anomalies of the renal system, including embryology, prevalence, and risk factors.
2. Describe the clinical presentation and management of renal and urinary tract anomalies, including which anomalies warrant further evaluation and the timing and utility of imaging modalities.
3. Develop an awareness of genetic syndromes affecting the kidneys and urinary tract with associated extrarenal manifestations.

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ABBREVIATIONS

ARPKD	autosomal recessive polycystic kidney disease
CAKUT	congenital anomalies of the kidney and urinary tract
ESRD	end-stage renal disease
MCDK	multicystic dysplastic kidney
PUV	posterior urethral valve
RBUS	renal and bladder ultrasonography
UPJ	ureteropelvic junction
UTI	urinary tract infection
UVJ	ureterovesical junction
VCUG	voiding cystourethrogram
VUR	vesicoureteral reflux

INTRODUCTION

Congenital anomalies of the kidney and urinary tract (CAKUT) include a wide spectrum of anomalies, with a reported incidence of up to 2% of births. (1) CAKUT account for almost one-fourth of all birth defects. (2) These are major causes of kidney disease in children and account for more than 40% of end-stage renal disease (ESRD). CAKUT are usually detected by routine prenatal ultrasonography, although some cases are not diagnosed until adulthood. (3)

When renal disease is suspected, a complete physical examination should be performed with particular focus on accurate blood pressure measurement; the abdomen for palpable kidneys and a distended bladder; the genitalia for the position of the meatus, penile abnormalities, and urine flow; and the back for signs of neural tube defects.

Genetics play a major role in the etiology of CAKUT as family history is identified in 10% to 50% of affected children (Table). (4)(5) Similarly, 23% of asymptomatic first-degree relatives are found to be affected on screening. (4) Many environmental factors, such as maternal medication exposure, folate and iron deficiency, and diabetes mellitus, also contribute to the expression of CAKUT.

PATHOGENESIS

Human kidneys develop in 3 stages: the pronephros, the mesonephros, and the metanephros. The pronephros and the mesonephros involute, and the metanephros

forms the kidney starting at the fifth week of embryogenesis. The kidneys lie in the pelvis initially, where they are adjacent to each other and facing ventrally. By the eighth week of embryogenesis, the kidneys migrate to their permanent location in the lumbar region, and by the tenth week, they start producing urine. At the time of birth, each healthy kidney measures approximately 4.0 to 4.5 cm in the longest dimension. With the co-expression of multiple signaling and transcription factors, there are reciprocal interactions between the metanephros and ureteric epithelium, resulting in the formation of nephrons and the collecting system. In the meantime, the bladder develops from the urogenital sinus. Malfunctioning of otherwise well-controlled signaling pathways and transcription factors can result in CAKUT. (6)

ANOMALIES OF THE KIDNEY

Renal Agenesis

Renal agenesis or aplasia is a missing kidney, with the former a result of embryologic lack of initiation of development and the latter of failure to progress after initial induction and subsequent involution. The incidence of unilateral renal agenesis is 1 in 1,000 to 1 in 3,000 live births, with males and the left side disproportionately affected. (7)(8) Diagnosis is usually made on routine prenatal ultrasonography without obvious physical examination signs or symptoms, although there is a 50% rate of coexisting urogenital abnormalities. During fetal development, the contralateral kidney undergoes hypertrophy as a compensatory response to the effects of a decreased nephron count. The single hypertrophied kidney hyperfiltrates and develops renal injury marked by hypertension, proteinuria, and reduced glomerular filtration rate. (9) Hence, serial ultrasonography and routine screening for hypertension and proteinuria into adulthood are warranted. In a study, the median age at renal injury was 14.8 years in a cohort of 151 pediatric patients with a solitary functioning kidney. (10) Bilateral agenesis has a male predominance and is rare, occurring in 1 in 7,000 to 1 in 10,000 births. (11) Those affected generally do not survive after birth because oligohydramnios leads to the Potter sequence, with multiple anomalies of the lung, limb, and face.

Renal Dysplasia

A dysplastic kidney occurs due to abnormal differentiation after initiation of normal development and is one of the most common causes of pediatric ESRD. Prevalence ranges from 0.1% on ultrasonography screening to 4% in infant autopsies. (12) Unilateral dysplasia is more common than bilateral dysplasia, and as many as 50% to 75% of cases are associated with urinary tract anomalies, such as horseshoe kidney, ureteral duplication, hydroureter, ureteropelvic junction (UPJ) and ureterovesical junction (UVJ) obstruction, and vesicoureteral reflux (VUR). (13) Although renal and bladder ultrasonography (RBUS) can capture a small hyperechoic kidney, histologic analysis can show undifferentiated tissue, nephron cell structure, and the presence of cartilage and cysts. Unilateral dysplasia has a good prognosis, barring the severity of comorbidities of concomitant anomalies. However, unilateral renal dysplasia also may lead to death due to the Potter sequence or chronic kidney disease.

Multicystic Dysplastic Kidney

The most common cause of a congenital solitary kidney is unilateral multicystic dysplastic kidney (MCDK). The prevalence of MCDK is 1 in 2,400 to 1 in 4,300 live births, and it disproportionately affects males and the left side. (14) Most MCDKs involute without intervention (60% by age 10 years) and have minimal functional nephrons or renal tissue. (15) Most cases of MCDK are detected prenatally via ultrasonography, and they are the second most common cause of newborn abdominal masses, with hydronephrosis being the first. There are many isolated cysts of differing sizes with dysplastic stroma associated with an atretic ureter on gross examination. The contralateral kidney may be affected by low-grade VUR in 15% to 28% of patients or by UPJ obstruction in 4% of patients. (15)(16) Serial postnatal RBUS is warranted to monitor for contralateral renal abnormalities, growth, and involution. If involution occurs, further RBUS is not necessary. Malignant transformation (Wilms tumor or renal cell carcinoma) is rare; therefore, nephrectomy of MCDK is no longer the treatment of choice. (14) Conservative management with regular screening for hypertension and proteinuria is needed in patients with MCDK.

Autosomal Recessive Polycystic Kidney Disease

Autosomal recessive polycystic kidney disease (ARPKD) is an inheritable genetic disorder with an incidence of 1 in 10,000 to 1 in 40,000 live births. (17) Although ARPKD can present later in life, it is typically thought of as an "infantile" disease. Prenatal ultrasonography shows bilateral enlarged hyperechoic kidneys and possibly oligohydramnios. Although 30% of newborns with ARPKD die after birth, postinfancy survival is 80% at 10 years. (17) Most infants develop difficult to control hypertension, and 20% to 45% have ESRD by age 15 years. (18) On examination at birth, patients have bilateral firm abdominal masses and possibly respiratory distress. Laboratory work will show increasing blood urea nitrogen and serum creatinine levels, indicative of renal insufficiency. The bilateral renal cystic disease and associated hepatic fibrosis lead to renal and hepatic

insufficiency. Although renal impairment can be variable, all patients have hepatic fibrosis.

Diagnosis is made by imaging and the infant having at least 1 of the following: both parents without renal cysts, a sibling with ARPKD, consanguineous parents, or hepatic fibrosis. Management of the infant may include respiratory support, hypertension control, or fluid and electrolyte support. Nephrectomy may be an option if the enlargement is severe enough to affect respiratory or digestive function. As the patient grows older, dialysis for advanced kidney disease or surgery for portal hypertension may be required.

Autosomal Dominant Polycystic Kidney Disease

Autosomal dominant polycystic kidney disease is the most common inheritable genetic renal cystic disease, with an incidence of 1 to 2 in 1,000 live births. (17) Genetic testing is usually reserved for unclear diagnosis or if a family member is being considered for kidney donation for transplant.

Although this disease generally presents later in life (30–40 years of age), it can present in infancy with a poor prognosis. Clinical presentations may range from abdominal masses in the newborn to abdominal or flank pain between the third and fifth decades of life. Hematuria, hypertension, or gastrointestinal symptoms may be seen in adults. Hypertension occurs in half of the patients by their third decade of life and in almost all patients with ESRD. Other complications include hematuria, urinary tract infection (UTI), cerebral aneurysm, or heart disease, with mitral valve prolapse as the most common valvular disorder. (17) Extrarenal manifestations occur most commonly in the liver, followed by cysts in the seminal vesicles, arachnoid membrane, and pancreas. (19) In contrast to ARPKD, macroscopic cysts are seen on ultrasonography. Asymptomatic children with affected parents should be screened annually with blood pressure measurements and urinalysis. Management includes supportive care for respiratory or renal issues in infancy and long-term medical management of renal insufficiency and hypertension. Patients presenting with flank or abdominal pain and/or fever should be evaluated for infection, urinary stones, or tumors. Nephrectomy may be performed in selective symptomatic patients.

ANOMALIES OF THE COLLECTING SYSTEM

Hydronephrosis

Hydronephrosis is the dilation of the renal pelvis and calyces secondary to increased urine retention. In 50% to 75% of the cases, hydronephrosis is transient or physiologic. Other causes, such as UPJ obstruction and VUR, have an incidence of 10% to 30%. Less common causes include UVJ obstruction, MCDK, or posterior urethral valves (PUVs). (20) A complete physical examination should be performed with particular focus on the lower abdomen for a distended bladder, genitalia for penile abnormalities and urine flow, and the back for signs of neural tube defects, which may be associated with a neurogenic bladder. (21) Blood work assessing kidney function is generally withheld, except in bilateral hydronephrosis and severe cases.

RBUS is useful in determining the severity of hydronephrosis, for which multiple classification systems exist. The urinary tract dilation classification system is widely used, which includes parameters on anteroposterior renal pelvic diameter, calyceal dilation, parenchymal thickness and appearance, and ureter and bladder abnormality. (20) The Society for Fetal Urology classifies diseases on the anteroposterior renal pelvic diameter and on the appearance of the kidney, ureter, and bladder. (22) Postnatally in a low-risk patient with unilateral hydronephrosis, RBUS can be performed 7 to 10 days after birth. (20)(23) Exceptions to delayed imaging include cases of oligohydramnios, urethral obstruction, bilateral high-grade dilation, and follow-up concerns. (23) In bilateral hydronephrosis or a high-risk infant with bladder dilation, delivery at a center with pediatric urology support is recommended. A high-risk maternal/fetal medicine specialist should be involved when there are findings of oligohydramnios. If the lower urinary tract is involved and there is bladder distention and/or bilateral hydroureteronephrosis, voiding cystourethrography (VCUG) should be performed to rule out PUVs, and initiation of antibiotic prophylaxis is recommended. (20)

Vesicoureteral Reflux

VUR is the most common pediatric urologic condition, affecting up to 30% of children with a febrile UTI and 0.4% to 1.8% of the general pediatric population. (24) In infants with prenatal hydronephrosis, 10% to 20% will have VUR. It is seen in 50% of children of parents with VUR and 25% of siblings with VUR. (25) Primary reflux is an embryonic anatomical defect and largely spontaneously self-resolves with elongation of the intravesical tunnel at the UVJ as the child grows. Secondary VUR occurs when increased intravesical pressures lead to retrograde urine flow past the normal UVJ. This can be due to disorders such as neurogenic bladder, PUVs, or bowel and bladder dysfunction in anatomically normal children. Correction of bowel and bladder dysfunction may resolve VUR. (26)

Management of VUR is controversial and may include antibiotic prophylaxis and/or surgical correction, taking into consideration patient age, severity, renal scarring, and parental preference. A meta-analysis that included the RIVUR (Randomized Intervention for Children with Vesicoureteral Reflux) and PRIVENT (Prevention of Recurrent Urinary Tract Infection in Children with Vesicoureteric Reflux and Normal Renal Tracts) studies demonstrated that although antibiotic

prophylaxis can reduce UTIs, it does not reduce kidney scarring. (27) Furthermore, antibiotic-resistant bacteria increasingly cause infections while the patient is on prophylaxis. Complications of VUR include pyelonephritis, renal scarring, hypertension, and chronic kidney disease.

Duplicated Collecting System

Duplications of the collecting system are the result of abnormal embryonic origins. Complete duplication is a double ureter, each draining separately into the bladder, whereas partial duplication presents with a bifid ureter with a single drain into the bladder. (28) VUR is usually present in the lower moiety of a duplicated collecting system, whereas the upper moiety is associated with an obstructive ectopic ureter, with or without a ureterocele. The prevalence of ureteral duplications is less than 1% of the general population and affects females 2 to 4 times more often than males. (29) Complications of a duplicated collecting system include VUR, obstruction, ectopic ureteral insertion, and ureteroceles. (28) Ultrasonography is the mainstay of diagnosis, both before and after birth, and is the most common modality for incidental findings of duplication. (30) VCUG can help determine VUR and voiding patterns. Management differs widely, based on anatomical abnormality, function, and VUR severity.

ANOMALIES OF THE BLADDER

Congenital Neurogenic Bladder

Bladder dysfunction results from disruptions of the normal voiding process by neurogenic, anatomical, or functional causes. (31) The most frequent neurogenic cause is spinal dysraphism, a spectrum of congenital abnormalities that includes open and closed forms. Folic acid deficiency is the major etiology for these disorders and leads to the failure of complete closure of the spinal neural tube. (32) Other less common risk factors include chromosomal abnormalities and gene mutations. (33) The estimated incidence of spinal dysraphism is approximately 0.1%. (34) However, the incidence has been declining worldwide during the past few decades due to better nutrition and supplementation for pregnant mothers and improved prenatal care. (35) Other neurogenic etiologies include sacral agenesis, cerebral palsy, and traumatic spinal cord injuries.

Myelomeningocele is the most common and severe form of neural tube defects and accounts for 85% of all open spinal dysraphisms. (36) A myelomeningocele involves an opening in the spine where both nerve roots and spinal cord tissue protrude into a fluid-filled sac on the back. (34) Due to incomplete closure of the vertebral column and the risk of exposed spinal canal contents, urgent evaluation and treatment are needed. As many as 60% of all cases have some form of bladder dysfunction and nearly all cases of myelomeningocele are associated with a neurogenic bladder. (36)(37) The initial evaluation should include a neurologic examination. Checking muscle mass, tone, spontaneous movements, deep tendon reflexes, and skinfolds can help determine neurologic function and localize the dysraphism level. Of note, the level of dysraphism is poorly predictive of bladder dysfunction, and a separate complete bladder evaluation is recommended. (38)

Secondary VUR occurs when increased intravesical pressures lead to retrograde urine flow past the normal UVJ. This can be due to disorders such as a neurogenic bladder, PUVs, or bowel and bladder dysfunction in anatomically normal children. Correction of bowel and bladder dysfunction may resolve VUR. (31)(38) Early determination of bladder function allows for more proactive treatment and will guide future surveillance and management. (31)(38)(39)

No intervention is necessary if complete bladder emptying is noted, regardless of the degree of bladder denervation. Any sign of bladder dyssynergy, incontinence, incomplete emptying, or urinary tract abnormalities warrants further intervention. Treatment is guided by the initial assessment and may include anticholinergic medications and intermittent or continuous catheterization. If symptoms worsen despite initial treatment, then surgical intervention is indicated. (39)(40)(41)(42) Failure to intervene early can lead to high-grade VUR and recurrent UTIs, leading to irreversible renal injury. Patients with congenital neurogenic bladder will need regular surveillance visits and imaging to monitor the urinary tract and bladder function. (38)(41) Patients born with spinal dysraphism can have secondary spinal cord injury over time associated with growth and fibrosis. (43) Prognosis for these patients has dramatically improved in the past 3 decades due to advancements in detection, minimally invasive therapies, and higher levels of urologic care. (38)

ANOMALIES OF THE URETHRA

Posterior Urethral Valves

PUVs are characterized by abnormal mucosal folds at the prostatic urethra level that function as a valve to obstruct urine flow. (44) They are the most common cause of congenital urethral obstruction, affecting 1 in 5,000 to 1 in 25,000 live male births. (45)(46) Prenatal ultrasonography may show bilateral hydronephrosis, a thick-walled bladder, dilated posterior urethra, and/or oligohydramnios. (45) If not diagnosed prenatally, PUVs can present in newborns with intrauterine growth retardation and a weak voiding stream. Older children can present with recurrent UTIs, urosepsis, renal failure, or incontinence. VCUG remains the gold standard for detecting PUV and will show the proximal urethra more

dilated than the urethra distal to the valve. A urethral catheter should be placed in the bladder to relieve the obstruction. (46) The treatment of choice is transurethral valve ablation, with diversion of the obstructed bladder via vesicostomy as an alternative. (47) PUVs continue to have high mortality and morbidity (48) and are the most common cause of obstructive uropathy leading to ESRD. (49)(50)

Eagle-Barrett syndrome, also known as prune belly syndrome, is a rare lower urinary tract malformation affecting 1 in 30,000 newborns, with 95% of patients being males. (51) Prenatal ultrasonography reveals findings similar to PUVs. Diagnosis at birth is established clinically with pathognomonic findings of deficiency of abdominal wall musculature with loose and wrinkled overlying skin and bilateral cryptorchidism. RBUS reveals dilation of the urinary tract secondary to urethral obstruction. (52) Nearly 50% of patients have associated cardiovascular, gastrointestinal, or orthopedic malformations, and all patients have some degree of renal dysplasia. (53) These patients have a high incidence of chronic kidney disease, with 40% to 50% of patients needing renal replacement therapy. (54)

Urethral Stricture

Congenital urethral strictures are a rare urethral anomaly. These patients can be identified prenatally with abdominal imaging findings similar to PUV. (55) After birth, patients present with obstructive and irritative symptoms, such as

TABLE. **Genetic Syndromes Associated with CAKUT**

SYNDROME	RENAL ABNORMALITY	EXTRARENAL MANIFESTATIONS
Turner	Renal agenesis, hypoplasia	Short stature, amenorrhea, webbed neck, cubitus valgus, hypogonadism
Down	Renal agenesis	Intellectual disability, hypotonia, congenital heart disease, clinodactyly
Patau	Renal agenesis	Holoprosencephaly, midline anomalies, cleft lip/palate
CHARGE	Many urinary tract malformations	Cardiac defects, cleft palate, hearing loss, coloboma, choanal atresia, genital anomalies
Di George	Renal agenesis, dysplasia, VUR	Cardiac defects, cleft palate, thymic aplasia, hypocalcemia, immunodeficiency
Renal coloboma	Renal hypoplasia, MCDK, VUR	Optic disc/nerve anomalies, hearing loss, CNS and genital anomalies, skin and ligamentous laxity
Orofaciodigital	Renal cysts	Lobed tongue, cleft palate, hypodontia, micrognathia clinodactyly
Branchio-oto-renal	Renal agenesis, dysplasia	Lateral cervical fistulas or cysts, ear pits, hearing loss
Renal cysts and diabetes	Renal dysplasia, cysts	Diabetes mellitus
Fraser	Renal agenesis, dysplasia	Cryptophthalmos, syndactyly, tracheal stenosis/atresia, ambiguous genitalia, craniofacial anomalies
Alagille	MCDK, renal dysplasia, mesangiolipidosis	Chronic cholestasis, cirrhosis, cardiac anomalies, butterfly vertebrae, dysmorphic facies
Bardet-Biedl	Renal dysplasia, calyceal malformations	Obesity, intellectual disability, retinal dystrophy, polydactyly
Beckwith-Wiedemann	Nephromegaly, nephrocalcinosis, medullary sponge kidney	Macrosomia, hemihyperplasia, macroglossia, omphalocele
Simpson-Golabi-Behmel	Variety of kidney and urinary tract malformations	Craniofacial anomalies, organomegaly, tumors, intellectual deficiency
Smith-Lemli-Opitz	Renal hypoplasia	Syndactyly, microcephaly, intellectual disability, heart, lung, and gastrointestinal abnormalities
Rubinstein-Taybi	Variety of kidney malformations	Highly arched eyebrows, high-arched palate, micrognathia, internally deviated toes and thumbs, hirsutism, heart and eye abnormalities

Abbreviations: CAKUT=congenital anomalies of the kidney and urinary tract, CNS=central nervous system, MCDK=multicystic dysplastic kidney, VUR=vesicoureteral reflux.

urinary retention, dysuria, or weak stream. Diagnosis is made with VCUG, and treatment is with urethral dilatation, endoscopic repair, or vesicostomy if severe. Acquired urethral strictures are usually due to urethral instrumentation but can have inflammatory and traumatic etiologies as well. (56)

Hypospadias

Hypospadias is the most common malformation of the male genitalia, with an incidence of 0.3% to 0.5% in live male births. (57) It results in abnormal ventral placement of the urethral opening anywhere along the penis, scrotum, or perineum. Hypospadias is a clinical diagnosis and is usually made during the newborn examination. Hypospadias can present in older children and teens with a weak, downward-angled stream and, when associated with chordee, painful sexual intercourse. (58) The examination should focus on identifying other associated anomalies, the exact anatomical location of the meatus, assessment of the foreskin, and the presence of chordee. Newborn circumcision is contraindicated because the foreskin may be used for urethral and penile reconstruction. Surgical repair is elective and is ideally performed at 6 to 12 months of age. (59)

Epispadias

Epispadias is a congenital malformation of the urethra that can present as an isolated finding or can be associated with bladder exstrophy, a condition referred to as the exstrophy-epispadias complex. Epispadias is found in nearly all cases of bladder exstrophy and has an incidence of 1 in 30,000 live births, with a male preponderance. (60) Epispadias presents as a partial or complete opening of the urethral meatus on the dorsal surface. In males, the urethral meatus can be located anywhere from the distal penile shaft to the pubic area. (61) In females, epispadias is associated with a bifid clitoris and anteriorly displaced vaginal orifice. (62) Isolated epispadias is much less common, with an incidence of 0.5 in 100,000 live births in males and an even lower incidence in females. (61) Epispadias is usually evident on examination at birth and is strongly associated with urinary incontinence. RBUS should be performed to assess the urinary tract. (62) Surgical repair should be pursued as early as possible for both sexes. To maintain bladder function and continence, these patients may need multiple surgeries throughout their lifetime. (62)

GENETIC SYNDROMES ASSOCIATED WITH CAKUT

There is increasing evidence of genetic abnormalities as the cause of CAKUT. Identifying the underlying genetic involvement may improve identification and management of extrarenal manifestations. There are more than 500 genetic syndromes with renal or urinary tract abnormalities. (5)

Summary

- Based on strong research evidence, hydronephrosis is the most common cause of abnormal prenatal ultrasonography and abdominal masses in newborns. The urinary tract dilation classification system and the Society for Fetal Urology classification system are widely used for disease severity classification and management. (20)(22)
- Based on strong research evidence, vesicoureteral reflux is the most common pediatric urologic condition, affecting up to 30% of children with a febrile urinary tract infection and 0.4% to 1.8% of the general pediatric population. (24) Management remains a controversial topic and is the subject of ongoing research.
- Based on consensus, ultrasonography is the imaging of choice for initial evaluation prenatally and after birth to evaluate congenital anomalies of the kidney and urinary tract and is the image of choice for the initial evaluation of various diagnoses, to classify severity of disease, and to guide future monitoring.
- Based on strong research evidence, posterior urethral valves are most commonly diagnosed prenatally with ultrasonography findings of bilateral hydronephrosis, dilated posterior urethra, and/or oligohydramnios. (45) Based on some research evidence as well as consensus, the treatment of choice is transurethral valve ablation. (47)
- Based on strong research evidence, hypospadias is the most common malformation of the male genitalia. Newborn circumcision is contraindicated in these patients, and surgical repair is ideally performed at 6 to 12 months of age. (60)

SUGGESTED QUALITY IMPROVEMENT PROJECT

- Improve the tracking of the multidisciplinary follow-up of the patient with urinary abnormalities.

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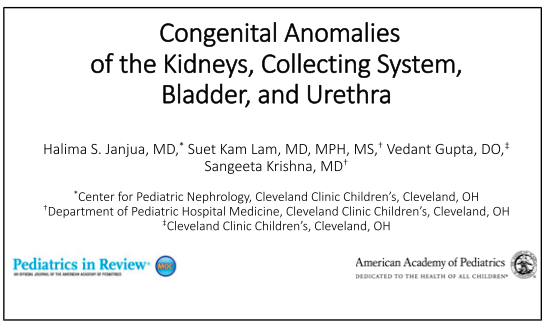

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1. You are seeing a 2-week-old female newborn for her first physical examination after discharge from the nursery. She was diagnosed prenatally with a solitary left kidney by prenatal ultrasonography. You decide to order repeated renal ultrasonography today. In explaining your reasoning to the parents, which of the following statements best describes your rationale for ordering the ultrasonography today?
   A. Only 10% of affected children have a problem in the solitary kidney, and this is just a precaution to make everyone feel better.
   B. Renal agenesis is associated with liver cysts 30% of the time, so you need the ultrasonography to look at the liver also.
   C. The solitary kidney is often affected by nephrocalcinosis and ultrasonography is needed to assess the extent of calcium deposits.
   D. The solitary kidney is typically smaller than normal because of a lack of growth factors from the other kidney, and follow-up ultrasonography is needed to check its growth rate.
   E. Up to 50% of children with 1 kidney can have a problem in the contralateral urogenital tract.

2. You are called to the newborn nursery because the nursing staff noted abdominal fullness on the right side of the abdomen while bathing a full-term male newborn infant. The baby is urinating normally, and there is no gross hematuria. On physical examination the baby is afebrile. His pulse is 120 beats/min, blood pressure is 100/60 mm Hg, and respiratory rate is 50 breaths/min. On physical examination you are able to palpate a large mass on the right side of the abdomen, and it does not cross the midline. The lungs are clear, but breath sounds are decreased on the right side. Which of the following is the most likely diagnosis in this patient?
   A. Hydronephrosis.
   B. Multicystic dysplastic kidney.
   C. Neuroblastoma.
   D. Renal cell carcinoma.
   E. Wilms tumor.

3. You are examining a 48-hour-old full-term male newborn in the nursery. The mother is only 16 years of age and did not have prenatal care because she was trying to hide the pregnancy from her parents and took no medications or supplements and had no prenatal ultrasonography. She occasionally smoked cigarettes and marijuana but stopped late in her first trimester when she learned she was pregnant. She used condoms inconsistently. Her medical history is significant for sexually transmitted infections, including gonorrhea and chlamydia, 6 months before her pregnancy. The baby is making approximately 6 wet diapers a day without any hematuria. On physical examination you note the presence of a deep sacral pit and you are unable to visualize its bottom. The muscle tone in the lower extremities is noticeably decreased. An evaluation is started. Which of the following factors most likely placed this infant at the greatest risk for possibly having this condition?
   A. Being first born.
   B. History of marijuana and tobacco use.
   C. History of sexually transmitted infections.
   D. Lack of intake of prenatal vitamins.
   E. Maternal age.

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4. A 15-month-old boy is brought to the clinic for follow-up. He is new to the practice because the family just moved to the area. The baby was not previously seen by a primary care clinician and received no immunizations so far due to the family being uninsured. The father just started a new job in the area that provided him with insurance. Physical examination is significant for macrosomia, hemihyperplasia, and macroglossia. Further testing of the kidneys and urinary tract system is warranted because these clinical findings may be associated with which of the following conditions?

   A. Epispadias.
   B. Medullary sponge kidney.
   C. Neurogenic bladder.
   D. Renal agenesis.
   E. Renal cysts.

5. A 5-year-old boy is brought to the emergency department with a temperature of 102.2°F (39°C) and emesis for the past 24 hours. He was born at full-term and has never been diagnosed as having any medical problems. He is up-to-date on his vaccines. Physical examination reveals an ill-looking child with a pulse of 100 beats/min, blood pressure of 70/50 mm Hg, and a respiratory rate of 30 breaths/min. He has diffuse abdominal tenderness. His white blood cell count is 18,000/μL (18×10^9/L). Urinalysis shows nitrates and leukocytes. Blood and urine cultures are pending. A fluid bolus is given, and he is started on intravenous antibiotics and admitted to the hospital for hydration. Which of the following imaging studies is the most appropriate next step in assessing the cause of his symptoms?

   A. Abdominal radiography.
   B. Computed tomography of the abdomen.
   C. Intravenous pyelography.
   D. Renal ultrasonography.
   E. Voiding cystourethrography.