Original Article

# Management of nephrolithiasis in autosomal dominant polycystic kidney disease — A single center experience

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## Abstract

**Purpose:** To evaluate available options for the management of nephrolithiasis in patients with autosomal dominant polycystic kidney disease (ADPKD).

**Materials and Methods:** Case files of all the patients with ADPKD treated in our hospital in the last 18 years were evaluated. Their demographic details, clinical presentations, investigations, treatments, and outcomes were critically analyzed.

**Results:** There were a total of 19 patients (23 renal units) with nephrolithiasis among 452 consecutive cases of ADPKD. Male-to-female ratio was 3.75:1. The mean age of the patients was 43.3 years (range 23 to 60 years). The most common presentations were pain and hematuria (27.7% each). Mean serum creatinine was 7.2 mg/dl (range 0.8-18.1 mg/dl) at presentation. The mean stone size was 115 $mm^2$ (range 36 to 980 $mm^2$). The majority of the stones were calyceal ($n = 10$). Ten renal units (nine patients) required intervention, while the rest were treated conservatively. Treatment offered included open nephrectomy for non-functioning infected kidney ($n = 1$), extracorporeal shock wave lithotripsy (ESWL, $n = 3$), ureterorenoscopy (URS, $n = 3$), and percutaneous nephrolithotomy (PCNL, $n = 3$). All patients undergoing URS and PCNL had complete clearance, while those undergoing ESWL had a residual stone. Two failed ESWL patients required an auxiliary procedure (retrograde intrarenal surgery, RIRS) and the other was kept under observation. Mean follow-up after treatment was 4.2 years (one month to six years). None of the patients had major complications.

**Conclusion:** Careful selection of the endourological procedure can give good results in patients of ADPKD with nephrolithiasis.

**Key Words:** Autosomal dominant polycystic kidney disease, nephrolithiasis, endourology

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## INTRODUCTION

Autosomal dominant polycystic kidney disease (ADPKD) accounts for 8-10% of the patients undergoing renal replacement therapy.[1] Although most patients exhibit an extended period of stability before a progressive decline in renal function, renal failure may be hastened by the onset of hypertension, infection, and nephrolithiasis. The incidence of nephrolithiasis in ADPKD ranges from 10-36%, and the stones produce morbidity by causing pain and obstruction and exacerbating the urinary tract infection.[2] Nearly half of the patients are symptomatic, of whom 20% ultimately require urologic intervention.[3] Ultrasonography is used primarily to diagnose such cases. With the advent of a CT scan, it is now the most commonly used modality for evaluation. Studies regarding the management of these patients are sparse in world literature. We present our experience in the management of nephrolithiasis in ADPKD patients.

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## MATERIALS AND METHODS

Medical records of patients with ADPKD since 1992 were reviewed. All patients were evaluated for urinary tract infection along with X-ray KUB (kidney, ureter, and bladder) [Figure 1] and ultrasonography [Figure 2]. In view of the deranged renal function, a non-contrast Computed Tomogram (CT) [Figure 3] was used for delineation of the local anatomy and size and location of the calculi. The type of intervention was chosen primarily on stone size, location, and presence of infection. Every case received consultation from the Nephrology Department as a part of the initial evaluation. Attention was paid to correct electrolyte imbalance and to treat urinary tract infection preoperatively. The obstruction was relieved in an urgent manner with either a DJ stent or percutaneous nephrostomy. Any stone more than 15 mm in the

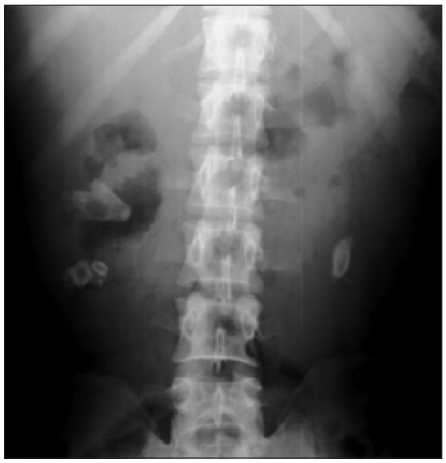

**Figure 1:** X-ray KUB of a patient with bilateral renal stones (Large renal shadows seen)

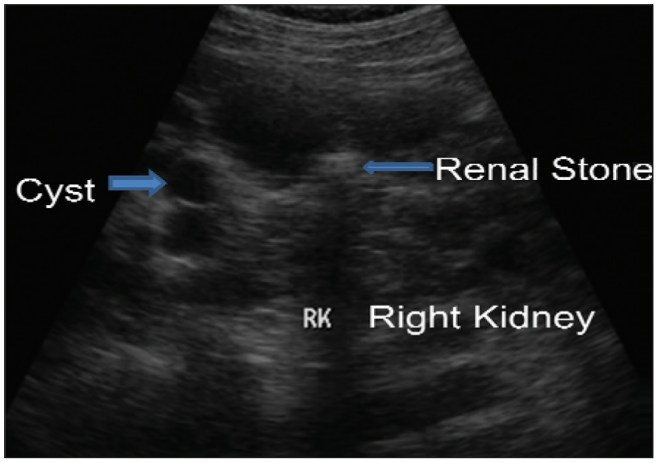


**Figure 2:** Ultrasonography of a patient with multiple cyst and renal stone

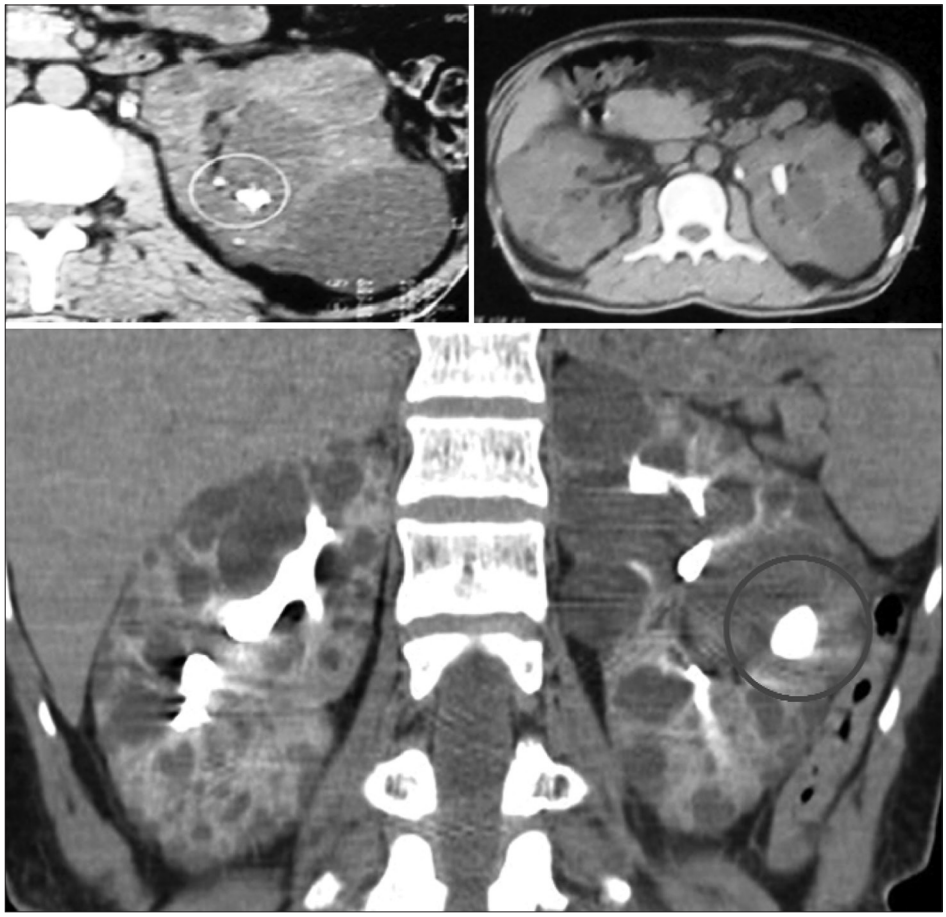

**Figure 3:** Showing CT scan of two patients with stones in left kidney

largest diameter was planned for percutaneous nephrolithotomy (PCNL). Small non-obstructing stones were treated conservatively. Conservative treatment included increased fluid intake, oral alkalization therapy, and regular follow-up. All patients were followed up with blood chemistries, urine examination, X-ray, and ultrasonography (USG) of the KUB region at one month, to locate any residual gravel. Subsequent follow-up comprised of three monthly visits.

## RESULTS

There were 19 patients (23 renal units) with ADPKD, who had nephrolithiasis. The patient demography is given in [Table 1].

Ten renal units in nine patients required interventions. The options offered were

1. Extracorporeal shock wave lithotripsy (ESWL)
2. Ureterorenoscopy (URS)
3. Retrograde intra renal surgery (RIRS)
4. Percutaneous nephrolithotomy (PCNL)
5. Nephrectomy

### Extracorporeal shock wave lithotripsy

ESWL was performed using Dornier Compact Delta in a single session in two patients. Shocks were limited to less than 1500 in one sitting. Only low energy shocks (Below 13 kV) were given to prevent cyst hemorrhage. Detail of the procedure is shown in [Table 2].

None of the patients experienced any hematuria or pain due to cyst hemorrhage.

### Percutaneous nephrolithotomy

Three patients underwent PCNL. Details are given in [Table 3].

Ultrasonography-guided puncture was attempted in order to prevent cyst rupture in two cases (One with multiple stones and another with a large pelvic stone). The third patient required a fluoroscopy-guided puncture as the pelvicalyceal system could not be identified properly because of multiple cysts. A total of four punctures and three tracts were used. Few stones were in inaccessible calyces. A puncture wash was found to be useful in such situations instead of making additional tracts, which could result in further nephron loss and increased morbidity. None of the patients required blood transfusion. The staghorn stone in the third patient was cleared in two stages.

### Ureterorenoscopy

Ureterorenoscopy was tried in two patients (three renal units). Details are given in [Table 4].

### LASER

One lady with a right lower ureteric stone was detected to have a stricture distal to the stone. LASER endoureterotomy with double J stenting stabilized the stricture part.

### Retrograde intra renal surgery

This procedure was performed in two patients for residual stones following ESWL. The stones were broken with a Holmium Yag LASER at an energy setting of 10-12 watts. A double J stent was kept for one month. Both the procedures were uneventful with patients being stone-free one month after the procedure.

### Nephrectomy

Nephrectomy was performed in one patient with calculus pyonephrosis, when his fever persisted even after putting in a percutaneous nephrostomy tube.

Overall, the mean hospital stay for all the patients undergoing intervention was five days (range four to twelve days).

**Table 1: Details of patient demography**

| | |
|---|---|
| No of patients | 19 |
| No of renal units involved | 23 |
| Male | 15 |
| Female | 4 |
| Average age | 43.3 years (23-60 years) |
| Hypertension | 17 patients |
| Average serum creatinine | 7.2 mg/dl (0.8-18.1 mg/dl) |
| No of patients with raised serum creatinine (>1.5 mg/dl) | 16 patients |
| No of patients requiring temporary dialysis | 7 patients |
| Symptoms | n = no. of patients |
| Hematuria | 5 |
| Pain | 6 |
| Anorexia | 3 |
| Vomiting | 3 |
| Weakness | 2 |
| Fluid overload | 2 |
| Fever | 1 |
| Asymptomatic | 2 |
| Mean stone size | 115 $mm^2$ (36-980 $mm^2$) |
| Stone locations | |
| Pelvic | 2 |
| Calyceal | 10 |
| Pelvicalyceal | 5 |
| Ureteric | 5 |
| Staghorn | 1 |

**Table 2: Details of extracorporeal shock wave lithotripsy**

| Patient | Stone size ($mm^2$) | Stone location | Total no of shocks | Result | Auxillary procedure | Follow up (year) |
|---|---|---|---|---|---|---|
| 1 | 209, multiple stones | Pelvis | 1800 | Incomplete clearance – 8 mm residual stone | RIRS | 3 |
| 2 | 99, single stone | Pelvis | 4000 | Incomplete clearance – 6 mm residual stone | No | 1 |
| 3 | 49 | Calyceal | 1500 | Incomplete clearance | RIRS | 1/2 |

**Table 3: Details of percutaneous nephrolithotomy**

| Stone size ($mm^2$) | Stone location | Result | Operating time (min) | Hemoglobin drop (gm/dl) | Serum creatinine drop (mg/dl) | Complications | Follow up (years) |
|---|---|---|---|---|---|---|---|
| 309 | Multiple calyceal | Complete clearance | 3p0.143 | 3.5 | 1.3 | Nil | 3 |
| 135 | Pelvic | Complete clearance | 95 | 0.5 | 0.4 | Post operative fever | 2 |
| 455 | Staghorn | Complete clearance | 135 | 1.2 | 0.9 | Pain in operating site | 1.5 |

**Table 4: Details of ureterorenoscopy**

| Stone size ($mm^2$) | Stone location | Result | Complications | Follow up (Months) | Comment |
|---|---|---|---|---|---|
| 153 | Rt lower ureter | Complete clearance | Nil | 6 | Needed LASER endoureterotomy for stricture in the ureter |
| 118 | Rt upper ureter | Complete clearance | Nil | 8 | Uneventful procedure |
| 72 | Lt upper ureter | Complete clearance | Nil | 8 | Uneventful procedure |

Subsequent to the conservative management, including percutaneous nephrostomy or dialysis, as indicated, the mean serum creatinine, prior to intervention, averaged at 2.4 mg/dl (range 1.1 to 18.1). The mean serum creatinine after intervention was 1.4 mg/dl. (range 1.3 to 2.1). Two patients were detected to have chronic kidney disease status on follow-up. However, they are yet to receive any renal replacement therapy after a follow-up of three years.

### Conservative treatment

Ten patients were treated conservatively. Of them, two patients were lost to follow-up and three patients underwent renal transplant. However, intervention for stone disease was not required in any of these patients in a mean follow-up of three years. Urine metabolic study was available in only five patients. Of these, three patients had only hypocitraturia, one had only hypercalciuria, and one patient had both. Stone analysis was available in only seven patients. Three had uric acid stones, while the others had calcium oxalate monohydrate stones.

## DISCUSSION

Autosomal dominant polycystic kidney disease is a common presentation in any nephro-urological set-up. Nephrolithiasis in such patients is seen in 10-36% of the cases.[2] In our series, the incidence of stone disease in ADPKD patients was 19/452 (4.2%). The exact etiology is still not completely elucidated. However, both the anatomic and metabolic factors are implicated as causative factors. Urinary stasis occurs due to large cysts. These patients have a low glomerular filtration rate (GFR), urinary volume, a lower level of magnesium, phosphorus, potassium, and citrate in the urine.[3-5] Unfortunately in our series, the result of urine metabolic evaluation was available only in five patients, to come to any conclusion. Diagnosis of nephrolithiasis in such patients is challenging due to a distorted anatomy, cyst wall, and associated parenchymal calcifications.[3] We believe a CT scan holds an edge over USG or intravenous urography in the diagnosis of stone disease.[6] A CT scan is helpful to detect radiolucent stones and to differentiate it from calcifications. A three-dimensional reconstruction of the stone helps in the planning of the percutaneous tract in PCNL. Intravenous urography is not possible in all patients due to the associated renal dysfunction. Plain X-ray KUB might not show all the stones, as most of them are radiolucent uric acid stones (56-71%).[7]

Asymptomatic patients can be managed conservatively, for example, oral alkalization therapy.[1]

However, half of the patients are symptomatic.[1] In our series only two patients (10.5%) were asymptomatic and one of them had a staghorn stone. Few decades ago all such patients had to undergo the trauma of open surgery.[3] All sorts of endourological processes were tried in such patients. ESWL is the most commonly performed procedure.[1] Success rates from 43 to 85% have been reported.[1] In the series published by Delakas in 1997, an average of 1800 shocks were given with low energy (Below 21 kV). Transient gross hematuria was the observation in the postoperative period. There was no cyst hemorrhage, decrease in hemoglobin or deterioration in the renal function. The stone-free rate was 85% at three months.[5] With improvement in the armamentarium, URS and RIRS have become exciting options in the management of these patients. Percutaneous management has been rarely performed in the published series.[1] Concerns of percutaneous puncture are cyst puncture, cyst bleeding, and infection. Also getting access to the pelvicalyceal system is difficult. Need for special smaller caliber instruments with extralength and not be overemphasized in view of altered anatomy and long and narrow infundibulum. However, Umbreit *et al.,* found PCNL to be safe and efficacious for patients of ADPKD with a large stone burden, despite increased operative complexity, need of multiple punctures, and repeat endoscopy.[7] We prefer an ultrasonography-guided puncture to a pelvicalyceal system, to avoid cyst puncture during PCNL. Being already with a reduced renal reserve, this group of patients needs special care. From this study, we would like to emphasize a few points.

As most patients are asymptomatic, suspicion is important

to recognize stones. Team approach including the treating urologist and nephrologist is important. Preoperative correction of electrolyte imbalance is essential, and if required dialysis. Preoperative correction of urinary tract infection and urgent de-obstruction of the system may be performed if indicated. Careful selection of the procedure should be done in an individualized fashion. Additional puncture in PCNL can be avoided by a puncture wash. Vigilant follow-up of patients and patient counseling/education regarding the disease are important.

There are multiple limitations of the study. Being a retrospective study, details of all patients were not available. Few patients were lost to follow-up. The number of patients was small.

## CONCLUSIONS

Team approach and careful selection of the endourological procedure can give good result in patients of ADPKD with nephrolithiasis. However, a further multi-institutional study is required for better understanding, better treatment, and better outcome of the condition.

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