Percutaneous Nephrolithotomy in the Horseshoe Kidney

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Abstract The horseshoe kidney (HSK) presents specific challenges when planning percutaneous intervention. The significant anatomic variation of the HSK requires the urologist to have a unique understanding of the altered anatomy in order to provide treatment that is both safe and effective. Variation in blood supply, malrotation of the HSK unit, relation to adjacent structures, and ectopic location must all be considered when undertaking percutaneous nephrolithotomy (PCNL) in the HSK. Because of the alteration in the percutaneous access tract required, flexible endoscopy should be performed at the conclusion of all PCNLs in the HSK. The expected complications and post-operative course for PCNL in the HSK is similar to that of the anatomically normal kidney. This chapter provides guidance to the urologist faced with the challenge of performing PCNL in the HSK. The primary purpose of this chapter is to describe the unique considerations required when performing PCNL in the HSK.

Keywords Percutaneous nephrolithotomy · Horseshoe kidney · Endoscopic technique · Nephrolithiasis · PCNL

1 Introduction

Horseshoe kidneys (HSK) are defined as bilateral functional renal moieties on both sides of the vertebral column which are fused together by an isthmus. The lower poles of the kidneys are connected by this isthmus, which may be positioned midline or slightly laterally. Lateral positioning will result in an asymmetric HSK. The isthmus is comprised of renal parenchyma in roughly 80% of cases, with the rest being comprised of fibrous connective tissue. The ureters remain uncrossed from the renal hilum to the urinary bladder and follow an anterior course up and over the isthmus, occasionally resulting in obstruction (Cook and Stephens [1977\)](#page-13-0). Fusion occurs at the lower pole in 90% of cases (Khougali et al. [2021\)](#page-13-1). The incidence of HSK is

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[©] The Author(s), under exclusive license to Springer Nature Switzerland AG 2023 J. D. Denstedt and E. N. Liatsikos (eds.), *Percutaneous Renal Surgery*, https://doi.org/10.1007/978-3-031-40542-6_19

approximately 1:500 in the general population (0.25%) and is twice as common in males (Schiappacasse et al. [2015\)](#page-13-2).

HSK can be associated with cardiovascular, gastrointestinal, skeletal, and genitourinary (GU) abnormalities. This chapter will briefly discuss GU abnormalities, as they are most common. These abnormalities include: vesicoureteral reflux (50%), ureteropelvic junction obstruction (35%), ureteral duplication (10%), cryptorchidism and hypospadias in 4% of male patients, and vaginal septum and bicornuate uterus in 7% of female patients (Schiappacasse et al. [2015\)](#page-13-2). HSK has also been associated with Patau and Gardner syndromes (trisomy 13 and deletion q15q22), up to 20% of Down and Edwards (trisomies 21 and 18) syndromes, and 60% of Turner syndrome patients (Natsis et al. [2014\)](#page-13-3).

The primary purpose of this chapter is to describe the unique considerations required when performing percutaneous nephrolithotomy in the horseshoe kidney.

2 Embryology

The normal embryological development of the kidney and ureter are well described. Three distinct structures are responsible for proper development: the pronephros, mesonephros, and metanephros. The pronephros and mesonephros degenerate in utero, and the metanephros ultimately develops into the mature kidney (Tanagho et al. [2013\)](#page-14-0). During this process, the developing kidneys ascend from the pelvis to the upper lumbar region. As they ascend, the kidneys rotate medially approximately 90 degrees. This results in hila that are directed anteromedially (Muttarak and Sriburi [2012\)](#page-13-4). Abnormal fusion of the developing kidneys causes an early arrest of the ascension process as cranial migration is prevented by the inferior mesenteric artery (IMA) (Baskin and Cunha [2021\)](#page-13-5). This arrest is key to understanding the anatomic position and relationships of HSKs. Figure [1](#page-2-0) illustrates these important anatomic differences.

Two theories have been described for HSK development. Classically, mechanical fusion of the metanephric blastema has been attributed to abnormal flexion or growth of the developing spine and pelvic organs. This is thought possible because the immature kidneys have no renal capsule, and this contact fusion results in a fibrous isthmus. Alternatively, it has been proposed that a teratogenic event occurs that results in abnormal migration of posterior nephrogenic cells that then fuse, creating an isthmus. It has been postulated that this process results in a parenchymatous isthmus (Schiappacasse et al. [2015;](#page-13-2) Natsis et al. [2014\)](#page-13-3).

3 Anatomic Considerations

Abnormal fusion of bilateral kidneys across the midline creates significant anatomic variation from normal. Most commonly, the isthmus lies anterior to the great vessels at the level of the third to fifth lumbar vertebra, just inferior to the IMA (Muttarak and Sriburi [2012\)](#page-13-4). The abnormal fusion also prevents normal renal rotation. This leaves the inferior poles oriented medially, with the renal pelvis located more anteriorly and/or laterally than normal (Schiappacasse et al. [2015;](#page-13-2) Muttarak and Sriburi [2012\)](#page-13-4).

The calyces are located in the upper two-thirds of each kidney and often point medially towards the spinal column, downwards, or both. The isthmus may be drained by an extrarenal calyx or an independent ureter. The ureteropelvic junction (UPJ) is positioned more superior than normal at the inner rim of the superior part of the kidney (Natsis et al. [2014\)](#page-13-3). The ureters cross anterior to the isthmus and then course inferiorly towards the bladder (Schiappacasse et al. [2015;](#page-13-2) Muttarak and Sriburi [2012\)](#page-13-4). An example of these anatomic abnormalities is provided by the contrasted CT scan in Fig. [2.](#page-3-0)

The blood supply of HSK is widely variable (Schiappacasse et al. [2015;](#page-13-2) Natsis et al. [2014;](#page-13-3) Muttarak and Sriburi [2012\)](#page-13-4). The isthmus is commonly supplied by a single vessel derived from the abdominal aorta (AA) (Natsis et al. [2014\)](#page-13-3). The renal arteries can originate from the AA, common iliac arteries (CIA), and/or the IMA (Muttarak and Sriburi [2012\)](#page-13-4).

Graves' classification was developed in attempt to characterize the most common variations in blood supply. This system classifies HSK arterial anatomy into one of

Fig. 2 CT abdomen/pelvis with contrast demonstrating common anatomic variation seen in HSKs. Note the position of the UPJs, malrotation of both renal units, and inferior displacement [original image]

six most observed variations. Each artery supplies its own area, with no collateral circulation between segments. Type 1a exhibits normal renal arterial pattern, with the upper, middle, and lower segments supplied by a single renal artery arising from AA. Type 1b occurs when the upper and middle segments are supplied by a single artery from the AA, while the lower segments are supplied by separate, single vessels from the AA. The lower segment arteries can arise from the AA by a common trunk, while the upper and middle segments are supplied by either a single (type 1c) or multiple (type 1d) RAs on either side. The isthmus may also be supplied by arteries that arise inferior to the fused segment, which can be unilateral or bilateral and may originate from the AA independently or via common trunk (type 1e). Lastly, the fused lower segments may derive supply on one or both sides from the CIA, hypogastric, or middle sacral arteries (type 1f) (Boatman et al. [1971\)](#page-13-7). Figure [3](#page-4-0) demonstrates these variations in arterial supply. Literature review demonstrates that most cases are types 1e (28%) and 1f (24%) (Natsis et al. [2014\)](#page-13-3). It is worth mentioning that HSK is often associated with IVC abnormalities. These include double IVC, left IVC, and preisthmic IVC. Pre-isthmic IVCs cause retrocaval ureters, which are a direct cause of hydronephrosis and UPJ obstruction (Natsis et al. [2014\)](#page-13-3).

Classically, urologists have been concerned by the potential of a retrorenal colon in HSK. However, studies have shown this occurs in $\lt 1\%$ of HSKs and has not been shown to affect PCNL outcomes in in these patients (Ding et al. [2015\)](#page-13-8).

4 Stone Disease in the Horseshoe Kidney

Nephrolithiasis is the most common complication in HSK and essentially all stone types have been described. The incidence ranges from 21 to 60% (Yohannes and Smith [2002\)](#page-14-1). Anatomic urinary stasis arising from the anterior orientation of the renal pelvis, abnormal ureteral course over the isthmus, and high ureteral insertion is

Fig. 3 Graves' classification of arterial variation in HSK. *U* upper, *M* middle, *L* lower, *AA* abdominal aorta, *RCIA* right common iliac artery, *LCIA* left common iliac artery (Natsis et al. [2014\)](#page-13-3)

believed to contribute to urinary tract infection and stone formation (Yohannes and Smith [2002\)](#page-14-1). It has also been proposed that the orientation of the calyces impairs drainage, resulting in stasis and stone formation. Stones are often multiple and there is significant increase in large staghorn stones (Natsis et al. [2014\)](#page-13-3). While stones can form anywhere within the upper urinary tract, the most common locations for stone formation are the renal pelvis and lower pole. It is also quite common to have multiple stones in multiple locations (Pawar et al. [2018\)](#page-13-9).

5 Other Surgical Management

The same treatment modalities used in the management of nephrolithiasis in normal kidneys can be considered in the HSK (Lavan et al. [2020\)](#page-13-10). However, some unique considerations must be made given the anatomic challenges.

Shockwave lithotripsy presents a reasonable option for management of small renal stones in HSK. Stone free rates range from 28%-80%, however many patients require > 1 intervention after SWL (Ding et al. [2015;](#page-13-8) Stein and Desai [2007\)](#page-14-2). This has been attributed to problems with energy localization for pelvic stones combined with poor fragment clearance secondary to impaired renal drainage. It is thus imperative to rule

out coexisting UPJ obstruction when considering SWL. Stones greater than 15 mm appear less likely to achieve stone-free status after SWL (Stein and Desai [2007\)](#page-14-2).

The use of ureteroscopy for the management of stones in HSK has been well described. Limited case series have reported stone free rates ranging from 70%- 88.2%. Generally, these required multiple procedures, were for stones < 2.0 cm, and were not assessed with postoperative computed tomography. Technical challenges arise during retrograde access of the collecting system due to the angle of deflection required by the more superolateral positioning of the ureteral insertion into the renal pelvis (Geavlete et al. [2022\)](#page-13-11).

6 Indications for PCNL in HSK

The main consideration when approaching stone disease in the HSK is the difficulty in gaining retrograde access that arises from the acute angle of deflection required by the more superolateral positioning of the ureteral insertion into the renal pelvis. Because of this, retrograde ureteroscopic single session stone-free rates in HSKs are typically $< 60\%$ (Ding et al. [2015\)](#page-13-8).

If retrograde management is ineffective, unsuccessful, or deemed impossible, antegrade management is the preferred approach. Stone-free rates for PCNL in the HSK range from 65.5% to 87.5%, further supporting antegrade management as the preferred initial approach (Vicentini et al. [2021\)](#page-14-3). Therefore, based on most data, antegrade management is preferred for most patients with nephrolithiasis in HSK to achieve stone-free status in a single operation. It should be noted that AUA guidelines currently recommend percutaneous nephrolithotomy as first-line therapy for staghorn stones and total renal stone burden > 20 mm (Assimos et al. [2016\)](#page-13-12). However, given the unique challenges associated with retrograde access in HSK, indications for PCNL in HSK can be expanded to include smaller renal stone burden.

7 Preoperative Evaluation

A complete history and physical is essential prior to proceeding with percutaneous access. Identification of contraindications precluding percutaneous access is paramount. Active urinary tract infection (UTI) and bleeding disorders are especially important to identify. It is also important to obtain a thorough surgical history when planning percutaneous access.

Regarding preoperative urine testing, both AUA and EAU guidelines state that urinalysis is required prior to any stone intervention and a urine culture should be obtained with clinical or laboratory signs of infection. Positive urine cultures should be treated with antibiotics until a sterile culture is obtained (Assimos et al. [2016\)](#page-13-12).

It is prudent to obtain a baseline complete blood count and serum electrolytes with renal function testing prior to proceeding with PCNL in HSK. This establishes

a baseline that can be followed post-operatively and could potentially identify preoperative conditions that may increase operative complications.

Preoperative imaging is required for proper surgical planning. AUA guidelines recommend clinicians obtain a non-contrast CT scan prior to performing PCNL (Assimos et al. [2016\)](#page-13-12). We prefer a CT stone-protocol on every patient prior to attempting PCNL in HSK. If planning a prone approach, ideally this CT is obtained with the patient in a prone position. This allows for evaluation of total stone burden, prone relationship to adjacent structures, and anatomic assessment of the HSK. Per AUA guidelines, in patients with complex stones or anatomy, clinicians may obtain additional contrast imaging if further definition of the collecting system and the ureteral anatomy is needed (Assimos et al. [2016\)](#page-13-12). Therefore, vascular contrast enhancement and/or CT urogram can also be considered in select patients.

8 Operating Room Setup

While recognizing that each surgeon will have preferences regarding the specific operating room set up for PCNL, we believe we can provide some general recommendations that we have found beneficial in our practice.

Surgeon safety and ergonomic considerations are fundamental for any endoscopic operating endeavor, and PCNL is no different. Radiation protection via lead impregnated aprons, thyroid shields, eyeglasses, etc. is imperative for any endourological surgery that uses fluoroscopic image guidance. All monitors used for the procedure (endoscope, X-ray) should be placed at eye level and in a location that requires minimal turning of the surgeon's head.

Our operating room set up is as follows. The surgeon, surgical assistant, and back table are positioned on the side to be treated. The endoscopic video monitor is positioned opposite the surgeon, near the patient's head. The X-ray monitor is positioned opposite the surgeon, near the patient's feet. The C-arm is positioned between these two monitors, above the patient's knees. Irrigation, lithotripter and/or LASER generators, suction, and other devices that may be required are placed at the foot of the bed.

9 Patient Positioning

The specific details regarding positioning for PCNL are covered in a separate chapter. For this reason, we provide limited commentary on positioning for PCNL in HSK. For both prone and supine positions, it is essential to ensure the patient is perfectly centered on the table. Deviations from center can cause difficulty with image interpretation as the C-arm orbits the patient, as well as possible instability and fall risk (Smith et al. [2019\)](#page-14-4).

Prone position is most commonly used for HSK as it provides several advantages, which include larger surface area for puncture site, more room for instrument movement, and possibly a lower risk of visceral organ injury. It also allows upper pole puncture to be performed more easily in the HSK (Osther et al. [2011;](#page-13-13) Pérez Fentes [2021\)](#page-13-14). Several disadvantages with prone positioning exist. Increased radiologic hazard to the urologist, patient discomfort, number of operating room staff needed for correct positioning, risk of pressure point injury, difficulty with retrograde access, circulatory and ventilator difficulties, as well as alteration in endocrine and pharmacokinetic effects have all been associated with prone positioning (Pérez Fentes [2021\)](#page-13-14). These are uncommon and we have rarely encountered positioning complications, even in the morbidly obese.

While supine PCNL in HSK is gaining popularity, it is still relatively uncommon (Osther et al. [2011](#page-13-13)**).** Therefore, the data on supine PCNL in HSK is quite limited, and most commentary is extrapolated from PCNL in anatomically normal kidneys.

Generally, supine PCNL appears to be as safe and effective as prone PCNL regarding stone free rates, transfusion, and complications. For a variety of factors related to repositioning, supine PCNL has been associated with reduced operative times. Supine positioning also provides easier access to the urethral meatus for retrograde access (Kumar et al. [2012\)](#page-13-15).

Five common supine positions have been described. Valdivia supine PCNL was the first described supine position for PCNL. Modifications to this position include: Galdakao-modified supine Valdivia (GMSVP), Barts Modified Valdivia, complete supine, and complete supine flank-free (Kumar et al. [2012\)](#page-13-15).

The data on supine PCNL specifically in the HSK is quite limited. Pérez-Fentes described a case report of one 42F with a complete staghorn stone in the right moiety of a HSK. The patient was treated with endoscopic combined intrarenal surgery and positioned in GMSVP and required 3 separate procedures to achieve stone-free status (Pérez Fentes [2021\)](#page-13-14).

Gupta et al. have described a case series of 5 supine tubeless PCNLs in HSKs. This series included 4 patients (one with BL nephrolithiasis) and all were operated on in GMSVP (Gupta et al. [2022\)](#page-13-16).

Sohail et al. published a case report of one 45 M with two 1.5 cm renal stones. Stone-free status was achieved with one puncture and one procedure in "complete supine flank-free" position (Sohail et al. [2017\)](#page-14-5).

Vincentini et al. reported a multicentric retrospective analysis of 106 PCNLs in HSKs. Approximately 37% were treated in supine position. There was no difference in transfusion, complication, and immediate success rates when compared to prone. Surgical time was significantly longer in the prone group (Geavlete et al. [2022\)](#page-13-11). Based on this literature, supine PCNL is considered a safe and effective in HSK while carrying a low complication rate. It may be considered an option for treating stones in patients with HSK (Vicentini et al. [2021;](#page-14-3) Pérez Fentes [2021;](#page-13-14) Kumar et al. [2012;](#page-13-15) Gupta et al. [2022;](#page-13-16) Sohail et al. [2017\)](#page-14-5).

10 Retrograde Injection

An open-ended ureteral catheter is placed via cystoscopy on the side to be treated. The timing of this placement depends on the planned operative position, as well as the type of operating table used. The ureteral catheter is then used to aid in retrograde imaging for identification of calyces when planning percutaneous puncture. It may also help reduce migration of stone fragments down the ureter during lithotripsy. We use a combination of contrast and air to completely characterize the calyceal anatomy. We have found air to be particularly useful in delineating the posterior calyces.

11 Percutaneous Puncture

The optimal calyx of entry is determined based on the preoperative CT scan, retrograde imaging, anatomic considerations, and stone location. The preoperative CT scan is useful for identifying a safe percutaneous window to avoid adjacent structures, while the intraoperative imaging guides percutaneous placement of the puncture needle.

The ultimate goal is to target a calyx that affords maximum stone clearance with a single puncture. Because of the downward displacement and malrotated axis of the HSK, calyxes tend to be oriented more inferiorly and posterolaterally, making subcostal puncture preferred in most cases (Stein and Desai [2007\)](#page-14-2). The malrotated HSK and its relationship to adjacent structures also results in a more medial cutaneous puncture site than in anatomically normal kidneys. Cutaneous puncture is generally made near the posterior axillary line or slightly medial. Upper pole access is most often chosen in HSKs as this allows access to upper pole calyces, renal pelvis, ureteropelvic junction, and proximal ureter, with minimal torque on the renal parenchyma and lower risk of injury to adjacent structures. Figure [4](#page-9-0) demonstrates fluoroscopic upper pole puncture yielding maximal collecting system access with minimal deflection angles. It should be noted that the puncture site is more medial than in a normal kidney, but access to the calyx is more lateral than in a normal kidney. This results in a longer than normal access tract, which is discussed in a later section.

As previously mentioned, the vascular anatomy of HSK is highly variable. However, blood vessels typically enter the kidney anteromedially and thus risk of vascular injury during percutaneous access is equivalent to that of a normal kidney. The exceptions to this are the arteries supplying the isthmus, which can arise laterally off the common iliac vessels (Boatman et al. [1971\)](#page-13-7).

Fig. 4 Intraoperative fluoroscopic image demonstrating upper pole puncture in a HSK. Note the minimal deflection angles required to access the entire collecting system [original image]

12 Fluoroscopic Guidance

The use of C-arm is imperative to understanding the 3-dimensional anatomy of the collecting system. The C-arm is orbited around the patient, both towards and away from the surgeon. A combination of spot and live images are used to understand the 3-dimensional anatomy of the collecting system.

Generally, the collecting system is opacified and distended with a combination of contrast and air. A scout film is then obtained in the anterior–posterior plane. A target calyx is chosen based on this image and the preoperative CT scan. A percutaneous access needle is then advanced slightly into the subcutaneous tissue in a straight line towards the desired calyx. Live rotation between different views can aid in understanding of the 3-dimensional relationships between calyxes and the renal pelvis. The C-arm is then rotated to an appropriate axis as to provide a direct, end-on view of the needle hub, shaft, and target calyx in a "bull's-eye" fashion. Once the appropriate trajectory is established, the C-arm is rotated to an axis that allows the surgeon to monitor the depth of needle insertion into the target calyx. Ideally, access is obtained along the axis of the calyx and through the papilla, thus avoiding the vascular infundibulum (Smith et al. [2019\)](#page-14-4). The inner stylet is removed, and access is confirmed by return of urine and/or contrast.

13 Tract Dilation

A hydrophilic, angled-tip guidewire is advanced through the lumen of the puncture needle. This wire should advance relatively easily. Aggressive probing should be avoided, as false tracts can be created. Because of the malrotated nature of the HSK and resultant acute angle of the UPJ, it can be exceedingly difficult to place the wire

down the ureter. Thus, the luxury of having a wire down the ureter might have to be sacrificed due to the technical difficulty that arises from the altered anatomy. Instead, one may have to accept a wire coiled in the renal pelvis as adequate access for tract dilation. It is imperative that the wire is at least in the renal pelvis prior to tract dilation. This can be confirmed with repeat pyelogram as needed.

Dilation of the tract can then be performed via the operative surgeon's preferred method. In our practice, a balloon dilator is inflated to a pressure of 20–30 atm with contrasted material. Periodic spot fluoroscopic images are obtained to ensure all "waists" have been fully expanded.

The nephrostomy sheath is then advanced over the balloon under live fluoroscopy until the tip of the sheath rests at the distal end of the balloon. Given the more anterior position of the HSK, the renal pelvis may be deeper in relation to the skin than in the anatomically normal kidney. This can result in the inability to reach the middle and lower calyces with rigid instruments, especially in obese patients. Preoperative measurements can help determine if an extra-long nephroscope and/or nephrostomy sheath may be needed to overcome this problem. If there is concern that the sheath will become buried in the subcutaneous tissue, another option is to place a stay suture through the end of the nephrostomy sheath. The sheath can then be buried in the subcutaneous tissue to achieve extra reach. The previously placed stay suture is then used to retrieve the buried nephrostomy sheath at conclusion. Additionally, extra-long rigid scopes may be necessary.

14 Nephroscopy

Once the nephrostomy sheath is in place, the balloon is deflated, removed, and the nephroscope is inserted with the lithotripter and suction deployed. The rigid nephroscope is used to remove as much stone as possible. As previously mentioned, the lower pole calyces can be very difficult to visualize in the HSK. Therefore, the rigid nephroscope may not be effective in removing the entire stone burden. It should be emphasized that use of the flexible endoscopy is mandatory to assure all calyces are interrogated, which is discussed in more detail in a later section.

15 Stone Extraction

We use a wide variety of methods for stone extraction. These include manual basket extraction, ballistic lithotripters, ultrasonic lithotripters, and combination lithotripters. In special cases requiring treatment via flexible nephroscopy, Ho:YAG LASER is our preferred energy for laser lithotripsy.

The presence of a HSK neither necessitates nor limits the use any specific lithotripter. Lithotripsy technique is determined more by stone location than overall stone burden. Given that HSKs tend to have increased stone burden and stones in

multiple calyces, there is increased requirement for flexible endoscopy to achieve stone-free status (Raj et al. [2003\)](#page-13-17). Thus, LASER is used more commonly when treating stones in the HSK. It is our opinion that this could increase the use of dusting technique as manual stone extraction via flexible endoscopy is not ideal. Suction devices can then be used after completion of dusting to increase stone-free rates.

16 Flexible Endoscopy

AUA guidelines state that flexible nephroscopy should be a routine part of standard PCNL, and we strongly believe PCNL in HSKs to be no different (Assimos et al. [2016\)](#page-13-12). Upper pole access in HSKs results in a tract that is longer than normal, which causes difficulty in reaching the middle and lower calyces (Stein and Desai [2007;](#page-14-2) Gupta et al. [2022\)](#page-13-16). It has been demonstrated that > 80% of PCNL in HSKs require flexible nephroscopy to adequately access stones in all calyces (Raj et al. [2003\)](#page-13-17). Ideally, flexible nephroscopy is used to reposition stones for removal via rigid nephroscopy. If repositioning is not feasible or unsuccessful, flexible nephroscopy can then be used to treat stones via laser lithotripsy. It is our recommendation and practice that flexible endoscopy should be performed at the conclusion of all PCNLs in HSKs.

17 Exit Strategy

In our opinion, drainage is required after any PCNL in HSKs. This is ultimately at the discretion of the operative surgeon and the type of drainage should be whatever is preferred. In our practice, a 16Fr tipless Foley catheter is left with the balloon inflated in the access tract. This provides a few advantages. Re-establishment of the access tract is easier if subsequent procedures are required. The larger diameter of the Foley provides maximal drainage, but generally causes the patient less pain than a larger or more rigid nephrostomy tube.

18 Complications

The expected complications that arise from PCNL in HSKs are similar to those with PCNL in anatomically normal kidneys. Complications include: bleeding, bleeding requiring transfusion, fever, and collecting system perforation. Less commonly, vascular injury, hydrothorax, sepsis, and visceral injury are seen. Current literature suggests that PCNL in HSK complication rates are comparable to PCNL in the anatomically normal kidney (Vicentini et al. [2021;](#page-14-3) Osther et al. [2011\)](#page-13-13). It could be

argued that, given the high rate of subcostal access, there is less risk of injury to the plural space when performing PCNL in the HSK as compared to PCNL in the anatomically normal kidney. Based on the current literature and our experience, we believe PCNL to be a safe and effective approach to the management of stone disease in HSKs.

19 Postoperative Considerations

Patients are observed in the hospital overnight after surgery. While it is our practice to obtain a non-contrasted CT of the abdomen/pelvis and an antegrade nephrostogram the morning of post-operative day one, these may not be required in all cases. The CT scan is useful in proving stone-free status and helps guide patient counseling if there are residual stone fragments. If stone fragments reside at the UPJ and/or renal pelvis, the kidney often will not drain postoperatively. In patients where there is concern for postoperative drainage, a nephrostogram may be of benefit prior to removing the nephrostomy tube. If the patient is deemed stone free, the drain is removed and the patient is discharged home on postoperative day one.

If residual stone is revealed on follow up imaging, these findings are discussed with the patient. Depending on the patient, stone burden, and other factors, we offer immediate or delayed antegrade versus retrograde ureteroscopy. In appropriate situations, observation may also be offered. Shared-decision making is imperative in this discussion and is often the most important factor when deciding how to proceed.

20 Follow up

Patients are seen in our clinic for a routine post-operative visit to perform a wound check and review stone analysis results. All patients are then seen at 6 month follow up intervals with renal ultrasound for stone surveillance.

21 Conclusion

The horseshoe kidney presents a unique operative challenge to the urologist when managing stone disease in this patient population. A thorough understanding of the embryology and anatomic variation that arises is crucial to the safe and effective treatment of these patients. With careful consideration of these differences, percutaneous intervention is safe and effective in the horseshoe kidney. Key points of percutaneous nephrolithotomy in the horseshoe kidney include: gaining upper pole access is preferred, ensuring the guidewire is within the renal pelvis or ureter prior to tract dilation is paramount, and performing flexible endoscopy at the conclusion of any percutaneous intervention in the horseshoe kidney is required. By understanding the anatomic variations and adhering to the principles described in this chapter, the urologist can safely and effectively approach stone disease percutaneously in most any patient with a horseshoe kidney.

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