

VOLUME 42

AUA

UPDATE SERIES

2023

LESSON 18

Surgical Considerations in Horseshoe and Anomalous Kidneys

Learning Objective: At the conclusion of this continuing medical education activity, the participant will be able to review the anatomical abnormalities associated with anomalous kidneys and describe the necessary adjustments and potential pitfalls when operating on these kidneys.

This AUA Update aligns with the American Board of Urology Module on Calculus, Laparoscopy-Robotics and Upper Tract Obstruction. Additional information on this topic can be found in the AUA Core Curriculum sections on Laparoscopy and Robotic Surgery, and Urolithiasis.



Wilson Sui, MD,¹ Marshall L. Stoller, MD,¹ and David B. Bayne, MD, MPH¹

¹Department of Urology, University of California San Francisco, San Francisco, California

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KEY WORDS: horseshoe kidney; ectopic kidney; cross-fused renal ectopia, anomalous

INTRODUCTION

Anomalous kidneys are routinely encountered in clinical practice, and due to their distorted anatomy, preoperative diagnostic imaging and innovative surgical approaches are required. These kidneys broadly can be categorized into failure of renal ascent, such as renal ectopia, occurring in an estimated 1 in 500-1,200 births,¹ and form/fusion abnormalities, such as cross-fused renal ectopia and horseshoe kidney, which occur in 1 in 400-600 births.² Though of course not congenital, renal transplants also should be considered in this category as their management often necessitates unique approaches in surgical management.

With anomalous kidneys, even in routine procedures such as ureteral stent placement and ureteroscopy, urologists' technical skills are tested. A well-thought-out plan is crucial to surgical success. These cases are an excellent opportunity for creative approaches and inventive use of surgical equipment. The goals for this Update are to review the anatomical considerations, necessary adjustments, and potential pitfalls when operating on the horseshoe kidney and other anomalous kidneys.

ANATOMICAL CONSIDERATIONS

Ectopic kidneys may be in a variety of locations including the pelvis, abdomen, contralateral, or crossed to an orthotopic position, or rarely in the thoracic cavity. There is a slight increase in occurrence on the left and there are no gender differences.³ While the exact mechanism(s) of this ascent abnormality is unknown, it leads to a renal pelvis that is typically located anterior to the renal parenchyma (instead of medial) due to incomplete rotation. Hydronephrosis is common, being reported in up to 50% of cases at either the ureteropelvic junction or ureterovesical junction.⁴ Rates of vesicoureteral reflux are 30%-50%.⁵⁻⁷

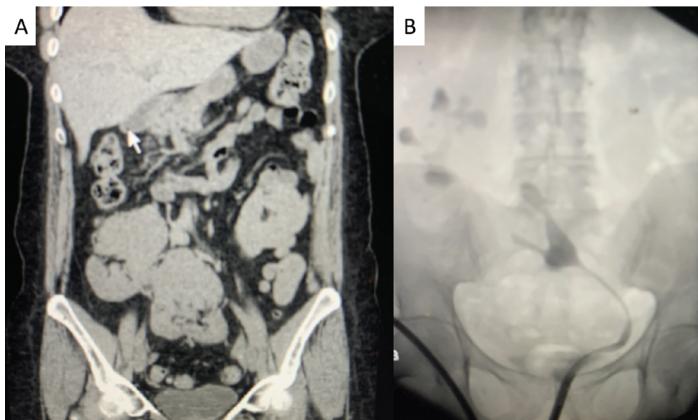


Figure 1. Left-to-right crossed-fused ectopia (A) with retrograde pyelogram showing the left ureter inserting orthotopically (B).

Despite the variety of locations where ectopic kidneys may reside, the ipsilateral ureter typically remains inserted into an orthotopic ureteral orifice unless there is an associated ectopic ureter as well (Figure 1). The vascular blood supply is typically anomalous with vascular takeoffs depending on the ultimate location of the kidney. While there is an association with agenesis, ectopia, or vesicoureteral reflux in the contralateral kidney, the majority of contralateral kidneys are normal.⁸ Probably the most important associations are with reproductive organ anomalies, reported in 15%-45% of these patients, notably uterine (uni- or bicornuate, atresia) or vaginal (underdevelopment, duplication) in females, and hypospadias or undescended testicles in males.^{1,5,9,10}

Crossed renal ectopia is a condition where the kidney is located on the opposite side of where its ureter inserts into the bladder. This is the second most common renal anomaly and 90% of these are fused to the other kidney.¹¹ While the exact embryological cause of this malformation is unknown, the superior pole of the crossed unit is typically fused with the inferior pole of the orthotopic unit, likely due to the crossover time causing it to lag behind its partner (Figure 1). Ascent ceases after the orthotopic kidney reaches its normal location or there is an anatomical barrier. Thus, there are many potential configurations to these fused units depending upon where they are fused, the degree of rotation, and their ultimate location. Abnormal vasculature (numbers of vessels and their origins) is common (Figure 2). Similar to simple ectopia, these abnormalities also are associated with other anomalies such as imperforate anus (20%), cryptorchidism, hypospadias, or vaginal/uterine irregularities.^{12,13}

Horseshoe kidneys are more common in males (2:1), occur in about 0.25% of the population, and are the most common kidney anomaly.² Aberrant fusion of the metanephric blastema leads to fusion of the inferior poles of the kidneys resulting in the incomplete ascent of these kidneys. The isthmus that forms between the 2 moieties is limited by the inferior mesenteric artery at the level of L3-L4. While most of these kidneys are fused at the lower pole forming this isthmus, the moieties may not be symmetrical, with 1 series reporting 38% left lateral and 22% right lateral fusions.¹¹

These kidneys are malrotated with anteriorly rotated renal pelvises and high ureteral insertion onto the ureteropelvic junction frequently leading to hydronephrosis. The blood supply includes not only the main renal hilum, but often anomalous vessels supplying the lower poles and isthmus (Figure 3). These vessels have been found to be derived from the aorta, inferior mesenteric, common iliac, external iliac, or sacral arteries.¹⁴ In utero, these embryonic vessels supply the kidney during ascent; however, the failure to completely ascend leads to persistence of these vessels. There are many associated conditions, with 1 institution reporting 50% of their patients as having extrarenal diseases, with gastrointestinal and vertebral sources being the most common.¹⁵ The most commonly associated syndromes include Turner syndrome

ABBREVIATIONS: end-stage renal disease (ESRD), percutaneous nephrolithotomy (PCNL), extracorporeal shock wave lithotripsy (SWL), ureteropelvic junction obstruction (UPJO)

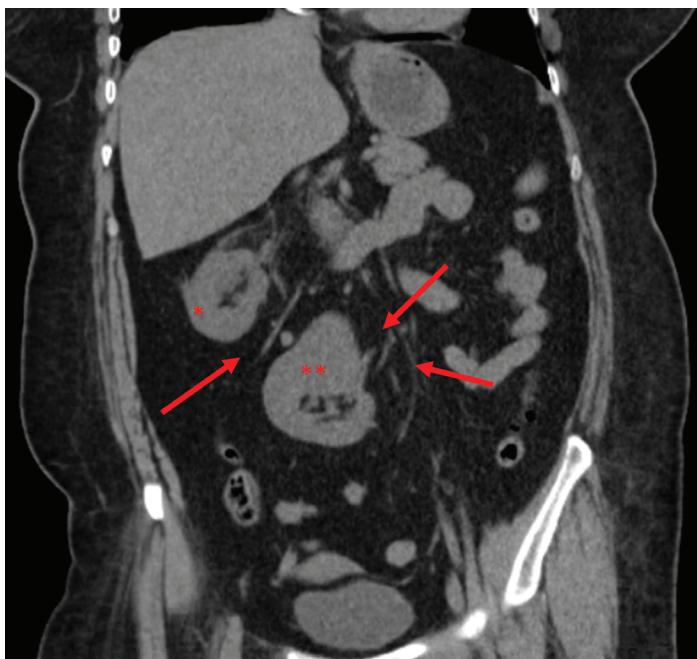


Figure 2. Orthotopic right kidney (*) in addition to left-to-right crossed ectopia (**) demonstrating the aberrant vasculature (denoted by arrows) due to the abnormal course of the crossed kidney during ascent.



Figure 3. CT of a horseshoe kidney with emphasis on marked vascular variability. Arrows denote supernumerary renal arteries branching off the common iliac arteries.

(horseshoe kidney in 60%), caudal regression syndrome, and VATER (for vertebral anomalies, anorectal malformations, tracheoesophageal fistula, esophageal atresia, renal anomalies) syndrome. Associated genitourinary anomalies are similar to those discussed for ectopic kidneys above.

The incidence of end-stage renal disease (ESRD) has been rising, and in 2016 there were over 120,000 newly reported cases of ESRD, with over 97% of these patients starting renal



Figure 4. Kidney transplant with severe tortuosity of the ureter. Retrograde access was quite challenging, and as such, a nephrostomy tube was placed for drainage.

replacement therapy and only 2.8% receiving a preemptive kidney transplant. Overall, 29.6% of all prevalent ESRD patients had a functioning kidney transplant.¹⁶ Urological complications after renal transplant occur in about 10% of cases. The estimated incidence of nephrolithiasis in a kidney transplant is 1%.^{17,18} The location of transplanted kidneys is typically in the right or left lower quadrants, with the vascular supply anastomosed to the external iliac vessels while the ureter is connected to the dome of the bladder. Accessing these kidneys retrograde requires cannulating the neo-ureteric orifice, which can be quite challenging due to its location on or near the dome of the bladder, and additionally, the ureter may be tortuous and take an unfamiliar course (Figure 4). Pediatric en bloc kidney transplants are utilized to increase the potential donor pool with good long-term outcomes.¹⁹ These are performed by anastomosing the donor aorta and inferior vena cava end-to-side to the recipient external iliac vessels and performing 2 ureteroneocystostomies or 1 anastomosis in a Wallace configuration. Depending on the orientation of the 2 kidneys, percutaneous access may be difficult or impossible due to the surrounding organs and vasculature (Figure 5).

RADIOLOGICAL CONSIDERATIONS

With the increasing utilization of noninvasive imaging, the incidental diagnosis of these anomalous renal units is becoming more common. Failure of imaging to demonstrate a kidney in its anatomical location should prompt the urologist to rule out ectopia with further imaging. Small or atretic ectopic kidneys may not be readily visualized on CT or ultrasonography. In these cases, magnetic resonance imaging may be the

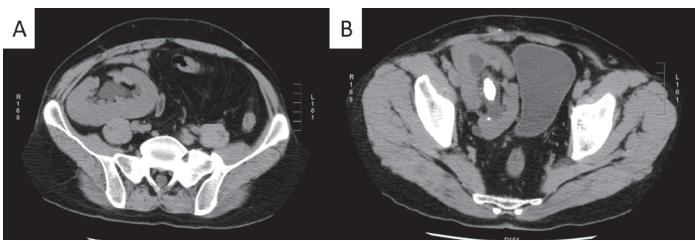


Figure 5. Two images of the same patient with a right pediatric en bloc renal transplant. A, The lateral kidney was malrotated, making access challenging. B, The medial kidney with a 2-cm renal pelvis stone that was amenable to right lower quadrant percutaneous access via ultrasound guidance.

only modality able to provide the anatomical detail in order to identify these structures. For horseshoe kidneys, the isthmus, though it may sometimes be thin, may be seen as a solid band of tissue even on ultrasound. **Functional imaging may show an uninterrupted strip of tissue across midline if the isthmus is large and bulky enough and has functioning parenchyma.** On cross-sectional imaging, the characteristic low lie and inward axis of the inferior poles of the renal moieties are typically demonstrated.

UROLOGICAL CONSIDERATIONS

As most horseshoe and ectopic kidneys are now found incidentally, one must be mindful that at least half of these patients will be asymptomatic.^{20,21} Patients with these findings should be reassured that, if found, they may not experience any deleterious consequences. **When symptomatic, the most encountered urological issues are predominantly related to poor drainage—hydronephrosis, calculi, and infection.** Malrotation leads to an anteriorly located renal pelvis, with the ureter inserting into the renal pelvis at a high location, leading to a so-called high ureteropelvic insertion. The ureter itself may also have an abnormal course causing obstruction.

While hydronephrosis is common, it is most often nonobstructive, and if so, can be observed. In 13%-34% of patients, however, the hydronephrosis will be secondary to ureteropelvic junction obstruction (UPJO), which may be due to a combination of high-inserting ureter, abnormal course anterior to the isthmus or renal parenchyma, or compression by an aberrant blood supply.^{22,23} Urinary tract infections occur in a third of these patients, likely again due to suboptimal drainage. Stone formation is also common with an estimated incidence of 36%.²⁴ **These stones are typically calcium-based calculi**

and the metabolic abnormalities of horseshoe kidney stone formers are similar to their non-horseshoe kidney counterparts, suggesting that it is the anatomical issue that drives stone formation in these patients.²⁵ Symptoms, however, may be vague and present atypically depending on the location of the kidney. These considerations will be discussed in more detail in the context of commonly encountered urological procedures.

ANATOMICAL CONSIDERATIONS, ADJUSTMENTS, AND POTENTIAL PITFALLS FOR SELECT UROLOGICAL PROCEDURES

Ureteroscopy. Noninvasive management of nephrolithiasis in horseshoe kidneys and other anomalous kidneys may be challenging, though feasible. As reviewed previously, these kidneys have higher rates of stone formation secondary to poor drainage. **As a result, one must be mindful of residual stone fragments that may not pass spontaneously after surgery and require further procedures.** One study reported that over 40% of patients with horseshoe kidneys undergoing ureteroscopy required a second procedure.^{26,27} Neither the surgical guidelines from the AUA²⁸ nor those of the European Urology Association²⁹ specifically address best practices for management of nephrolithiasis in anomalous kidneys. At our institution, we favor ureteroscopy or percutaneous approaches over extracorporeal shock wave lithotripsy (SWL) in most situations based on lower rates of stone clearance due to an anteriorly placed renal pelvis. A review of ureteroscopy for horseshoe kidneys showed that the initial stone-free rate was 55.6%-100% for flexible ureteroscopy vs just 22.7%-60% in SWL.^{30,31} For SWL in a horseshoe kidney, we highly encourage patients to lie prone after treatment to facilitate stone passage. For ectopic kidneys, a different group found that the stone-free rate for ureteroscopy was only 20% for renal stones in ectopic kidneys vs 94% for ureteral stones, highlighting the difficulty in accessing some of these ectopic kidneys endoscopically.³²

While the absolute number of calyces is typically similar in horseshoe or ectopic kidneys compared to normally positioned renal units, the unfamiliar lie may make them disorienting to inspect. **The acute angulation of the calyces, especially in the lower pole, makes stones in this area very difficult to access and has been shown to be a risk factor for clearance failure.**^{33,34} Some of these stones may be best approached percutaneously. The ureters may take an unexpected course from the kidney to the bladder, leading to tortuosity, making retrograde endoscopic access difficult. To alleviate this, ureteral access sheaths or stiffer wires may be useful in straightening tortuous ureters. Care should be taken when placing these, however, as there is a higher risk of perforation given the unusual ureteral course. In terms of ureteral stents, varying lengths should be on hand and fluoroscopy targeted more caudal than typical for horseshoe and low-lying ectopic kidneys. On the other hand, longer stents may be needed in cross-fused ectopia.

The literature and our own experience would suggest that there is not a higher complication rate in treating these anomalous kidneys. The complication rates for horseshoe and ectopic kidneys were 7%-24% and 7.4%, respectively.^{30,32} The most commonly reported postoperative complications were renal colic

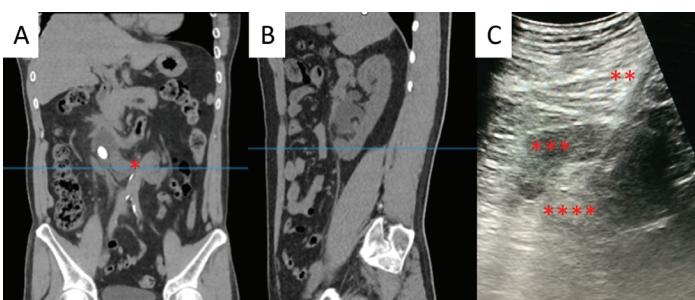


Figure 6. Superior pole access for horseshoe kidney with an anteriorly placed renal pelvis and stone. Isthmus (*) is visible in midline. Horseshoe kidney in coronal (A) and sagittal (B) reconstructions. C, Ultrasound-guided access into upper pole. The access sheath (**) can be seen traversing the renal parenchyma (***) into the collecting system (****).

and infection-related issues. These cases are technically challenging, leading to longer operative times with the mean range of 60-90 minutes.^{27,32} As a result, it is useful to set an operating time limit to reduce the risks of prolonged anesthesia and to minimize the risk of infectious complications, especially if a second procedure is proactively planned.

Percutaneous nephrolithotomy. Access to anomalous kidneys requires careful planning and an understanding of the associated anatomical irregularities. The incomplete ascent of horseshoe kidneys routinely requires an upper or mid kidney puncture as they are typically located in the lower abdomen, making the lower pole inaccessible (due to their medial orientation), and the upper pole typically has the shortest skin-to-stone distance (Figure 6). In addition, a more posterior puncture is needed due to the malrotation compared to typically positioned kidneys. In patients with normal anatomy, upper pole punctures may require a supracostal approach, leading to pneumothorax or hydrothorax. The relatively inferior displacement of the upper poles in horseshoe kidney leads itself to decreased risk of these particular complications. An upper pole puncture typically allows access to the associated lower pole calyces, albeit they may require longer flexible scopes. The resultant inward tilt of the lower pole makes them almost impossible to access directly, and aberrant vasculature posterior to the isthmus (aorta and inferior vena cava) complicates access. When access is obtained through these locations, the tract itself may be long, leading to instrument issues and other calyces being relatively inaccessible due to the limited mobility. Variations in the surrounding viscera not typically encountered with normal kidneys include a retrorenal location to the colon, which can make posterior access challenging (Figure 7).

Access is less predictable due to the variety of locations kidneys may reside, from simple to cross-fused ectopia. In general, one should be vigilant about the hazards of surrounding viscera and vasculature and mindful of limitations posed by bony structures. While historically access has been fluoroscopically guided, in our practice, the utilization of intraoperative ultrasonography helps to alleviate some of these anatomical issues as adjustments can be made based on real-time imaging. In rare instances, severely malrotated kidneys may preclude any access through the renal parenchyma, therefore necessitating pyelolithotomy via open or laparoscopic approaches. After access, depending on the location of the anomalous kidney, sometimes a long sheath and nephroscope are required if the kidney is located deep in the pelvis, for example. The longer distance may lead to higher rates of access lost, bleeding, and operative time. Due to these anatomical anomalies, high-quality cross-sectional imaging is critical for preoperative planning. At our institution, patient positioning for horseshoe kidney access is typically prone to ensure access to the upper poles that may require a more medial puncture that is difficult or impossible to achieve while patients are in a supine position. This is because the trajectory of the needle is usually more posterior-anterior and the target is typically near the paraspinous muscle, which is impossible to access in the supine position. Other positions—modified dorsolithotomy, supine, and flank—are options as well, depending on patient habitus, stone location, and provider comfort. Care must be



Figure 7. Horseshoe kidney with left moiety hydronephrosis with adjacent colon presenting a potential hazard to posterior access.

taken to account for the increased mobility of the horseshoe kidney due to posterior to anterior migration in contrast to a lateral-to-medial migration hindered by the great vessels in anatomically normal kidneys. Due to this, balloon dilation is preferred over sequential dilators.

Overall, the outcomes for percutaneous nephrolithotomy (PCNL) of horseshoe kidneys are favorable, with stone-free rates in contemporary studies ranging from 81%-88%.³⁵⁻³⁷ In many cases, PCNL may be the preferred approach over ureteroscopy due to the anatomical difficulties described in the prior section. A propensity score-matched analysis of pooled patient data from 20 centers comparing flexible ureteroscopy vs PCNL for anomalous kidneys found that stone-free rates favored PCNL in horseshoe and ectopic kidneys.³⁸ In our experience, which is supported by the literature, though these kidneys are technically more challenging to access and treat, there does not seem to be a higher complication rate.³⁹⁻⁴¹

Pyeloplasty. UPJO is common in horseshoe kidneys, with an incidence of 15%-33%.⁴² The etiology is typically related to the high insertion of the ureter onto the renal pelvis, unusual course of the ureter especially over the isthmus or renal parenchyma, and anomalous vasculature, the latter of which may act as obstructing crossing vessels. While endopyelotomy is an option for these patients, we favor pyeloplasty as the frequency of anomalous crossing vessels may leave these patients at an increased risk of acute bleeding or delayed failure.⁴³ Outcomes for pyeloplasties of these anomalous kidneys have been reported to have a 55%-100% success rate, though these are primarily composed of single-institution reports.⁴⁴⁻⁴⁷

The surgical approach—open, laparoscopic, robot assisted—to these operations is up to provider preference, equipment availability, and comfort. While the main renal vessels may enter the renal moiety cranially, as mentioned previously, it is critical to identify irregular vasculature as these vessels may act as the fulcrum for obstruction. Care should be taken not to injure these vessels because they are end arteries. The anterior location of the renal pelvis may be advantageous when considering pyeloplasty or pyelolithotomy as this may be more accessible. Some cases of anomalous kidneys necessitate surgical creativity, especially with reconstruction. We present a case of a completely anteriorly rotated ectopic pelvic kidney

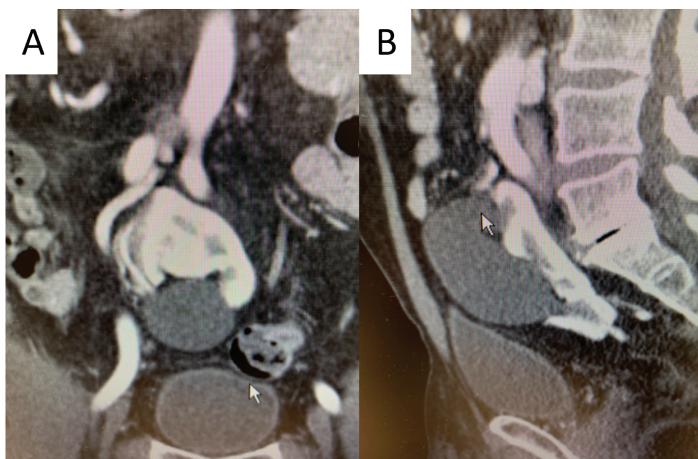


Figure 8. Malrotated ectopic right kidney with aberrant vascular anatomy and ureteropelvic junction obstruction in coronal (A) and sagittal (B) reconstructions. Close proximity of urinary bladder was amenable for a laparoscopic pyelovesicostomy.

with a UPJO (Figure 8). Due to the proximity of the renal pelvis to the bladder, we performed a pyelovesicostomy, with an excellent postoperative outcome.

Partial nephrectomy and heminephrectomy. The incidence of renal cell carcinoma is no higher in horseshoe or other anomalous kidneys than in the general population.^{48,49} The general principles and steps to the operation are no different than a normal kidney; however, the complicating issues are the unfamiliar surrounding organs, anomalous vasculature, and, in horseshoe kidneys, management of the isthmus. When considering access and placement of ports for minimally invasive surgery for anomalous kidneys, one must consider not only the location of the kidney, but also the surrounding organs/vessels, which may now complicate the dissection. In contrast to a normally positioned kidney, the isthmus typically precludes the inferior-to-superior dissection most familiar to urologists. **Therefore, in horseshoe kidneys, dissection may**

best be approached from the superior pole, identifying the main renal hilum and then dissecting down toward the renal isthmus. Once the main vasculature of the kidney is ligated, demarcation based on turgor and color can be undertaken. In our experience, for horseshoe kidneys, ports must often be placed more caudal and medial; however, the primary vasculature typically originates in the normal location of the aorta and vena cava. Other modifications will vary depending on location when operating on an ectopic kidney.

Preoperative imaging is crucial. A conventional CT scan with and without contrast is typically sufficient; however, the use of a CT angiogram to identify vasculature may sometimes be helpful (Figures 2 and 3). **Attention should be paid to the location of the isthmus as sometimes it is not located directly in the midline (Figure 9).** A CT urogram is useful in situations where the collecting system of one moiety is close to or crosses midline in order to avoid accidental entry. While the pelvis is likely a location familiar to most urologists, one must be cognizant of the surrounding organs and proceed cautiously with the dissection and exposure. **When clamping the arteries for a partial nephrectomy, especially for a mass located near the isthmus, one must consider that the main arterial blood flow to that portion of the renal moiety is derived from this variable vasculature.** If a heminephrectomy is being performed, there are many ways to divide the isthmus depending on its thickness. In our experience, an energy device or stapler usually suffices; however, for a thick isthmus, division followed by oversewing (same technique as with partial nephrectomies) may be required.

While most patients with horseshoe and anomalous kidneys are asymptomatic, these patients are routinely encountered in clinical practice. Any provider planning to perform a procedure on these renal units should be aware of the irregular vasculature and unfamiliar neighboring structures that may complicate the operation. Preoperative surgical planning is therefore critical, and high-quality imaging should be obtained. Ultimately, anomalous kidneys challenge the surgeon to be resourceful and employ innovative surgical thinking.

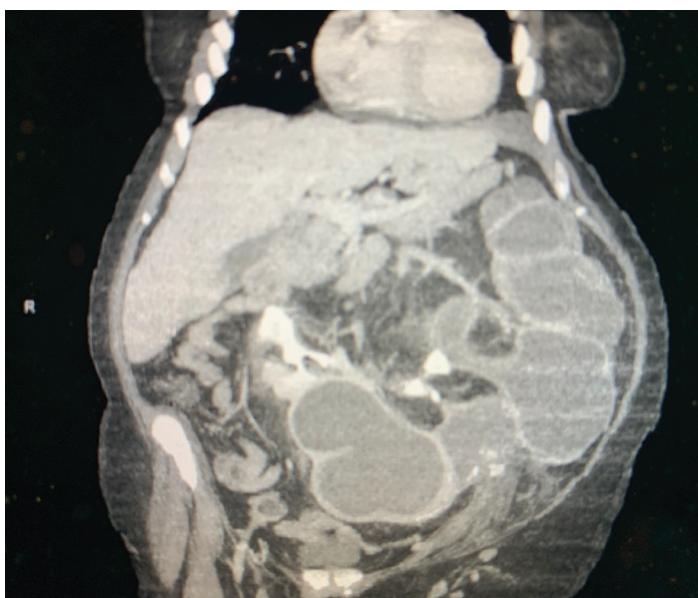


Figure 9. Horseshoe kidney with severe pyonephrosis of left collecting system secondary to obstructing renal pelvis stone. Note the isthmus was located to the right of midline due to the massive pyonephrosis. Left heminephrectomy was performed via laparoscopic approach after drainage with multiple nephrostomy tubes.

DID YOU KNOW?

- Most patients with horseshoe and anomalous kidneys are asymptomatic, and when symptoms occur they are usually related to poor drainage (urinary tract infection, nephrolithiasis, UPJO).
- Horseshoe kidneys are characterized by failure of ascent leading to a lower abdominal location, and malrotation leading to anteriorly rotated renal pelves.
- Operating on horseshoe and anomalous kidneys is complicated by unfamiliar surrounding organs and irregular vasculature.
- High-quality preoperative imaging is critical in operative planning, and the use of real-time ultrasound intraoperatively may be useful, especially when obtaining percutaneous access. Creative surgical thinking is key, especially when approaching ectopic or cross-fused kidneys.

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Study Questions Volume 42 Lesson 18

1. A patient with a 3-cm renal mass located near the isthmus of a horseshoe kidney is undergoing partial nephrectomy. With the main renal hilum clamped, there is substantial bleeding during the mass excision. An unclamped anomalous artery feeding the mass may be derived from the
 - a. Marginal artery of Drummond
 - b. Inferior mesenteric artery
 - c. Superior mesenteric artery
 - d. Celiac artery
2. In cross-fused renal ectopia, the ____ of the crossed unit is typically fused to the ____ of the orthotopic unit.
 - a. Superior pole, inferior pole
 - b. Inferior pole, superior pole
 - c. Lateral lobe, inferior pole
 - d. Superior pole, lateral lobe
3. The complication more likely to occur when obtaining access to a normal kidney as opposed to a horseshoe kidney is
 - a. Retrorenal colon injury with posterior calyx access
 - b. Pneumothorax with upper pole puncture
 - c. Vascular injury with upper pole puncture
 - d. Renal hilum injury with mid pole puncture
4. A 40-year-old female with a history of left lower quadrant abdominal pain is found to have a right-to-left cross-fused ectopic kidney with hydronephrosis. When performing a retrograde pyelogram, the most likely location of the right-to-left moiety ureteral orifice is
 - a. Orthotopic on the right
 - b. Ectopic on the right
 - c. Ectopic on the left
 - d. On the dome
5. A 50-year-old male with a horseshoe kidney performs a 24-hour urine as a part of a metabolic stone workup. As compared to a stone former with a normal kidney location, the most likely abnormal urinary analyte is
 - a. Hypocitraturia
 - b. Hyperoxaluria
 - c. Hypercalciuria
 - d. No difference