Urol Res (2006) 34:291–298
DOI 10.1007/s00240-006-0059-z

ORIGINAL PAPER

Treatment of calculi in kidneys with congenital anomalies: an assessment of the efficacy of lithotripsy

Adel R. Al-Tawheed · Khaleel A. Al-Awadi · Elijah O. Kehinde · Hamdy Abdul-Halim · Akram M. Hanafi · Yusuf Ali

Received: 10 November 2005 / Accepted: 15 May 2006 / Published online: 29 June 2006

Abstract We studied the effectiveness of extracorporeal shock wave lithotripsy (ESWL) in the treatment of stones in kidneys with congenital anomalies to determine factors that may affect the results. Patients found to have renal calculi in kidneys with different types of congenital anomalies were treated using ESWL. All patients were investigated by intravenous urography (IVU) to confirm the diagnosis. J stents were inserted prior to therapy in renal units with calculi exceeding 1.5 cm in diameter. Complications encountered and factors affecting success using this treatment modality were analysed. Twenty-five patients (18 males, 7 females) were studied between August 1988 and July 2005. There were nine patients with horseshoe kidneys, eight with ectopic kidneys, three with malrotated kidneys, two with duplex renal system, and one patient each with polycystic kidneys and hypoplastic kidney. The IVU showed 31 isolated calyceal or renal pelvic stones with mean stone burden of 1.44cc. All 25 patients were treated by lithotripsy. Twenty-four (77.4%) renal units (in 19 patients) were completely cleared of stones, 2 (6.5%) renal units (2 patients) were partially cleared of calculi and the procedures failed in 5 (16.1%) renal units (4 patients). Out of five renal units in which the procedures failed, open surgery was performed in three renal units and percutaneous nephrolithotomy (PCNL) was performed in two. None of the 25 patients developed any major complications. No significant adverse changes in renal function tests were observed at 3-month follow-up. The stone-free rate was influenced and reduced by stone size and location in the pelvi–calyceal system. Calculi in kidneys with congenital anomalies may be treated successfully by ESWL as a first-line therapy in the majority of patients. With position modifications, localization of stones may be facilitated and disintegrated. The outcome in patients so treated does not differ significantly from that in those with normal kidneys

Keywords Congenital renal anomalies · Calculus · Treatment · Lithotripsy

This paper was presented as a poster at the 11th European Symposium on Urolithiasis, Coburg, Germany. June 16–18, 2005. Poster No ESWL 8.

A. R. Al-Tawheed · K. A. Al-Awadi · H. Abdul-Halim · A. M. Hanafi · Y. Ali
Department of Surgery (Division of Urology), Mubarak Al-Kabeer Hospital, Safat, Kuwait

E. O. Kehinde (✉)
Department of Surgery (Division of Urology), Faculty of Medicine, Kuwait University, P.O Box 24923, 13110 Safat, Kuwait
e-mail: ekehinde@hsc.edu.kw

Introduction

The incidence of stone disease among patients with congenital renal anomalies varies widely and depends on the type of renal anomaly. Twenty percent of patients with horseshoe kidneys and polycystic kidney diseases develop renal stones [1]. Poor urine drainage and stasis are the major causes of stone formation in congenital renal anomalies; rather than metabolic and genetic causes which may assist stone formation in polycystic kidney disease [2, 3]. Therapeutic options in such cases are: extracorporeal shock wave lithotripsy

(ESWL), open surgery, percutaneous nephrolithotomy (PCNL) and laparoscopic approach [2, 4]. Although ESWL has dramatically changed the management of urolithiasis, the unique anatomical properties of kidneys with congenital anomalies present substantial obstacles to the use of ESWL in these disorders [5]. In this study, we present our experience with the treatment of calculi in patients with different types of congenital renal anomalies using ESWL. We analyse the factors that influenced the stone-free rates in such patients.

Patients and methods

From August 1988 to July 2005, patients attending the Stone Clinic, Mubarak Al-Kabeer Hospital, Kuwait with different types of congenital renal anomalies and urolithiasis were studied. Complete blood count (CBC), renal and liver function tests, serum electrolytes, coagulation profile as well as urine culture were performed in all patients. Intravenous urography (IVU) was performed in all patients and spiral computed tomography was performed in three cases. Patients with coagulopathy, previous renal reconstructive surgery or renal function of affected kidney < 15% were excluded from the study. Treatment of patients with ongoing UTI was delayed until infection was successfully treated. A 'J' stent was inserted if renal calculus was more than 1.5 cm in diameter or if there was poor drainage of the renal unit on IVU or renogram. Renogram was requested if poor renal function was suspected by the finding of reduced cortical thickness on ultrasound of kidneys, ureters and bladder (KUB) or of a marked delay in the excretion of dye on IVU.

Patients were treated by extracorporeal piezoelectric lithotripsy (EPL) using a Richard Wolf Piezolith 2300 machine (Knittingen, Germany) from 1988 to 2001 or extracorporeal shock wave lithotripsy (ESWL) using a Siemens lithostar C (Erlangen, Germany) from 2002 to 2005. The latter device uses an electromagnetic acoustic source shockwave generator. All patients had treatment either in the supine, lateral or prone position or slight modifications of these positions if the shadow of the spine made localization difficult. The treatment protocol consisted of a maximum of 4,000 and 3,000 shocks per session for EPL and ESWL respectively. The energy setting was 16 kV. Treatment sessions ranged between 30 and 45 min. No analgesia was given except in patients who were unable to tolerate the procedure. Lithotripsy sessions were conducted as an outpatient service once a week. The effect of ESWL on renal calculus size was assessed by plain X-ray and ultrasound examination of KUB. If after five sessions of ESWL there was no substantial fragmentation of the calculi for any reason, treatment with ESWL was discontinued. Renal function tests, serum electrolytes as well as urine culture were done at the completion of stone clearance followed by removal of the double J stent under local anesthesia, IVU and ultrasound of KUB and blood tests were repeated at 3-month intervals. A patient was declared stone-free if there was complete clearance of disintegrated stones or residual fragments less than 0.4 cm in diameter on X-ray of KUB if the calculus was radio-opaque or ultrasound of KUB if radiolucent. Treatment failure was diagnosed if the stone was refractory to disintegration or inadequate drainage was encountered. If incomplete clearance of fragmented stone occurred, the patient was considered to have had a partial response. The subsequent treatment of patients who failed to respond to ESWL was either PCNL or open surgery. Obstructed ureters were treated by ureteroscopy (URS). All results were recorded and analysed.

Results

During the 17-year study period a total of 63 patients with calculi in kidneys with congenital anomalies were treated in our stone clinic. Of the 63 patients, 21 had a stone burden less than 6 mm and were managed conservatively. Twenty five patients had a stone burden > 9 mm and were treated primarily with ESWL. Eight patients had renal function less than 20% and were managed conservatively or by nephrectomy if infection supervened or patients complained of pain. Prior to 1995, two patients were excluded from receiving ESWL because of coagulopathy. Seven patients presented with a large renal stone burden in kidneys with congenital anomalies and were subjected to primary open surgical intervention. The 25 patients who were subjected to ESWL, the subject of this study, were further analyzed regarding efficacy and outcome of ESWL.

Of the 25 patients (with 31 renal units) there were 18 men and 7 women with a mean age of 34 (range 3–64) years. Eleven (with 15 renal units) were managed by ESWL and EPL was performed in 14 patients (16 renal units). Although we did not perform a comparative study, our unit's experience was that the ESWL was more effective at stone disintegration in both normal and abnormal kidneys. Nine patients had horseshoe kidneys (Fig. 1), nine had different types of ectopic kidney (Fig. 2), three had malrotated kidneys (Fig. 3), two patients had duplex renal systems and one patient

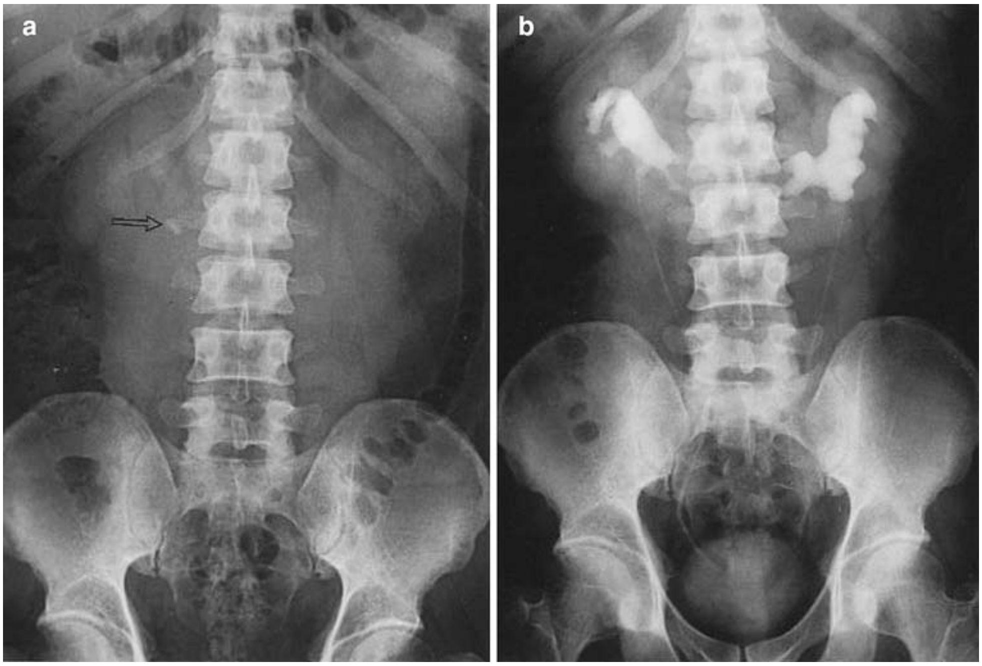

Fig. 1 Intravenous urography showing horseshoe kidneys with a calculus on the right side indicated by an *arrow*

each had polycystic and hypoplastic kidney. Table 1 shows the different locations of these stones and their distribution in different types of congenital renal anomalies. The mean size of stone was 1.5 (range 0.8–2.1) cm. Out of the 25 patients treated by lithotripsy, in 17 patients the stone sizes ranged between 9 and 15 mm and no 'J' stents were inserted prior to lithotripsy. A double 'J' stent was inserted prior to lithotripsy in 8 patients with stone size > 1.5 cm.

The average number of sessions per stone was 4.4 and 4.7 for ESWL and EPL respectively. The mean ISD number of shocks given was 1,727 ± 150 and 3,666 ± 111 for ESWL and EPL respectively. Out of 31 treated renal units, stone-free status was achieved in 24 renal units (77.4%). Partial response was achieved in two renal units (6.5%) as shown in Tables 1 and 2. In five renal units (16.1%) there was no stone clearance. Table 2 also shows that better clearance was achieved

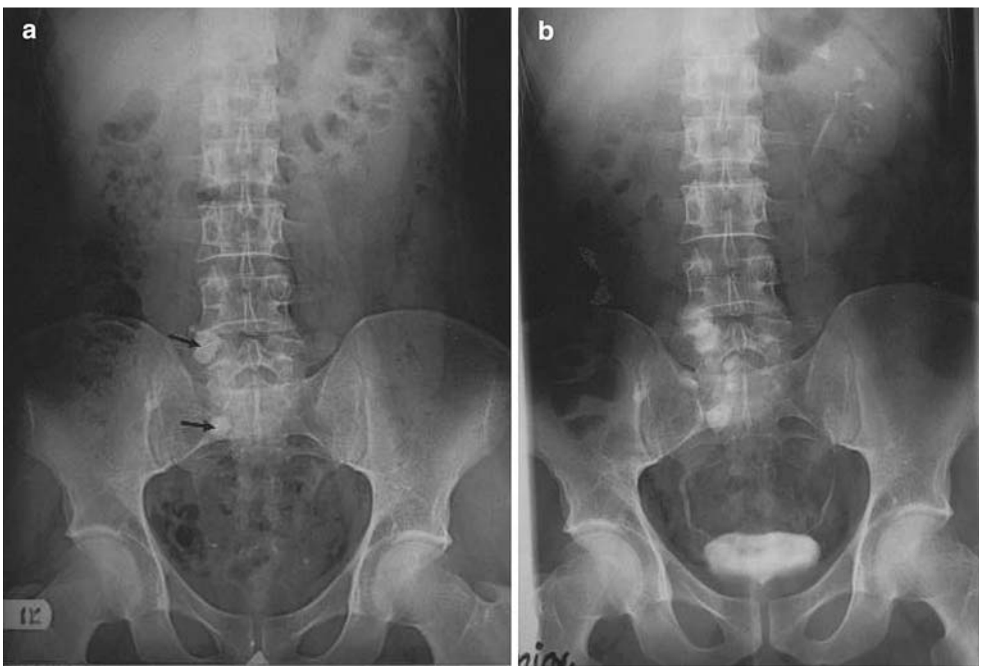

Fig. 2 Intravenous urography showing a right pelvic kidney with calculi indicated by *arrows*

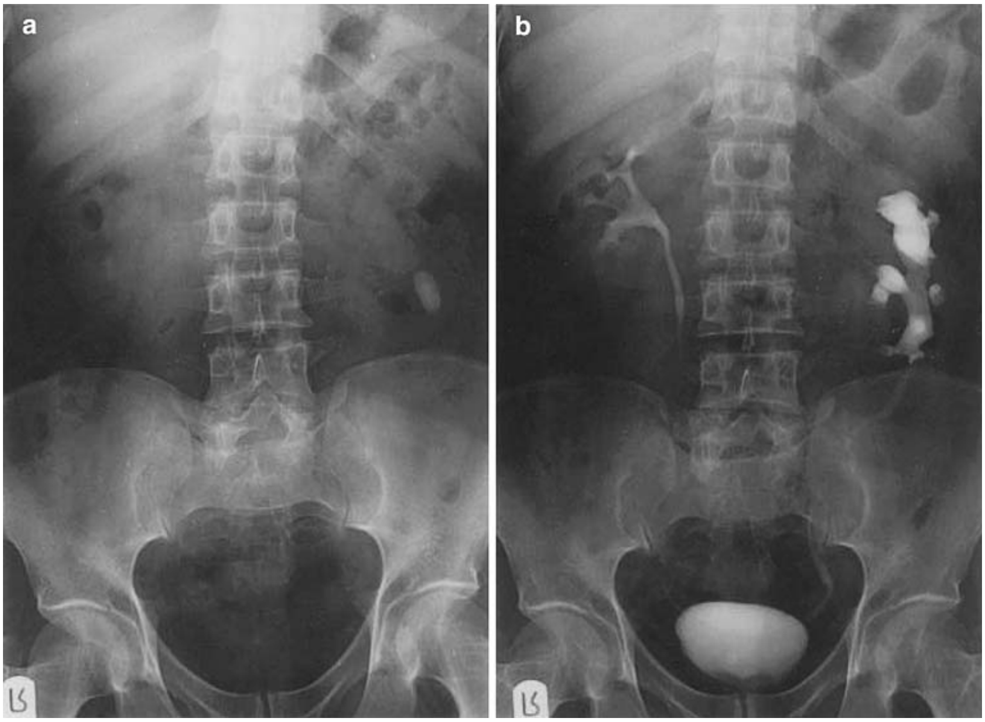

Fig. 3 Intravenous urography showing a malrotated left kidney with a calculus

Table 1 Type of congenital renal anomalies and outcome of treating the calculi using lithotripsy in 25 patients treated with ESWL

Type and number of patients with congenital renal anomaly	Number of renal units	Stone clearance status			% Stone free rate	Subsequent management for failed ESWL treatment
		Free	Partial	Failure		
Horseshoe kidneys (9)	13	8[a]	2	3	76.9	Open surgery × 2 PCNL × 1
Ectopic kidneys (9)	10	10	0	0	100	0
Malrotated kidneys (3)	4	2	0	2	50	Open surgery × 1 PCNL × 1
Duplex renal system (2)	2	2	0	0	100	0
Polycystic kidney (1)	1	1	0	0	100	0
Hypoplastic kidney (1)	1	1	0	0	100	0
Total (25)	31	24	2	5	83.9	5

[a]Four renal units required URS before becoming stone free

for calculi in the renal pelvis or upper pole. Out of 24 renal units considered stone-free, 4 required ureteroscopy and Dormia basket to evacuate fragments of calculi from the ureter. In five renal units that were considered failures, one patient had bilateral stones in a horseshoe kidney, one had unilateral stone in a horseshoe kidney and two renal units with malrotated pelvis also had one stone. The patient with a malrotated renal pelvis had a stone 2 cm in largest diameter; however, the shadow of the spine made localization difficult. Patients who failed ESWL were treated subsequently with different methods. There was open surgery on one side and PCNL on the other side in a patient with bilateral stones in a horseshoe kidney. In three other patients (three renal stones) open surgery was carried out in two patients and PCNL in one patient. All the patients were followed up as outpatients and none had any deterioration in renal function after a mean follow-up of 39 (range 3–204) months.

Table 2 Relationship between stone location and stone clearance using ESWL

Stone location	*N*	Stone clearance		
		Complete	Partial	Failure
Upper calyx	6	6	0	0
Lower calyx	10	6	2	2
Pelvis	15	12	0	3
Total	31	24	2	5

Positional modification was a key challenge in treating calculi in kidneys with congenital anomalies. To facilitate stone location four out of ten patients with calculi in pelvic or ectopic kidneys were put in a prone

Fig. 4 Patient with calculus in a pelvic kidney placed in a prone position to facilitate stone localization

or semi-prone position, as shown in Fig. 4. Three out of nine patients with horseshoe kidneys and one patient with crossed ectopia were put in lateral positions, before the stones could be localized prior to giving the shock waves as shown in Fig. 5.

Discussion

Common congenital renal anomalies include horseshoe kidneys, malrotated kidneys, renal ectopia, renal agenesis and polycystic kidneys [6]. Horseshoe, ectopic, malrotated and polycystic kidneys may be associated with obstruction, stasis and subsequent stone formation. The high insertion of the ureter, its anterior course over the isthmus, and the variation in the arterial and venous vasculature may singly or in combination contribute to stasis which may increase the chance of infection and/or calculus formation in patients with horseshoe kidneys [2, 5]. However, as Evans and Resnik [6] have noted, metabolic factors may also be involved. Prior to the 1980s, open surgery was the only modality of therapy to remove stones in abnormally placed kidneys with reconstruction of the pelvi-

Fig. 5 Patient with calculus in a horseshoe kidney placed in a lateral position to facilitate stone localization

ureteric junction, and with potential complications [7, 8]. In the early 1980s, with the introduction of PCNL in the management of stones, special precautions had to be taken in carrying out these procedures in abnormally located kidneys due to anatomic and vascular variations. Nowadays, PCNL does not carry any greater risk in these kidneys than that reported in normal kidneys [9].

The introduction of ESWL in the management of stones in horseshoe kidney and other renal anomalies has further reduced the potential complications associated with open surgery or PCNL [10, 11]. Localization of stone and drainage of disintegrated stones are major considerations to be borne in mind in dealing with abnormally placed kidneys. In this study, ESWL was performed either in supine, prone or lateral positions with some tilting using a pillow under the contralateral side as shown in Figs. 4 and 5. To facilitate drainage, a preoperative J stent was inserted in eight patients (nine renal units) with a low-lying pelvis. Out of nine patients with horseshoe kidneys (13 renal units), ten renal units were rendered free of stones (7 patients, 77.8%). The remaining three renal units required ureteroscopy as a complementary treatment to extract residual stone fragments. One patient had bilateral renal stones with significant residual stones that required PCNL in one kidney and open surgery in the other kidney. Open surgery was performed on one patient when both ESWL and PCNL had failed. Postural drainage and ureteral stenting might facilitate fragment passage. The abnormal anteriorly directed outflow of urine and calculus fragments over the fused renal parenchyma in horseshoe kidneys is believed to be a potential problem when these patients are treated with ESWL. However, in this study, follow-up data indicate that 10 of 13 (76.9%) horseshoe renal units were rendered stone-free. Our results are similar to those of Smith et al. [2], who reported a 79% stone-free rate in their series, and Bhatia et al. [9], who reported a stone-free rate of 94.4% for stones in horseshoe kidneys treated by ESWL. Malrotated kidneys have the same anatomical configuration as horseshoe kidneys and for the same reasons patients with malrotated kidneys are prone to renal calculi formation. Two renal units out of four were rendered stone-free (50%) and significant residual stones were encountered in the other two patients. Out of these latter two patients one required open surgery, while the other was treated successfully using PCNL. We achieved 100% stone-free rates in: ten stones in different types of renal ectopia (Fig. 2), two stones in duplex kidneys and one stone each in a polycystic and a hypoplastic kidney. This result is similar to that of Bhatia et al. [10], who reported that eight patients with stones in duplex system were rendered 100% stone-free at 3-month follow-up post-ESWL. Our overall stone clearance rate was 84% (26/31–83.9%). Therefore, clearance of stones as well as the use of auxiliary methods to accomplish this goal was comparable to the results of treating calculi in normal kidneys if the size of the stone is considered [11–16]. These data indicate that the stone-free rate is also influenced by the type of renal anomaly, being about 100% in patients with ectopic kidneys and duplex kidneys, compared to 50–76.9% for patients with horseshoe or malrotated kidneys as shown in Table 1.

The treatment of renal calculi in anomalous kidneys can also be achieved using PCNL, ureteroscopy or laparoscopy apart from ESWL [2, 9, 10, 17–21]. The stone clearance rate using PCNL in most centres is about 88% which, although similar to that achieved in calculus in horseshoe kidneys treated by ESWL (about 76–94%), is a far more challenging method of treatment with potentially more complications compared to ESWL [2, 9, 17]. Hence, for calculi in horseshoe kidneys, ESWL where feasible should be the first choice of treatment as its use as indicated by our results and those of others is associated with less morbidity, as it is also less invasive compared with PCNL [9, 17]. However, if the patient has a large stone burden in an anomalous kidney, to reduce the incidence of steinstrasse it can be debulked using PCNL followed by 'J' stent insertion and treatment of residual calculi using ESWL [17, 18]. Recently Weizer et al. [19] reported their experience on the use of the ureteroscopy with laser lithotripsy in the management of calculi in anomalous kidneys. In patients with horseshoe kidneys they reported a stone clearance rate of 75%, which is similar to the stone clearance rate obtained by us and others using ESWL [2, 9, 10]. The minimally invasive nature of ESWL compared to that of ureteroscopy would appear to favour the former over the latter. However, it is pertinent to point out that although similar stone-free rates can be achieved using ESWL, PCNL and ureteroscopy, the stone-free rate is achieved in one or two sessions using the latter two techniques whereas in ESWL, multiple sessions are often required. Furthermore, ESWL can have adverse long-term consequences for renal function and may result in hypertension [22]. Ureteroscopy and PCNL do not as a rule produce these long-term complications [22]. In patients where ESWL is relatively contraindicated such as in grossly obese patients, patients with deformed habitus or patients with coagulopathy, ureteroscopy and laser

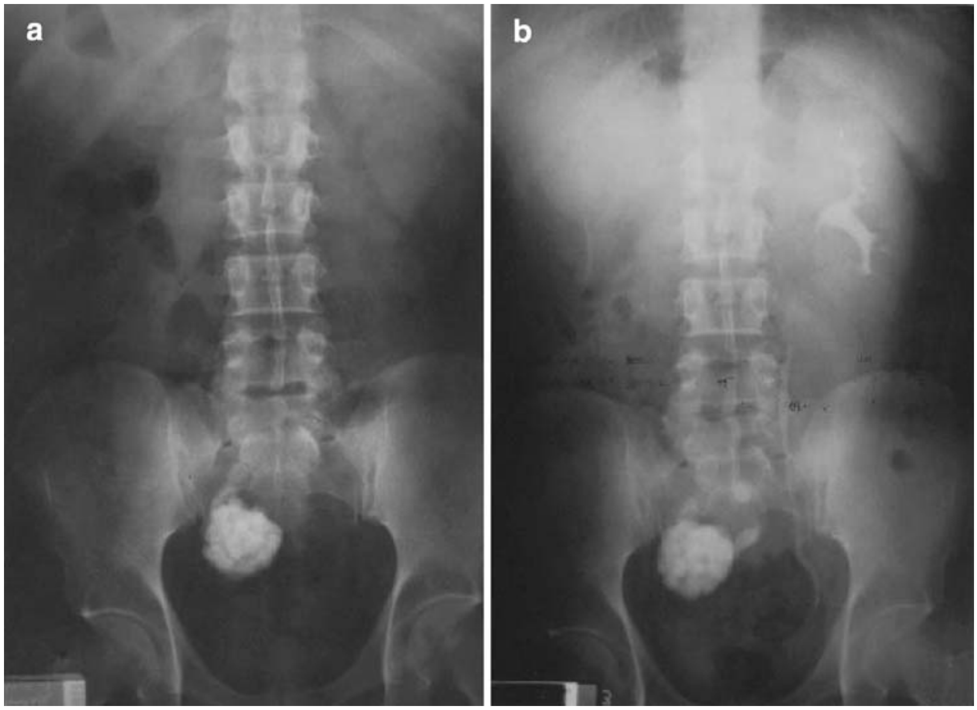

Fig. 6 Intravenous urography showing a right pelvic kidney with a large stone burden with right PUJ obstruction and 25% right renal function, treated successfully by one-stage open pyelolithotomy and Anderson–Hynes pyeloplasty

lithotripsy would appear to be the best treatment option [19]. Laparoscopy alone or in combination with PCNL may also be tried in some cases of calculus in anomalous kidneys before resorting to open surgery [20, 21].

From our experience, the most important factor affecting the efficacy of ESWL in the treatment of calculi in kidneys with congenital anomalies is the ease of positioning of the patient to deliver the shock waves to the calculi [14, 15]. If the vertebral column is in the way of the shockwaves as in some patients with horseshoe or malrotated kidneys, ESWL may not be effective in treating such patients. On the other hand, in all other anomalies of the kidneys like ectopic kidneys or kidneys with duplex system where the spine does not interfere with the direction of the shock waves, 100% stone clearance rate can be expected [14]. However, patients with horseshoe kidneys or malrotated kidneys may be likely candidates for open surgery ureteroscopy or PCNL if, after a trial period, ESWL is not effective for stone disintegration. Laparoscopy is increasingly being used for some of these cases in some units. Similarly, if a patient presents with a large stone burden in a kidney with a congenital anomaly with borderline ipsilateral renal function and if there is an additional reason for open surgical intervention, primary open surgery may still be the best treatment option even today. In such cases, before proceeding to open surgery, laparoscopy is worth a trial in these patients. Figure 6 illustrates one out of seven such cases that we managed by primary open method during the study period. The patient had a large stone burden in an ectopic kidney with 25% function and a PUJ obstruction. The kidney was ectopic and deep in the pelvis; an initial attempt at laparoscopy failed, due to difficult access. He was treated successfully by a one-stage open retroperitoneal pyelolithotomy and Anderson–Hynes pyeloplasty.

Conclusions

Extracorporeal shock wave lithotripsy should remain the first choice of treatment for calculi in kidneys with congenital anomalies because among all available options, the stone-free rate is similar to those achieved by PCNL or ureteroscopy. Its use is least invasive and associated with possibly the least immediate morbidity. However, multiple sessions are often required before patients become stone-free when using ESWL in these patients compared to 1–2 sessions when PCNL or ureteroscopy is used. Also, long-term complications like renal dysfunction and hypertension are sometimes encountered in patients treated by ESWL, while these complications are rarely seen in those treated by the other methods. Patients with horseshoe or malrotated kidneys require special positioning to ensure that the shock waves can be delivered effectively to the calculi. Surgical intervention is indicated in stones which are refractory to disintegration by lithotripsy.

References

1. Bauer SB (2002) Anomalies of upper urinary tract. In: Walsh PC, Retik AB, Vaughan ED Jr, Wein AJ (eds) Campbell's urology, 8th edn, vol 2, Chap 38. W.B. Saunders Co, Philadelphia, pp 1880–1913
2. Smith JE, Van Arsdalen KN, Hanno PM, Pollack HM (1989) Extracorporeal shock wave lithotripsy treatment of calculi in horseshoe kidneys. J Urol 142:683–686
3. Hoenig DM, Shalhav Al, Elbahnasy AM, McDougall EM, Clayman RV (1997) Laparoscopic pyelolithotomy in a pelvic kidney: a case report and review of the literature. J Laparoscopic Surg 1:163–165
4. Locke DR, Newman RC, Steinbock GS, Finlayson B (1990) Extracorporeal shock wave lithotripsy in horseshoe kidneys. Urology 35:407–411
5. Pitts WR, Muecke EC (1975) Horseshoe kidneys: a 40 year experience. J Urol 113:743–751
6. Evans WP, Resnick MI (1981) Horseshoe kidney and urolithiasis. J Urol 125:620–629
7. Jaetschek G, Kunzel KH (1988) Percutaneous nephrolithotomy in horseshoe kidneys :applied anatomy and clinical experience. Br J Urol 62:117–125
8. Culp OS, Winterringer JR (1955) Surgical treatment of horseshoe kidneys: comparison of results and various types of operations. J Urol 73:747–752
9. Bhatia V, Biyani CS (1992) ESWL for calculus disease in horseshoe kidney: our experiences and current status. Japanese J Endourol ESWL 5:1–10
10. Bhatia V, Biyani CS (1993) Calculus disease in duplex system—role of extracorporeal shockwave lithotripsy. Urol Int 50:164–173
11. Chaussy C (1988) ESWL: past, present and future. J Endourol 2:97–105
12. Holmes SAV, Whitfield HN (1991) The current status of lithotripsy. Br J Urol 68:337–344
13. Ng LG, Yip SK, Wong MY, Lau TN (2001) Clinics in diagnostic imaging. Hydronephrotic horseshoe kidneys with multiple calculi. Singapore Med J 42:540–544
14. Serra AC, Moreno RP, Baron FR, Garcia de Vicu FM et al Current management of calculi in horseshoe kidneys. Scand J Urol Nephrol 34(7):114–118
15. Slavkovio A, Vlajkovic M, Radovanovic M, Siric Z, Stefanovic V (2001) ESWL in the treatment of stone in children with horseshoe kidney. Int Urol Nephrol 32:327–329
16. Desai MR, Jasani A (2000) Percutaneous nephrolithotripsy in ectopic kidneys. J Endourol 14:289–295
17. Raj GV, Auge BK, Weizer AZ, Denstedt JD, Watterson JD, Beiko DT et al (2003) Percutaneous management of calculi within horse shoe kidneys. J Urol 170:48–51
18. Sheir KZ, Madbouly K, Elsobky E, Abdelkhalek M (2003) Extra corporeal shock wave lithotripsy in anomalous kidneys. 11-year experience with two second-generation lithotripters. Urology 62:10–15
19. Weizer AZ, Springhart WP, Ekeruo WO, Matlaga BR, Tan YH, Assismos DG et al (2005) Ureteroscopic management of renal calculi in anomalous kidneys. Urology 65:265–269
20. Holman E, Toth C (1998) Laparoscopically assisted percutaneous transperitoneal nephrolithotomy in pelvic dystopic kidneys: experience in 15 successful cases. J Laparoendosc Adv Surg Techn A8:431–435
21. Zafar FS, Lingerman JE (1996) Value of laparoscopy in the management of calculi complicating renal malformations. J Endourol 10:379–383
22. Krambeck AE, Gettman MT, Rohlinger AL, Lohse CM, Patterson DE, Segura JW (2006) Diabetes mellitus and hypertension associated with shock wave lithotripsy of renal and proximal ureteral stones at 19 years of follow up. J Urol 175:1742–1747