Urolithiasis in kidneys with abnormal lie, rotation or form

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Current Opinion in Urology 2011, 21:145–153

Purpose of review
The treatment options in urolithiasis in kidneys with abnormalities of form and location can be challenging due to abnormal anatomy (calyceal and renal orientation), relative immobility interfering with movement of equipment, and abnormal relation with other visceral organs.

Recent findings
In this review, we focus on the different techniques and results of various treatment modalities. The approach to managing these stones should be individualized. We also allude to the results of a few recent series and emphasize various treatment options.

Summary
Ultrasound helps in gaining access in ectopic kidneys, in addition to being a diagnostic tool. Computerized tomography is pivotal in helping to decide the management and choosing the method of treatment in anomalous kidneys. Flexible ureteroscopy can be a useful tool in stones less than 2 cm in size with the availability of smaller flexible ureteroscopes and access sheaths. However the surgeon should consider complete 'on table' clearance in these patients as the drainage is likely to be impaired. Ultrasound guided percutaneous approaches for ectopic kidneys should be performed by surgeons well versed with it. Laparoscopic assisted percutaneous nephrolithotomy has shown good clearance rates with minimal morbidity and less likelihood of ancillary procedures. Although adequate fragmentation can be achieved with extracorporeal shock wave lithotripsy, the drainage of fragments might be impaired due to the anatomical abnormalities. The choice of shock wave lithotripsy as a treatment option should be made prudently.

Keywords
ectopic kidney, horseshoe kidney, percutaneous nephrolithotomy

Curr Opin Urol 21:145–153
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0963-0643

Introduction

Urolithiasis has affected the human race for centuries and the options in managing stones range from open to minimally invasive surgery, which includes extracorporeal shock wave lithotripsy (ESWL), percutaneous nephrolithotomy (PCNL) and flexible ureterorenoscopy. The treatment options in urolithiasis with malrotated kidneys and ectopic kidneys can be challenging. The renal anomalies range from agenesis, dysplasia, fusion, rotation, and ectopia, to aberrations of the pelvicalyceal system [1•]. In this article, we review the management of urolithiasis in kidneys with abnormal lie, rotation or form. The salient points to note and be concerned with in these patients are abnormal anatomy (calyceal and renal orientation), relative immobility interfering with movement of equipment, abnormal relation to other visceral organs and abnormal vasculature [1•].

We focus on technical issues and results in various approaches in managing these patients.

Imaging

The key imaging modalities to be employed include computerized axial tomography (CT) and ultrasound scan of the abdomen. An ultrasound scan should ideally be done by the operating surgeon as it may help the surgeon to decide the method of access, for instance, if a good acoustic window is seen while scanning an ectopic pelvic kidney, it suggests that a ultrasound guided puncture is possible. There are concerns regarding the relationship of anomalous and ectopic kidneys with the surrounding structures and organs. There have been suggestions to include CT scan as a routine investigation to avoid injury to these structures [2]. Horseshoe kidneys have a higher incidence of retrorenal colon, due to a defect in the normal development of lateral conal fascia resulting in deficiency of mechanical support to the colon [1•]. Maheshwari *et al.* [3] have shown that contrast enhanced CT in a horseshoe kidney showed major vessels entering the kidney along the posterior surface. It also revealed an intrarenal location of the pelvis and a

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stone was in awkward intrarenal location. These findings helped the surgeon to decide laparoscopy as the preferred modality for gaining access. This case highlights the fact that CT can help in planning punctures in these anomalous kidneys. Additionally a preoperative CT has the advantage of predicting the outcome of ESWL treatment as shown by the effectivity quotient in various series [4•]. Ectopic kidneys pose a problem for any planned intervention due to their anomalous blood supply. So these kidneys are prone to iatrogenic injuries because of its varied vasculature and anatomic relationships. These complications can be avoided or reduced by a preoperative CT planning with angiography [5,6•].

Ectopic kidney and fused kidney

Percutaneous renal access can be gained either by guidance of ultrasound, laparoscopic vision, or fluoroscopy or various combinations of these modalities.

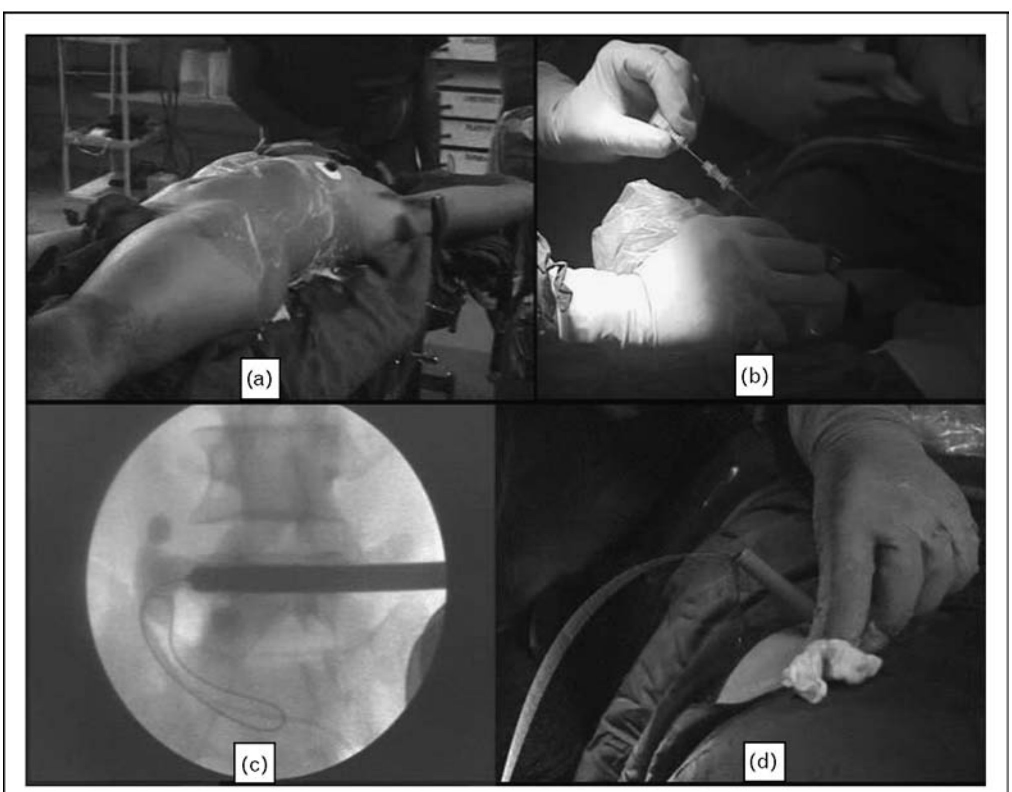
Ultrasound guidance

The procedure is usually done under general anaesthesia. After retrograde catherization, the pelvicalyceal system is distended and opacified. The patient is placed in a supine oblique position with a bolster under the ipsilateral hemipelvis. Pressure on the ultrasound probe is used to displace the bowel away from the puncture line. The dilatation is performed with telescopic dilators (Fig. 1). A contrast study is done at the end of the procedure. In a study by Desai and Jasani [7] none of the patients had any bowel injury. All patients had complete stone clearance on follow-up. The average haemoglobin drop was 0.9/dl. The average hospital stay

Key points

- CT is pivotal in deciding the management and choosing the method of treatment in anomalous kidney.
- Laparoscopic assisted PCNL has shown good clearance rates with minimal morbidity and less likelihood of ancillary procedures.
- Although adequate fragmentation can be achieved with shock wave lithotripsy (SWL), the drainage of fragments might be impaired due to the anatomical abnormalities.

Figure 1 Technique of percutaneous nephrolithotomy in ectopic pelvic kidney with ultrasound guidance



(a) Supine position with bolster under the ipsilateral hip. (b) Pressure on the ultrasound probe pushes the bowel away. (c) Tract dilatation is done with telescopic dilators under fluoroscopic guidance. (d) Amplatz sheath helps to maintain the low pressure in the pelvicalyceal system.

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was 5.2 days. In an update on this series, in 16 cases there was one instance of bowel injury; however, the authors had started tackling more complex stones (12 simple and four complex) [8].

It may be noteworthy that apart from these authors, no other series has demonstrated similar results, suggesting that this technique should only be employed by those expertly versed in ultrasound guided percutaneous access.

Ultrasound guided percutaneous nephrolithotomy in a fused L-shaped kidney

A 68-year-old lady presented with bilateral staghorn calculi in a left to right crossed L-shaped renal fusion ectopia anomaly. In a supine position with a slight tilt on the right side, ultrasound was used to gain access to the middle calyx of the right kidney. The majority of the staghorn stone bulk was thus cleared with further separate punctures and access tracts for the lower and upper calyces using an extra long nephroscope (Fig. 2).

Laparoscopic guidance for access

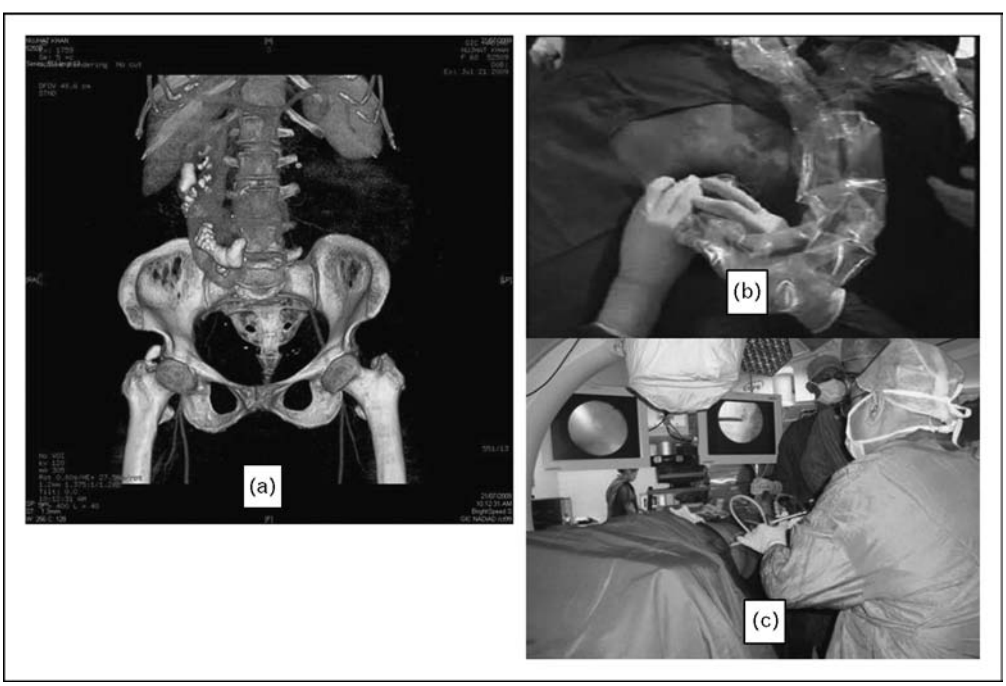
This approach for management of stones in an ectopic kidney gained acceptance after the interest generated by the first report from Eshghi *et al.* [9] for a similar case. The procedure is performed in a Trendelenburg position after placing a ureteric catheter. Once pneumoperitoneum is established, a 10-mm port is placed in the umbilicus, two 5-mm ports are placed, one in the mid-clavicular line at the level of the umbilicus and one in the iliac fossa. The puncture needle is introduced under laparoscopic and fluoroscopic guidance, once clear fluid is aspirated, the rest of the procedure is performed as in standard PCNL [10•].

Goel *et al.* [11] described two different approaches to this technique: in one of the patients with a prior history of open pyelolithotomy, the anterior surface of the kidney was exposed by mobilizing the overlying sigmoid colon laparoscopically and the percutaneous tract was established into the desired calyx under combined laparoscopic and fluoroscopic control. In the second patient, the tract was established between the major mesenteric vessels without any mobilization of the bowel.

It is a well known fact that kidneys with anomalous form which include horseshoe kidney have a varied vascular anatomy [12]. The stones in an ectopic kidney present a unique challenge to the operating surgeon and the method of access should be selected judiciously by the surgeon. A variety of approaches have been described for such cases. A retroperitoneoscopic approach has been described by Troxel *et al.* [13] for ectopic kidneys. A drawback of this approach is that the retropubic space, which is often transgressed in multiple surgeries, may need to be utilized. Although beneficial it may make the case challenging.

Watterson *et al.* [14] has reported PCNL performed through the greater sciatic foramen. Prior to the

Figure 2 Clearance of staghorn stone bulk with extra long nephroscope in a L-shaped kidney



(a) Computed tomography scan helps to delineate the relation with the surrounding structures. (b, c) Ultrasound is used for gaining access.

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procedure, a flexible ureteroscopy was attempted; however due to the ureteral angulation it was not successful. A CT did not show any intervening bowel or aberrant vessel. The kidney was punctured through the greater sciatic foramen under fluoroscopy and dilated to 30 Fr. Postoperative CT did not reveal any extravasation. The authors noted that the foramen was bound by the sacrum, ischium and the iliac bones. The tract should be as close to the sacrum as possible to avoid vascular and neural structures. The nephrostomy tract existed in the mid-buttock. From a technical standpoint, the transgluteal distance is more than the distance to the kidney in flank position, and a full-length amplatz sheath (18 cm) was required. Thus the authors suggested that this approach may not be suitable in obese patients.

Matalaga *et al.* [15] have described a transhepatic PCNL in a patient who had multiple previous interventions, this patient also had hepatomegaly with the caudate lobe extending over the ectopic kidney. The tract was dilated till 30 Fr and the stones were removed with a combination of rigid and flexible nephroscopes. The authors in this case did not report any complications. In the same article, they described a case of transiliac PCNL for ectopic kidney in a patient of menigocele. The patient had undergone an ileostomy as a part of 'Sharrads' procedure for hip dislocation. During the procedure a round osteotomy was created in the iliac bone through which the psoas was transposed. A CT showed that the calculi were adjacent to the opening in the left iliac bone with no interposed viscera. An 18-gauge needle was passed under biplanar fluoroscopy through the targeted calyx into the left kidney. The tract was dilated till 30 Fr and stone removal performed. A few remaining fragments required additional flexible nephroscopy and two additional accesses. The authors noted that there are a few patients who are not good candidates for laparoscopic assisted PCNL, such as those with past history of abdominal surgery and those with difficult body habitus. Results are shown in Table 1 [16–20,21•,22–24].

The advantage of this approach is that each step is done under vision hence minimizing the chance of bowel and vascular trauma. The disadvantages of this approach are the need for skills and equipment in both laparoscopy and endourology; in addition, this approach does not guarantee a perfect calyceal puncture.

Horseshoe kidney

Stones in horseshoe kidneys can be tackled by PCNL, SWL, and/or flexible ureterorenoscopy.

Percutaneous nephrolithotomy

The access to the upper pole in kidneys placed in a normal position (orthotopic) requires a supracostal puncture, with its attendant complications. However it is worth noting that as these kidneys are low, placed upper calyx puncture in these cases does not require a supracostal puncture [18,20]. Raj *et al.* [18] noted that longer nephroscopes were required in such an access to clear all the stone bulk in these kidneys. In a series by Symons *et al.* [20] 17% of patients required more than one puncture. At our centre we prefer to employ multiple tracts and use flexible nephroscopy for stones in inaccessible calyces. However, there might be a trade-off in between using longer nephroscopes or flexible nephroscopes and employing multiple punctures for clearing stones. Results are shown in Table 1.

Role of laparoscopy

The laparoscopic approach for destroying stones in horseshoe kidneys is described either for pyelolithotomy or guiding percutaneous renal access (Fig. 3) [3,25]. Maheshwari *et al.* [3] have described a case wherein a preoperative CT scan was suggestive of an unfavorable vascular and pelvicalyceal anatomy for routine renal access, and the patient was offered laparoscopic guided access. The stone was in the dilated isthmic calyx of the kidney. Thus, a routine laparoscopy ran the risk of vascular and bowel injury. A laparoscopic guided approach was employed. Once pneumoperitoneum was established the bowel was reflected out of harms way. The puncture needle was passed through the skin incision just below the umbilicus. The tract was dilated with telescopic dilators.

Role of shock wave lithotripsy and flexible ureterorenoscopy in anomalous kidneys

SWL and flexible ureterorenoscopy pose unique technical challenges due to anatomical considerations in these kidneys.

Shock wave lithotripsy

Anatomic anomalies in horseshoe kidney are responsible for certain limitations in treating stones with SWL in these kidneys. The inherent problems are as follows [26,27]:

(1) The anterior position of the pelvis in horseshoe kidneys precludes the precise localization of the stone at F2. This may lead to inadequate fragmentation.
(2) Stones in medial calyces can be obscured by the spine.
(3) It may be difficult to localize the stone in prone position and hence supine position might be useful.
(4) Collecting system anomalies such as incomplete rotation, posterior location of the pelvis, fused lower poles, high insertion of the ureter, variation in vascular anatomy hinder drainage and promote stasis,

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Table 1 Results for various treatment modalities

Author	Technique and number of patients	Stone size and location	Stone-free rate	Remarks
PCNL and pyelolithotomy in ectopic kidneys				
Hoenig *et al.* [16]	Laparoscopic assisted pyelolithotomy ($n=0$)	Renal pelvic stone	100%	No complications
Eshghi *et al.* [9]	Laparoscopic assisted PCNL ($n=1$)	Staghorn	100%	No complications
Maheshwari *et al.* [3]	Laparoscopic assisted PCNL ($n=3$)	3.5, 5.5 cm and multiple stones	100%	No complications
Goel *et al.* [11]	Laparoscopic assisted PCNL ($n=2$)	600 mm^2	100%	No complications
Desai and Jasani [7]	Ultrasound guided ($n=9$)		100%	No complications
PCNL in horseshoe kidneys				
Jones *et al.* [17]	15	ND	77.8%	Two required SWL
Raj *et al.* [18]	24	448 mm^2 (63–1800)	87.5%	Two required SWL and one instance each of ureterocolonic fistula and pneumothorax, three of obstruction
Shokeir *et al.* [19]	34	664 mm^2 (264–2408)	73.5%	Eight required ESWL, one URS, one each of septaecemia, obstruction and colonic injury
Symons *et al.* [20]	47	614 mm^2	88%	Minor 15% (pyrexia), bleeding requiring clot evacuation
Liatsikos *et al.* [21•]	15 patients	17 staghorn calculi	82%	Major complications −20%, minor complications −46.6%
ESWL in anomalous kidneys				
Tunc *et al.* [22]	45 horseshoe kidneys,14 pelvic kidneys and 4 crossed fused ectopia) (Siemens lithostar)	ND	66.7% (horseshoe) 57.2% (pelvic), 25% (crossed fused ectopia), 56.7% for malrotated kidneys	ND
Lampel *et al.* [23]	37 horseshoe kidney (Dornier HM3)	14 mm	76%	Nine required ancillary procedures
Symons *et al.* [20]	6 renal units (Dornier compact delta)	50–225 mm (mean 149 mm^2)	Four successfully fragmented	One required two sessions and one was incompletely fragmented even after two sessions
Shier *et al.* [24]	49 horseshoe kidneys, malrotated in 120, duplex in 29 ((Dornier MFL 5000 and Toshiba Echolith)	13.54 mm	Overall stone-free rate −72.2%	Steinstrasse in seven patients

ND, no data; PCNL, percutaneous nephrolithotomy.

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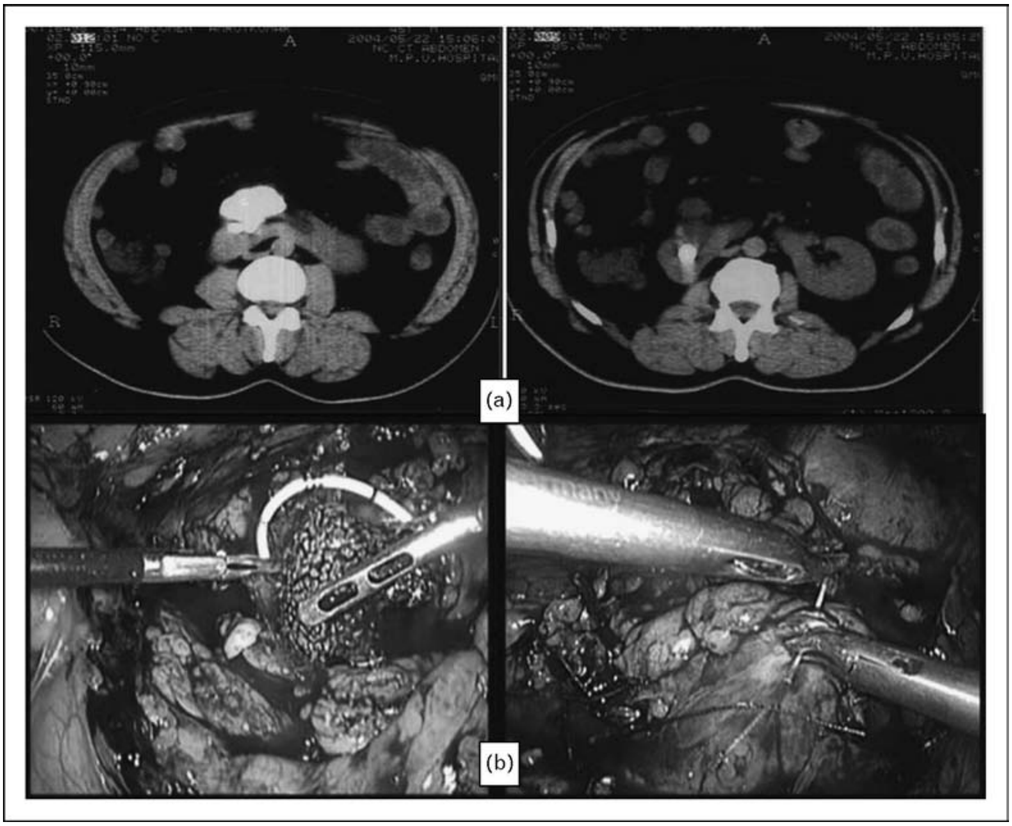
Figure 3 Complete clearance of stones in the isthmus of a horseshoe kidney using laparoscopic pyelolitomy



(a) Computed tomography scan shows that the calculus is lying at the isthmus of the horseshoe kidney. (b) Laparoscopic pyelolithotomy helps in complete clearance of stones.

and hence decrease the chance of success after the procedure.

The suggested modifications for improving the results in these stones include placing a double-J stent and delivering the shocks at a higher power. The position in which the shocks are to be delivered can also be altered (supine or prone depending on the localization of the stone). The results of SWL are as shown in Table 1.

Flexible ureterorenoscopy

Flexible ureterorenoscopy is a valuable option in cases with horseshoe kidney. It has the potential to allow laser fragmentation of stones into dust that may otherwise be unable to pass spontaneously due to aberrant anatomy.

Despite excellent results of flexible ureterorenoscopy in orthotopic kidneys, the literature is not abundant with series showing the effectiveness of this modality in managing renal calculi in anomalous kidneys. The overall stone-free rate in a study by Weizer *et al.* [28] has been 75% in both ectopic and pelvic kidneys. They treated four patients with horseshoe kidney and four patients with pelvic ectopic kidney. The average stone burden was 1.4 cm, a 7.5 Fr flexible ureterorenoscope was used in all cases with holmium laser lithotripsy and nitinol basket graspers. In a report by Fayad, he treated four patients with retrograde holmium laser lithotripsy. All had a more than 3 cm stone in the renal pelvis of the ectopic kidney. Three patients were rendered stone-free at 3 months follow-up. One patient required open surgery for complete clearance. The laser was kept at 1–1.5 J power with a frequency ranging from 15 to 20 Hz. There was no major complication. Symons *et al.* treated two patients in such kidneys with flexible ureterorenoscopy. One patient had a single pelvic stone (63 mm^2), whereas the other had four stones (9281 mm^2). Stone fragmentation was complete in all [20]. Andreoni *et al.* [29] reported a case wherein an access sheath had been used for fragmentation and removal of stones in a horseshoe kidney.

(1) The key issues in managing and selecting flexible ureterorenoscopy as a treatment modality are flexible ureterorenoscopy in a few series has been shown to be effective in stones less than 20 mm.
(2) Miniature scopes, nitinol baskets and other accessories, have a potential to improve the results in the future.

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(3) All attempts should be made to extract all the fragments in these patients, as the drainage is compromised in these kidneys.

Percutaneous nephrolithotomy in spinal deformity and musculoskeletal disorders

Shrivasatv *et al.* [6•] reported their experience with PCNL in musculoskeletal deformity and ectopic kidney. The mean stone size was 27.4 mm (16–37 mm). Six patients had severe kyphoscoliosis, two had achondroplasia. The stone-free rate in this study was 92.3%. The authors stressed that PCNL in these patients required preoperative planning with CT and the access should be image guided. Laparoscopic guided PCNL can be performed in these patients when access to the collecting system is not possible with image guidance.

In our opinion, the points to be taken in consideration in these patients are as follows (Figs 4 and 5):

(1) Preoperative CT helps to evaluate the presence of vascular and other visceral structures.
(2) Adequate precautions should be taken for padding the pressure points.
(3) Supine PCNL should be considered to prevent fractures and neurological postoperative problems.
(4) Ultrasound-guided PCNL helps in preventing bowel injury and injury to adjacent organs in expert hands.
(5) Judicious use of flexible instrumentation for complete stone clearance in a single sitting.
(6) Such cases should be operated on at centers with a significant amount of expertise in percutaneous, laparoscopic and flexible ureterorenoscopic surgery.

Metabolic basis of stone formation in anomalous kidneys

The question of stone formation in anomalous kidneys is unresolved. Anatomic causes such as high insertion of ureter, and ureteropelvic junction obstruction are thought to be the cause due to resultant stasis, obstruction and infection. A study by Raj *et al.* [30] showed that patients had metabolic abnormalities predisposing to stone formation. It was noted that hypovolemia, hypercalcemia

Figure 4 Preoperative evaluation in a patient with kyphoscoliosis and osteogenesis imperfecta undergoing percutaneous nephrolithotomy

(a) Plain radiograph kidney ureter and bladder (KUB) showing left renal calculi in a patient with kyphoscoliosis and osteogenesis imperfecta. (b) Ultrasound shows a good acoustic window. (c, d) Computed tomography scan helps in delineating the surrounding anatomy.

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Figure 5 Positioning and intraoperative steps in a patient with kyphoscoliosis and osteogenesis imperfecta undergoing percutaneous nephrolithotomy

(a) Severe kyphoscoliosis. (b) Supine position for percutaneous nephrolithotomy. (c) Amplatz sheath in position. (d) Complete clearance.

and hypocitraturia are the most common metabolic defects. The authors also suggested that patients with calculi in these kidneys should be candidates for metabolic evaluation in order to initiate preventive measures.

Conclusion

The approach to managing these stones should be individualized. The factors to be taken into consideration are the stone bulk, the location of the stone, the vascular anatomy and the anatomy of the pelvicalyceal system. Ultrasound helps in gaining access in ectopic kidney apart from being a diagnostic tool. CT is pivotal in deciding the management and choosing the method of treatment in anomalous kidneys.

CT will also give the attenuation values and be a deciding factor in deciding for SWL or flexible ureterorenoscopy. Flexible ureterorenoscopy will be a useful tool in small stone size burdens with the availability of smaller flexible ureterorenoscopes and access sheaths. However, the surgeon should consider complete 'on table' clearance in these patients as the drainage is likely to be impaired. Ultrasound scan guided approach for ectopic kidneys should be done by surgeons well versed with it. Laparoscopic assisted PCNL has shown good clearance rates with minimal morbidity and less likelihood of ancillary procedures. Although adequate fragmentation can be achieved with shock wave lithotripsy (SWL), the drainage of fragments might be impaired due to the anatomical abnormalities. The choice of SWL as a treatment option should be made prudently.

Acknowledgement

There are no conflicts of interest.

References and recommended reading

Papers of particular interest, published within the annual period of review, have been highlighted as:
- • of special interest
- •• of outstanding interest

Additional references related to this topic can also be found in the Current World Literature section in this issue (p. 170).

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