

# Polycystic Kidney Disease and the Presence of a Pyocyst

Raed Bargout, MD  
Christopher A. Smith, MD

**P**atients with autosomal dominant polycystic kidney disease (ADPKD) sometimes develop an infection in one or more of the cysts present in their kidneys. The infection of a cyst (pyocyst) within a polycystic kidney is a well-known and serious complication of ADPKD.<sup>1</sup> This article describes the clinical course of a 46-year-old woman with ADPKD who developed an infection in at least one of the cysts in her kidneys. The article discusses difficulties pertaining to the diagnosis and treatment of a pyocyst complicating ADPKD.

## CASE PRESENTATION

### Initial Presentation and Management

A 46-year-old woman with a recent diagnosis of ADPKD and a 4-day history of fever, urinary frequency, and dysuria presented to our clinic for evaluation. Except for a family history of ADPKD (relating to her father), there was no other remarkable information in her medical history. Upon evaluation, the patient had no gross hematuria or flank pain. She was initially treated with a 7-day outpatient course of oral ciprofloxacin. However, her symptoms persisted despite the therapy, and she later developed mild right flank pain. She was subsequently admitted for further evaluation.

### Continued Evaluation and Management

**Physical examination.** After the patient was admitted, a physical examination was performed, yielding the following results: temperature, 103°F; pulse, 105 beats/min; and blood pressure, 110/75 mm Hg. An abdominal examination revealed mild bilateral flank tenderness and a liver span of 16 cm. The results of the remainder of the physical examination were unremarkable.

**Laboratory evaluation.** Results from a complete blood count and blood and serum chemistry evaluations are provided in **Table 1**. A urinalysis yielded the following results: no protein in the patient's urine; urine pH, 5.5 (normal, 5.0–9.0); 69 erythrocytes/high-power field (hpf) (normal, 0–2 erythrocytes/hpf); and

81 leukocytes/hpf (normal, 0–5 leukocytes/hpf). Examination of the urinary sediment did not show evidence of casts. An initial urine culture yielded more than 100,000 colony-forming units (CFUs) of *Escherichia coli* per mL that were susceptible to ciprofloxacin. Blood cultures remained negative for bacteria after 5 days.

**Imaging studies.** A radiograph of the kidney, ureters, and bladder was obtained and showed no evidence of renal stones. Computed tomography (CT) scans of the abdomen and pelvis showed multiple low-attenuation lesions (cysts) in both the liver and kidneys (**Figure 1**). In the liver, the cysts were from 1 to 4 cm in size; in the kidneys, they were from 1 to 10 cm. There was no evidence of a renal parenchymal or perinephric abscess. Ultrasonography confirmed the presence of enlarged kidneys, containing multiple cysts of various sizes; some of the cysts showed internal echo, indicating either cystic hemorrhage or infection (**Figure 2**).

**Initial management.** The patient was started on intravenous ciprofloxacin. However, after 7 days of intensive antibiotic therapy, there was only a slight improvement in her symptoms, and her fever persisted. Repeated blood cultures were consistently negative for bacteria, whereas subsequent urine cultures continued to grow *E. coli* bacteria that were susceptible to ciprofloxacin. A second abdominal CT scan and a gallium scan failed to provide any useful additional information.

**Diagnosis and treatment.** The patient's clinical presentation prior to admission and the urine culture and urinalysis results strongly suggested an infectious process rather than hemorrhage into a cyst. Because of the absence of a clinical response after appropriate antibiotic

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*Dr. Bargout is a Senior Resident, Division of Internal Medicine, Cook County Hospital, Chicago, IL. Dr. Smith is an Assistant Professor of Medicine, Rush Medical College/Cook County Hospital, Chicago, IL, and an Attending Physician, Division of General Internal Medicine, Cook County Hospital, Chicago, IL.*

**Table 1.** Results from a Complete Blood Count and Blood and Serum Chemistry Evaluations of the Case Patient

Component Measured	Result	Reference Value
Hemoglobin mass concentration (g/dL)	12.2	12.0–16.0
Total leukocyte count (cells/ $\mu$ L)	$17.6 \times 10^3$ *	$4.0 \times 10^3$ – $10.0 \times 10^3$
Platelet count (platelets/ $\mu$ L)	$424 \times 10^3$	$140 \times 10^3$ – $440 \times 10^3$
Blood urea nitrogen level (mg/dL)	74	8–20
Creatinine level (mg/dL)	3.5	0.6–1.4
Serum sodium level (mEq/L)	135	136–146
Serum potassium level (mEq/L)	4.4	3.5–5
Alkaline phosphatase level (U/L)	245	36–92
Aspartate aminotransferase level (U/L)	76	0–40
Alanine aminotransferase level (U/L)	61	5–35
$\gamma$ -glutamyltransferase level (U/L)	132	3–60
Total bilirubin level (mg/dL)	0.3	0.3–1.2

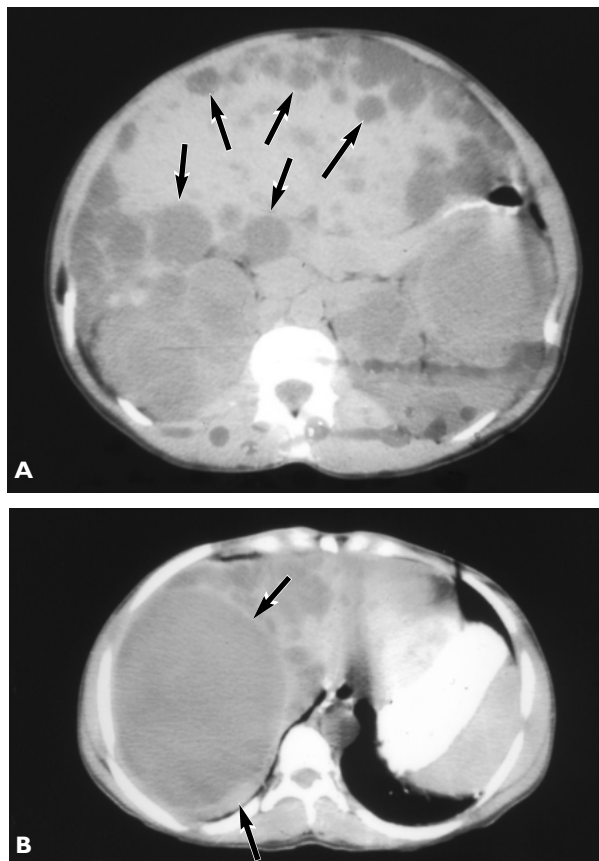
\*93% Neutrophils.

therapy for pyelonephritis, a diagnosis of pyocyst was made.

The patient was discharged home and treated on an outpatient basis with a long course of oral ciprofloxacin. This was done despite her persistent fever because the majority of patients with ADPKD and a pyocyst will eventually respond to a prolonged course of oral antibiotics. We considered percutaneously draining the cyst but decided against it because of the difficulty in correctly identifying an infected cyst. On a follow-up evaluation at the clinic 4 weeks later, she reported a complete resolution of her symptoms.

## DISCUSSION

Fifty percent to 75% of patients with ADPKD will experience one or more symptomatic lower urinary tract infections during their lifetime,<sup>2</sup> and these patients usually respond quickly to a standard course of antibiotics. In contrast, upper urinary tract infections



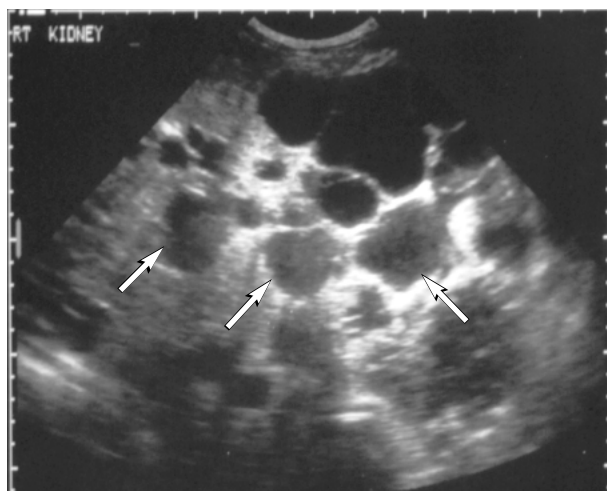
**Figure 1.** Computed tomography scans of the abdomen of the case patient. (A) Multiple cysts in the liver (arrows). (B) 10-cm cyst in the right kidney (arrows).

are less common and tend to have a more complicated course. They are often difficult to diagnose and treat.

## Diagnosis of a Pyocyst Complicating ADPKD

Diagnosing a pyocyst in patients with ADPKD can be difficult because such a cyst can produce signs and symptoms similar to those resulting from hemorrhage into a cyst or acute pyelonephritis. For example, patients with a pyocyst and ADPKD may experience acute flank pain. However, acute flank pain can also indicate hemorrhage into a cyst and acute pyelonephritis, as well as a number of other disorders (Table 2).

Diagnostic imaging tests are not reliable for distinguishing pyocysts, acute pyelonephritis, and hemorrhage into a cyst. CT scans of the abdomen are good for determining the presence or absence of renal or perinephric abscesses.<sup>3</sup> However, neither CT scans nor sonograms can be used to reliably distinguish uninfected cysts, pyocysts, or hemorrhage into a cyst in patients with ADPKD. Gallium scans have a low sensitivity



**Figure 2.** Sonogram of the right kidney (transverse view) of the case patient, showing multiple cysts with internal echoes (arrows), suggesting hemorrhage or infection.

and specificity for localizing an infected cyst<sup>4</sup>; results of gallium scans are positive in only approximately 50% of cases of pyocysts.<sup>4</sup>

Hemorrhage into a cyst should be suspected when there is an abrupt onset of abdominal or flank pain with the absence of dysuria or other urinary symptoms. In contrast, a pyocyst or pyelonephritis is likely to be present when the patient experiences an abrupt onset of pain in conjunction with dysuria and frequent fever. Moreover, a pyocyst or pyelonephritis is likely to be present if positive results are obtained from urine cultures testing for the presence of bacteria; however, urine culture results can also be negative for bacteria in patients with pyocysts.<sup>4</sup> This is because many cysts do not communicate with the rest of the urinary tract; thus, a patient with a pyocyst may not experience lower urinary tract symptoms (eg, dysuria) and bacteriuria. Also, pyuria in the absence of infection occurs relatively frequently (45% of cases) in patients with ADPKD. However, the presence of leukocyte casts on urinalysis is highly suggestive of pyelonephritis. The presence of a pyocyst should be strongly considered in any patient who shows an incomplete response after 7 days of treatment for pyelonephritis with ampicillin/gentamicin or fluoroquinolones.

#### Treatment of a Pyocyst Complicating ADPKD

Antimicrobial therapy is the mainstay for treatment of a pyocyst. Favorable patient outcomes are most often achieved with the use of fluoroquinolones, trimethoprim-sulfamethoxazole, or chloramphenicol.<sup>5,6</sup> All of these antibiotics have been shown to have better penetration into the cysts than other agents have.

**Table 2.** Differential Diagnosis of Acute Flank Pain in Patients with Polycystic Kidney Disease

Acute hemorrhage into a cyst
Acute pyelonephritis
Bleeding inside renal adenocarcinoma
Cyst rupture
Papillary necrosis
Pyocyst
Renal abscess/perinephric abscess
Renal infarction
Renal stone

There is no evidence that combinations of antibiotics are any more successful than monotherapy, and the optimal duration of therapy is controversial. Patients are often treated for 4 to 6 weeks and usually have a high response rate.<sup>4</sup> If the infection recurs after antibiotic withdrawal, therapy should be restarted and administered for up to 3 months or longer.

Percutaneous drainage and surgical drainage of the pyocyst are alternative options for treatment and have been described in the literature.<sup>7,8</sup> However, the difficulty in accurately identifying a pyocyst through imaging studies limits the effectiveness of these options. In severe cases of recurrent or refractory infections, nephrectomy may be required as a last resort, especially for patients who are about to undergo hemodialysis (which is often the case for 35% to 45% of patients age 60 years or older) or renal transplantation.<sup>9</sup>

#### CONCLUSION

The presence of a pyocyst within a polycystic kidney is a well-known and serious complication of ADPKD. The clinical course of the patient presented in this report illustrates the challenge in diagnosing and treating this condition and emphasizes the importance of considering a prolonged course of antibiotic therapy when the infected cyst cannot be identified. Physicians must consider the diagnosis of an infected cyst when caring for a febrile patient with ADPKD and must be familiar with the available therapeutic options. **HP**

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