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Urolithiasis in the horseshoe kidney: a single-centre experience

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OBJECTIVE

To report the operative management and subsequent stone-free rates of patients with urolithiasis in a horseshoe kidney and treated at one centre.

PATIENTS AND METHODS

We retrospectively reviewed all patients presenting to our centre with a horseshoe kidney and urolithiasis over a 15-year period. The stone burden, surgical management, complications and stone clearance rates were recorded.

RESULTS

In all, 55 patients with urolithiasis in horseshoe kidney were treated. Percutaneous nephrolithotomy (PCNL) was used in 60 renal units in 47 patients. Five patients had extracorporeal shock wave lithotripsy (ESWL), two had flexible ureteroscopy and one had a laparoscopic pyelolithotomy for a stone extending into the isthmus. PCNL was used for large stones (mean digitized surface area = 614.32 mm^2) and required one to four stages to achieve an overall stone clearance rate of 88%. Stones were cleared at one sitting in 77% of PCNL procedures, completely cleared in two-thirds of patients treated by ESWL, and in both who had flexible ureteroscopy and the one treated with laparoscopic pyelolithotomy. Complications were minimal, with 15% minor and 3% major complications in the PCNL group only.

CONCLUSION

Appropriate management of urolithiasis within the horseshoe kidney depends not only on stone burden, but also on stone location, calyceal configuration and malrotation. Stones can be cleared successfully in almost all patients providing that all techniques are available to the operating surgeon.

KEYWORDS

horseshoe kidney, urolithiasis, percutaneous nephrolithotomy, ESWL, laparoscopic nephrolithotomy

INTRODUCTION

The horseshoe kidney is the most common renal fusion anomaly, affecting 1 in 400 of the population [1]. Originating from abnormal fusion of the lower poles of the developing kidneys, the ascent of the kidneys is subsequently arrested by the inferior mesenteric artery. As a result, normal rotation of the kidneys is prevented, leading to malrotation with anterior displacement of the collecting system. Ureteric insertion onto the renal pelvis tends to be both superiorly and laterally displaced. Although most horseshoe kidneys are asymptomatic, the anatomical abnormalities predispose to impaired drainage of the collecting system, urinary stasis and an increased incidence of PUJ obstruction, infection and calculus formation.

Urolithiasis is the most common complication of horseshoe kidney, with a reported incidence of 21–60% [2]. The surgical treatment of stones in horseshoe kidneys has developed endourologically, just as for stones in anatomically normal kidneys, with open surgery now practically obsolete. Percutaneous nephrolithotomy (PCNL) and ESWL are the most commonly reported methods for treating stones in horseshoe kidneys. More recently, flexible ureteroscopy and laparoscopic pyelolithotomy have also been shown to be effective and valuable management options. We report our single-centre experience of stone management in symptomatic patients with horseshoe kidneys.

PATIENTS AND METHODS

We retrospectively reviewed patients with stones in a horseshoe kidney presenting to our centre between July 1991 and February 2008; in all, 55 were treated for urolithiasis in a horseshoe kidney within that period. The patients were treated using several methods, including PCNL, ESWL, flexible and rigid ureteroscopy, and laparoscopic pyelolithotomy.

The choice of treatment depended on stone size/location and the appearance of the pelvicalyceal system on preoperative imaging. In most cases with a large stone bulk, we find PCNL a safe and effective treatment option in horseshoe kidneys. However, in some cases of lower calyceal and isthmus stones, the nephroscope might not reach the stone, either due to length limitations or because of the angulation present within the pelvicalyceal system. A further limitation to the use of PCNL might be in severely malrotated kidneys where all calyces face anteriomedially. In these situations we opted for laparoscopic pyelolithotomy as the most appropriate treatment to clear the stones. We reserve ESWL for solitary stones with evidence of free drainage in a suitable pelvicalyceal

TABLE 1 *Characteristics of the 55 patients presenting with stones in horseshoe kidneys*

Characteristic	*n* (%) or *n*
No. stone bearing renal units	69
Mean (range) age, years	36.5 (7–60)
Male	49
Female	6
Presentation	
Pain	40 (73)
Haematuria	4 (7)
Pain and haematuria	6 (11)
Fever	1 (2)
Anuria	1 (2)
Unknown	3 (5)
Mean serum creatinine level, mg/dL	0.97
Urine culture	
Sterile	49
Escherichia coli	4
Proteus	1
Klebsiella	1
Associated PUJ obstruction	3
(previous pyeloplasty)	1
(bilateral endopyelotomy)	1

system, and flexible ureteroscopy for small primary or recurrent stones.

Patient demographic data were collected, with characteristics of the stones, preoperative investigations, operative details, complications, stone clearance rates and follow-up. Stone analysis and metabolic analysis has been used at our centre since 2002, and data were available only for 2002–2008.

As well as location, we calculated stone digitized surface area (DSA), as previously described by Raj *et al.* [3], computed as the sum of the products of the maximum dimensions of stones on a plain X-ray. For example, for a stone with dimensions of 5×7 mm the DSA was 35 mm^2. For a patient with two stones of 5×7 and 7×10 mm, respectively, the DSA was $35 + 70 = 105$ mm^2.

RESULTS

Table 1 summarizes the preoperative findings for the 55 patients ; the mean patient age was 36.5 years, with a male to female ratio of 8:1. Within these patients there were 69 stone-bearing renal units; 13 patients had bilateral urolithiasis and one patient presented twice. Before CT was available in our centre in 2006, all patients were preoperatively imaged using ultrasonography (US) and IVU. Once CT became available it has been used routinely in all patients with a horseshoe kidney to accurately determine stone location and volume before surgery.

Stone characteristics by subsequent stone management are shown in Table 2; 47 patients (86%) had PCNL to manage their stone, with 13 requiring bilateral PCNL, giving a total of 60 renal units. Upper calyceal access was used in 29 (48%) PCNLs. In 21 procedures a solitary upper calyceal tract was used, whilst in eight patients the upper-pole access was combined with mid- or lower-pole access in a multi-tract procedure.

The number of PCNL stages required for stone clearance depended on both stone configuration and size (Table 3). In six patients with a staghorn calculus only two were rendered stone-free in a one-stage PCNL; two required a two-stage procedure, one a three-stage and one a four-stage PCNL. Of the 17 patients who had multiple stones, 12 (71%) were stone-free in one sitting; this compares with 30 (86%) of 35 who had neither multiple nor staghorn stones. The number of tracts required, operative time, mean decrease in haemoglobin level and hospital stay all correlated positively with the number of PCNL stages required, increasing as the number of stages increases.

The overall stone-free rate in the PCNL group was 88%; in four patients small residual fragments were treated conservatively (mean DSA 18.9 mm^2); in one the residual stone bulk was felt to be significant and the patient was advised to undergo ESWL.

Details of the five patients treated by ESWL are shown in Table 4; six relatively small (mean DSA 149 mm^2) solitary renal stones were treated. IVU before treatment showed prompt bilateral excretion of contrast medium in all patients. US showed mild hydronephrosis in two patients with a renal pelvic stone, and in one with a lower calyceal stone. The remaining renal units had no evidence of hydronephrosis. Stones were completely cleared in four patients at the 1-month follow-up. Of the remaining two patients, one had residual fragments of <4 mm and one of >4 mm; both patients were treated conservatively.

Two patients had flexible ureteroscopy; one had a single pelvic stone (DSA 63 mm^2) and the second had four stones, with one each in the pelvis, superior calyx, middle calyx and isthmus (DSA 281 mm^2). Neither patient had any ureteric calculus. In both patients stone fragmentation was complete. One further patient with bilateral renal calculi in horseshoe kidneys (DSA 80 and 12 mm^2) was found to have a solitary functioning left kidney with a large left distal ureteric calculus. In view of the minimal symptoms and significant comorbidities, after ureteric stone clearance via rigid ureteroscopy, the renal stones were treated conservatively.

One final patient had laparoscopic pyelolithotomy for a large staghorn calculus extending into the isthmus (Fig. 1). The pelvicalyceal system configuration was unsuitable for PCNL. The stone was completely cleared within an operative duration of 3 h. There were no complications and the patient was discharged four days after surgery.

The stone composition was determined in 12 of the 55 patients; all stones were predominantly calcium oxalate dihydrate. Eleven patients had a metabolic analysis; seven had hyperoxaluria, eight hypercalciura, two hypocitraturia and three hypomagnesuria. None had a normal metabolic study, with all patients having at least one abnormal factor.

TABLE 2 Stone characteristics by subsequent stone management

Characteristic	PCNL	ESWL	Ureteroscopy Flexible	Ureteroscopy Rigid	Laparoscopic pyelolithotomy
N patients	47	5	2	1	1
N renal units treated	60	6	2	1	1
Mean stone DSA, mm^2	614.3 (99–3850)	149.2 (50–225)	172 (63–281)	44 (28–60)	1662
Multiple stones present, *n* (%)	16 (27)	0	1	1	1
Staghorn, *n* (%)	6 (10)	0	0	0	0

TABLE 3 PCNL results by number of PCNL stages required

Variable	No. of stages: 1	2	3	4
No. of patients	46	12	1	1
Mean stone DSA, mm^2	448.7 (99–2100)	1070 (225–3850)	1950	975
% or *n*/*N* single tracts (max tract no.)	94 (2)	2/3 (3)	0 (4)	0 (6)
Mean (range) operative duration, min	63.0 (30–120)	97.3 (40–195)	180	Unknown
% or *n*/*N* complete clearance	87	4/6	1/1	Residual fragments
Mean decrease in haemoglobin, g/dL	1.2 (0–2.3)	1.4 (0.2–3)	2	5
Complications*	p/o pyrexia; 4 (m) i/o bleeding, 1 (m) JJ stent, lost to FU, 1	p/o pyrexia, 2 (m) PC system perforation 1 (M) Bleeding requiring clot evacuation, 1 (M)	p/o pyrexia, 1 (m)	p/o pyrexia, 1 (m)
Mean hospital stay, days	4.9 (2–11)	9.6 (5–14)	10	20

**Complications were characterized as major (M) if they required additional intervention or resulted in prolonged hospitalization, and minor (m) if they could be conservatively managed with no additional intervention. p/o, postoperative; i/o, intraoperative; FU, follow-up.*

TABLE 4 The results of ESWL in five patients

Variable	Patient: 1	2	2	3	4	5
Stone site	Upper calyx	Pelvis	Pelvis	Lower calyx	Pelvis	Pelvis
DSA, mm^2	50	225	196	88	216	120
ESWL sittings, *n*	1	2	1	1	2	1
Total shocks, *n*	1000	3000	2000	1000	3000	1500
Maximum shockwave energy, kV	13	13	13	13	13	13
Clearance at 1 month follow-up	Incomplete	Complete	Complete	Complete	Incomplete	Complete
Total follow-up, months	1	1	1	3	3	1

DISCUSSION

Historically, the most common cause of stone formation in the horseshoe kidney was believed to be secondary to UTI and urinary stasis related to the abnormal anatomy of the PUJ and aberrant ureteric course. However, Evans and Resnick [4] found a high incidence of treatable metabolic abnormalities in a study of eight patients with a horseshoe kidney, and other authors also identified a high incidence of metabolic abnormalities within those with a horseshoe kidney [5,6]. There were few patients who had a metabolic analysis in the present study, as the facility for analysis was not available until 2002. Despite this, all of those who had a metabolic analysis had at least one metabolic abnormality. Our experience is in keeping with that of Raj *et al.* [7], who found an average of 2.8 metabolic abnormalities per 24 h urine collection in 11 patients with a horseshoe kidney. The findings in the present series support the theory that metabolic abnormalities, in addition to anatomical variations causing urinary stasis, contribute significantly to stone formation in the horseshoe kidney.

In patients with normal renal anatomy, access to the upper pole calyces during PCNL often requires a supracostal approach, with the associated risk of pleural injury. However, in the horseshoe kidney upper-pole access is

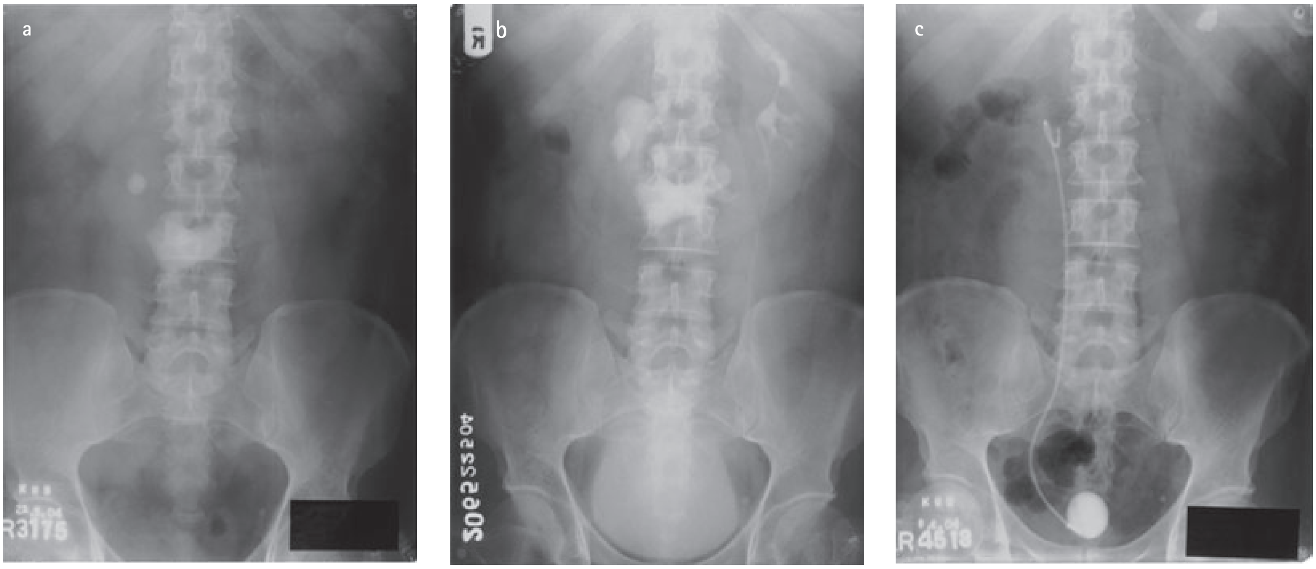

FIG. 1. Radiographs of a patient who had a laparoscopic pyelolithotomy; (a) is the plain film before surgery (b) the IVU 20 min after surgery and (c) the plain film after surgery.

relatively safe due to the inferior displacement of the kidneys away from the pleura. Furthermore, upper-pole access is a valuable aid to stone clearance, as the alignment of the nephroscope with the long axis of the kidney aids manipulation of the scope into the upper calyces, renal pelvis, lower calyces, PUJ and proximal ureter. Our experience confirms the safety of the upper-pole approach to the horseshoe kidney. We used upper-pole access in 48% of PCNL procedures, with no associated pleural injury. In a study of 24 patients with stones in a horseshoe kidney, Raj *et al.* [3] reported upper-pole access in 63%, with one pneumothorax. In that study it was also noted that the use of long rigid nephroscopes and flexible nephroscopes was usually required for a satisfactory stone clearance rate, given as 87.5%. It has not been our experience that long nephroscopes or flexible scopes are a necessity in treating patients with horseshoe kidneys by PCNL; in the present series, a flexible nephroscope was used in only one patient. However, the reason for this difference in opinion is probably to be due to there being more multiple tract PCNLs in the present series. We used multiple tract procedures in 10 patients (17%) compared with just one (4%) in the series by Raj *et al.* [3]. Supporting this observation, in a series of 12 horseshoe kidney treated by PCNL, Al-Otaibi and Hosking [8] reported the routine use of a second nephrostomy tract, which was noted to be essential for complete stone removal. These authors noted that flexible nephroscopy was particularly helpful in patients with a horseshoe kidney, but concluded that urologists using PCNL in horseshoe kidneys should be prepared to use multiple nephrostomy tracts and to establish these tracts during surgery as necessary; we agree with this statement.

To our knowledge, the present series represents the largest single-centre experience of managing urolithiasis within the horseshoe kidney. Although our experience with ESWL was limited the stone-free rate was four of six (stones) after ESWL, and in keeping with the largest published series to date, that gave stone-free rates of 67–76% [9–14]. In general, our preference for stone treatment in horseshoe kidneys has been very much towards PCNL and our stone-clearance rate of 88%, with minimal complications, shows the safety and success of this approach at our institution. Other authors have also found PCNL to be the treatment of choice in these patients, who often have a very significant stone bulk. Stone clearance rates in series of 12–45 renal units are 75–87.5% [3,8,15].

Flexible ureteroscopy in horseshoe kidneys has been reported in a few patients to date. Andreoni *et al.* [16] reported the use of an access sheath to aid flexible ureteroscopy in a horseshoe kidney and more recently and Weizer *et al.* [17] showed flexible ureteroscopy to be effective in four patients, with clearance in three. We consider flexible ureteroscopy to be a valuable treatment option in horseshoe kidneys, allowing the fragmentation and removal of stones which might not otherwise pass freely from the aberrant drainage system.

The laparoscopic approach has also been reported in the stone-bearing horseshoe kidney, and with good effect. One group [18] used laparoscopy to guide a PCNL, whilst Nambirajan *et al.* [19] described a successful bilateral laparoscopic pyelolithotomy. The use of laparoscopy is particularly helpful with a very large stone burden at the isthmus, where negotiating a rigid nephroscope is impossible and the stone burden makes flexible nephroscopy impractical.

In conclusion, our experience shows that PCNL is a safe and effective treatment for stones in horseshoe kidneys, providing excellent stone-clearance rates. This study also shows that for smaller stone burdens, ESWL and flexible ureteroscopy are viable treatment options with acceptable stone clearance rates. However, appropriate management of urolithiasis within the horseshoe kidney depends not only on stone burden, but also stone location and the presence of further anomalies such as malrotation. Thus, in some circumstances alternatives to PCNL, ESWL and flexible ureteroscopy must be considered. Our experience confirms laparoscopic pyelolithotomy to be a useful adjunct to the more standard management approaches in this challenging group of patients. Stones can be cleared successfully in almost all patients, providing all techniques are available to the operating surgeon.

CONFLICT OF INTEREST

None declared.

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Abbreviations: **PCNL**, percutaneous nephrolithotomy; **DSA**, digitized surface area; **US**, ultrasonography.