

Percutaneous Nephrolithotomy In Patients With Polycystic Kidneys.

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ABSTRACT

Introduction: Urolithiasis is known to affect 20-28% of patients with Autosomal Dominant Polycystic Kidney Disease, of whom 50% are symptomatic and 20% require definite urologic intervention. The management of urolithiasis includes oral dissolution therapy, extracorporeal shock wave lithotripsy and surgical treatment. Percutaneous nephrolithotomy (PCNL) has been reported as a method of stone treatment by several experts. In this study we report our experience with PCNL in patients with autosomal dominant polycystic kidney disease (ADPKD) and the outcome of the same.

Materials & Methods: We retrospectively reviewed the outpatient, inpatient records and imaging of all patients with autosomal dominant polycystic kidney disease undergoing PCNL at our centre.

Results: During the study period, a total of 17 patients with a mean age of 47 ± 5.6 years were diagnosed to have renal calculi with ADPKD and underwent PCNL for the same. The mean serum creatinine was 1.8 ± 0.4 mg% and mean

size of stones was 18 ± 4.6 mm. No major intra-operative complications were noted. Stone clearance rate with PCNL alone was 94.1%.

Conclusions: The PCNL in ADPKD is safe and effective but can be associated with postoperative bleeding requiring transfusions. Infection of the cysts may be a cause for fever, and prolonged leak.

Keywords: Percutaneous Nephrolithotomy, Urolithiasis, Autosomal Dominant Polycystic Kidney Disease.

INTRODUCTION

Autosomal dominant polycystic kidney disease (ADPKD) is the most common form of renal cystic disease, with an incidence of approximately 1 in 400 to 1000 live births. [1] It accounts for 8–10% of end stage renal disease. Urolithiasis is known to occur in 20–28% of patients, of whom 50% are symptomatic for stone disease and 20% require definite urologic intervention. [2, 3] Both anatomic and metabolic factors are known to be the causes of stone formation in ADPKD. The management of urolithiasis includes oral dissolution therapy, extracorporeal shock wave lithotripsy (ESWL) and surgical treatment. [2-4] Over the past few decades open surgery has given way to minimally invasive procedures such as ESWL (Extracorporeal shockwave lithotripsy) and percutaneous nephrolithotomy (PCNL). ESWL is a safe and effective method of treatment for stones less than 15mm in size. [5-7] PCNL has established itself as a method of stone treatment for large stone bulk in anatomically normal appearing kidneys. Vishwajeet Singh et al [8] reported their experience with PCNL in 22 patients (26 renal units) with ADPKD. The overall success rate of PCNL was 82.1% and PCNL with extracorporeal shock wave lithotripsy for clinically significant residual fragments was 92.85% respectively. Significant haematuria requiring blood transfusion was noted in 9 patient, postoperative fever due to cyst infection in 4 and paralytic ileus in 3. The authors concluded that PCNL in patients with ADPKD was safe and effective. In this paper we report our experience with PCNL in patients with ADPKD.

MATERIALS & METHODS

We retrospectively reviewed the outpatient, inpatient records and imaging of all patients with autosomal dominant polycystic kidney disease undergoing PCNL at our centre. This

study was permitted by the Institutional/University ethical committee. Patient demographics such as age at presentation, gender, symptoms, imaging, size of stones, preoperative blood biochemistry was noted. Pre-operative coagulation profile was noted. The type of anaesthesia administered, intraoperative complications, blood transfusions, post-operative outcome and complications were also noted. Adjuvant procedures such as post-operative ESWL, repeat PCNL, DJ stenting and other interventions were also noted.

RESULTS

During the study period Jan 2000 - Dec 2019, a total of 17 patients (15 males, 2 females) with a mean age of 47 ± 5.6 years (range 39 – 54) were diagnosed to have renal calculi with ADPKD and underwent PCNL for the same. Haematuria remained a most common symptom. Other symptoms included pain, fever, lumbar tenderness and dyspepsia. All the 17 patients were known hypertensives on treatment. Twelve (70.6%) of these were diagnosed to have polycystic kidneys prior to diagnosis of renal calculi. The mean serum creatinine was 1.8 ± 0.4 mg% (range 1.6 – 3.3), all the patients had proteinuria and urine culture showed *Escherichia coli* organism in three (17.6%) of the patients. Renal ultrasonography revealed bilateral polycystic kidneys. Plain CT revealed bilateral calculi in four (23.5%) patients and unilateral calculi in the remaining patients (Figure 1). The mean size of stones was 18 ± 4.6 mm (range 13-31). In view of raised renal parameters it was decided to perform PCNL in all the patients. Blood grouping and cross matching was done in all. None of the patients needed pre-operative transfusions. Bleeding time, clotting time were within normal range in all, however INR (International Normalized Ratio) was elevated in six (35.2%) of the patients, but not severe enough to prevent or postpone surgery. PCNL was performed using either an 18 Fr. or 26 Fr. Nephroscope, under fluoroscopic control (Figure 2). It was possible to retrieve the stone with a single puncture in 14 (82.4%) patients and with two punctures in the remaining three (17.6%). The mean duration of surgery was 95 (range 85-120) mins. Stone clearance rate with PCNL alone was 94.1%. No major intra-operative complications were noted. Three (17.6%) patients needed post-operative transfusions. The mean drop in haemoglobin was 1.6 ± 0.6 (0.5-2.8) gm%. Post-operatively ten (58.8%) patients had reddish urine for four days (Table 1). No severe haematuria was noted. Serum creatinine was raised post-operatively in 12 (70.6%) patients, which came down to over a period of two weeks. Following removal of nephrostomy, prolonged leak was noted in five (29.4%) patients, which stopped on its own in a week's time. Only one (5.9%) patient needed an adjuvant ESWL session to treat a residual fragment. The double J stent kept after PCNL was removed 2-3 weeks following the procedure.

Figure 1.

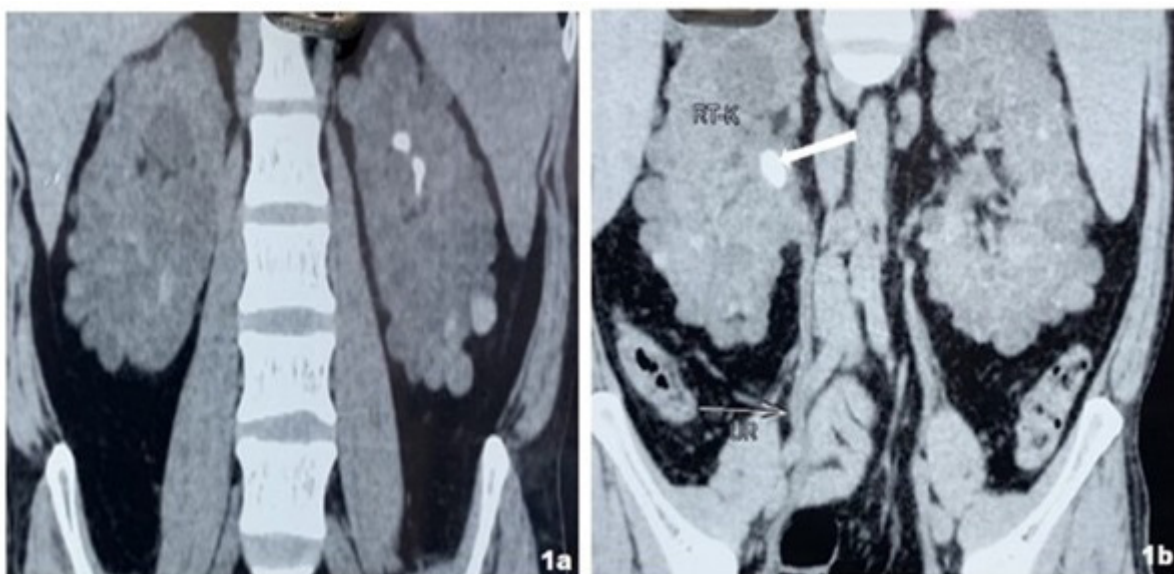


Figure 1a: Non-contrast computed tomography of KUB region shows both the kidney are enlarged in size, the renal parenchyma is replaced by multiple cysts.

Figure 1b: Right kidney shows a pelvic calculus (white arrow) approximately measuring 0.7 x 2.2 x 1.4 cms. (Avg. HU +1460) causing moderate hydronephrosis.

Figure 2.

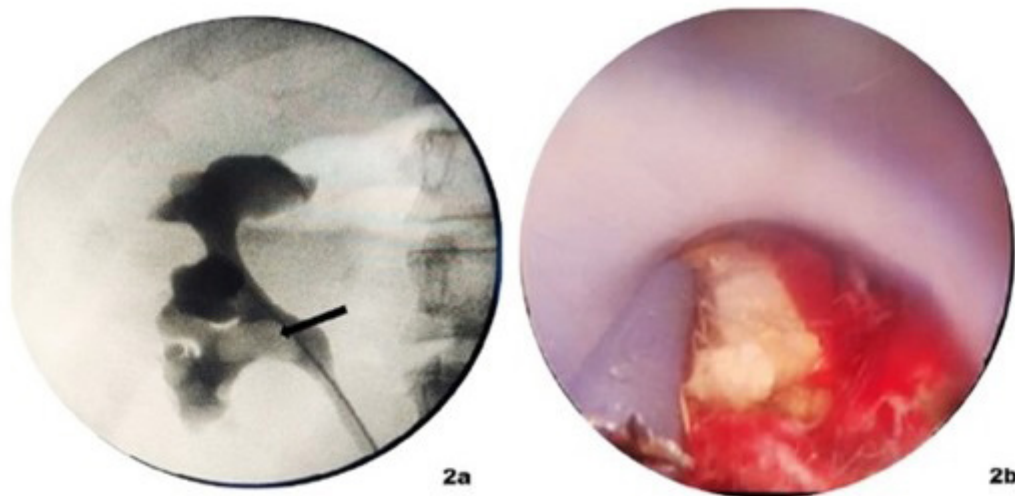


Figure 2a: Intra-operative fluoroscopy image shows a retrograde pyelogram depicting pelvicalyceal anatomy and stone (black arrow) in the renal pelvis seen as a filling defect

Figure 2b: Endoscopic view of percutaneous nephrolithotomy shows stone being fragmented with ballistic (pneumatic) lithotripsy.

Table 1: Complications classified according to modified Clavien system.

No	Complications	n (%)
1	Grade 1 Prolonged urinary leak from PCN site	5
2	Grade 2 Bleeding needing transfusions	3
3	Grade 3a	-
4	Grade 3b	-
5	Grade 4a Raised renal parameters needing ICU care	1
	Total	9

DISCUSSION

Patients with ADPKD are prone to develop urinary stones. Urolithiasis is known to aggravate renal dysfunction and damage thereby accelerating renal failure in these patients. [9] Calcium oxalate and uric acid are the common components of the renal stone in these patients. The expanding renal cysts distort the intrarenal calyceal anatomy thereby causing urinary stasis. This facilitates the formation of crystals and aggregation of these leads to formation of calculi. It has been noted that the levels of urinary oxalate and the process of urinary crystallization is significantly higher in patients with ADPKD and urolithiasis. Nikolov et al [10] retrospectively studied patients with ADPKD and reported a high proportion of uric acid stones. Composition of the stone and low urine pH, suggested that metabolic, along with mechanical factors were responsible for the occurrence of urolithiasis in these patients. The renal calculi in patients with ADPKD can be diagnosed using ultrasonography (USG) and/or computed tomography (CT). Studies have shown that renal USG may under detect urolithiasis in patients with ADPKD because of the frequent occurrence of renal calcifications. [7] Unenhanced helical CT provides an excellent option to distinguish renal calculi from that of calcifications within the cysts. [11] Nishiura et al [7] looked for the presence of renal stones in patients with ADPKD using both renal ultrasound and unenhanced helical CT scan in 125 patients. CT scan detected calculi in 32 patients, including 20 whose previous ultrasonography had revealed no calculi.

Percutaneous nephrolithotomy and ESWL remain the mainstay of treatment in these patients. ESWL has been associated with poor results in patients with ADPKD with stone-free rates ranging between 25–46% at 3 months. [12, 13] PCNL has been the procedure of choice for large renal calculi and so it would be to treat the stones in patients with ADPKD. PCNL has been commonly performed under fluoroscopic guidance in patients with ADPKD, however some have used ultrasonography guidance so as to avoid the cysts. [14] PCNL is difficult in these patients as the calyceal spaces are elongated by the compressive effect of the cysts. Use of ultrasound guidance may make it difficult to aim at the calyces because of the multiple cysts. Use of methylene blue along with contrast agent would be a good method to confirm the puncture. [15] Continuous efflux of blue

liquid would be the definite proof that the desired calyx has been punctured. Some have used ultrasound contrast agent to puncture the right calyx however the use of ultrasound contrast agent seems to be not as good as it is claimed to be. [14] Whenever it becomes difficult, it is better to direct the puncture at the stone directly. Baishya et al [2] reported on the available options for the management of renal stones in 19 patients with ADPKD. Mean serum creatinine was 7.2 mg/ dl (range 0.8-18.1 mg/dl) at presentation. The mean stone size was 115 mm² (range 36 to 980 mm²). Ten Renal units (nine patients) required intervention, of which percutaneous nephrolithotomy was performed in three. PCNL had complete clearance (**Table 2**).

Table 2: Comparison with other series of PCNL in patients with ADPKD.

Parameters	Our Study	Baishya et al. [2]	Singh et al. [8]	Srivastava et al. [16]
No of Patients	17	19	22	22
Renal units	17	23	26	25
Age (mean)	47.5	43.3	38	39.7
Serum Creatinine mg/dL	1.8	7.2	-	-
Puncture	Single 14, Multiple 3	PCNL- 3 patients USG-2 Fluoroscopy-1	Single stage-12, Two stage-10	Single 20, multiple 5
Stone clearance	Complete	Complete	-	
Success rate	PCNL alone 94.1%	PCNL alone 100%	PCNL alone 82.1%	Single stage 88%, Second stage 100%
Hb drop (gm/dl)	1.6	0.5-3.2	-	-
Blood transfusion	3 (17.6%)	none	9 (32.2%)	3
Other complications	Prolonged leak	none	Fever 16%, haematoma 16%, hydrothorax 16%	Fever 16%

Srivastava et al [16] studied the efficacy and safety of PCNL in 22 patients with ADPKD. PCNL was done in 25 renal units among 22 patients. Sixteen patients had chronic kidney disease, and the average stone burden was 2.4 cm $\pm$ 0.8 cm. The overall success rate (complete stone clearance/ residual fragments $\leq$ 4 mm) was 88% (22/25). Two patients required relook PCNL for residual stone and one required ESWL for the incomplete clearance. Mean preoperative serum creatinine in group I was 0.9 $\pm$ 0.1 mg% and in group II 3.1 $\pm$ 1.2 mg%. There was improvement in serum creatinine in group II (1.4 $\pm$ 0.5 mg% postoperatively). Three patients required blood transfusion (13%) and four patients had fever postoperatively (18%).

Similarly, a number of articles have reported PCNL to be safe and effective in patients with ADPKD. [14, 15] PCNL is a well-established procedure and can be widely used for a wide range of stones with minimal comorbidity and satisfactory results. Auxiliary procedures in the form of a second PCNL, ureteroscopy and ESWL may be necessary especially in cases of large renal stone burden. Bleeding is a known complication, needing blood transfusion during or after PCNL. In our series 17.6 % of the patients needed post-PCNL transfusions. Prolonged leak from the PCN site was noted in our series, probably due to rupture of cysts during PCNL or from infected cysts. Our study has its limitations in that, it is a retrospective study. A prospective study is necessary to know

the progression of these patients to chronic kidney disease and to know if our treatment has helped to temporarily pause this process.

CONCLUSION

Percutaneous nephrolithotomy in patient with ADPKD is safe and effective modality for management of renal calculi but it can be associated with postoperative bleeding requiring transfusions. Infection of the cysts may be a cause for fever, and prolonged urine leak. Careful selection of patients is extremely important to minimise complications

Conflicts Of Interest: None

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