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Case Report

# When Polycystic Kidneys Become a Surgical Burden: Bilateral Nephrectomy in Advanced Autosomal Dominant Polycystic Kidney Disease - A case report from Azerbaijan

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## ABSTRACT

Bilateral nephrectomy remains an important yet selectively utilized intervention in advanced autosomal dominant polycystic kidney disease (ADPKD), particularly in patients with end-stage renal disease (ESRD) and severe symptom burden. Progressive cystic enlargement can lead to debilitating flank pain, recurrent hemorrhage, abdominal distension, and reduced quality of life, often necessitating surgery when conservative measures fail.

This report highlights the clinical role of bilateral nephrectomy in effectively controlling symptoms and reducing disease burden. In cases of massively enlarged kidneys, an open surgical approach may be required, making careful perioperative planning essential for favorable outcomes.

Beyond symptom relief, bilateral nephrectomy also creates anatomical space for future renal transplantation, serving both therapeutic and preparatory purposes. When performed in appropriately selected patients, it is associated with significant clinical improvement and acceptable risk.

This case is notable for the massive renal burden (17 kg total kidney weight) necessitating bilateral nephrectomy, highlighting surgical decision-making in resource-limited settings and reinforcing its role as a definitive symptom-relieving intervention rather than solely a preparatory step for transplantation.

***Keywords****: Autosomal dominant polycystic kidney disease, Bilateral nephrectomy, End-stage renal disease, Hemodialysis, Renal transplantation, Polycystic liver disease.*

## INTRODUCTION

Autosomal dominant polycystic kidney disease (ADPKD) is one of the most common inherited renal disorders, primarily associated with mutations in the PKD1 and PKD2 genes. It is characterized by progressive bilateral kidney enlargement due to the development of numerous fluid-filled cysts, which gradually replace normal renal parenchyma and impair renal function. Clinical manifestations such as hypertension, hematuria, and flank pain often appear early, and many patients eventually progress to end-stage renal disease (ESRD) in adulthood [1, 2, 3].

Renal transplantation remains the definitive treatment for ESRD; however, removal of native kidneys may be required in specific situations. Indications for nephrectomy include persistent or severe pain, recurrent cyst infection or hemorrhage, nephrolithiasis, and the need to create adequate space for future transplantation [4, 5]. While unilateral nephrectomy is more common, bilateral nephrectomy may be necessary in patients with extensive involvement of both kidneys. This case describes a patient with advanced ADPKD who required bilateral nephrectomy due to significant symptom burden and massive kidney enlargement.

### Case Presentation

A 43-year-old man with a known diagnosis of autosomal dominant polycystic kidney disease (ADPKD), incidentally identified in 2011, was followed regularly until progression to end-stage renal disease (ESRD), requiring initiation of hemodialysis in October 2025 due to worsening uremic symptoms; his medical history included prior bilateral inguinal hernia repair, and his social history was notable for occasional smoking with no alcohol use. A strong family history of ADPKD was present in his father and multiple siblings.

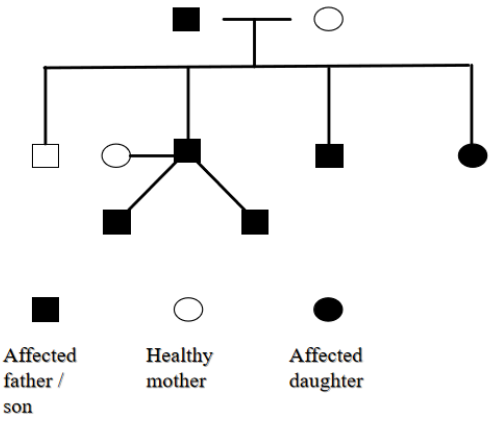


***Figure 1: Pedigree chart illustrating family history of Autosomal Dominant Polycystic Kidney Disease (ADPKD)***.

In the months prior to admission, he developed progressively worsening right-sided flank pain, later involving the left side, along with intermittent gross hematuria, increasing abdominal girth, nausea, and generalized fatigue. On examination, he appeared uncomfortable, with a blood pressure of 150/100 mmHg. Abdominal palpation revealed large, tense bilateral flank masses consistent with markedly enlarged kidneys.

Laboratory evaluation showed advanced renal failure, with serum creatinine of 6.64 mg/dL and blood urea nitrogen of 60.4 mg/dL. Hemoglobin was 9 g/dL, consistent with chronic kidney disease-related anemia, and C-reactive protein was elevated at 109.6 mg/L, suggesting inflammation. Histopathological examination was not performed due to logistical constraints; however, intraoperative and radiological findings were consistent with advanced ADPKD, with no gross evidence of malignancy.

Contrast-enhanced computed tomography demonstrated markedly enlarged kidneys occupying most of the retroperitoneal space, with numerous cysts of varying sizes. A 7 × 5 cm cyst in the lower pole of the right kidney and a 12 × 10 cm cyst in the midportion of the left kidney showed hyperdense components consistent with intracystic hemorrhage. Multiple small hepatic cysts were also noted, consistent with polycystic liver disease, without evidence of malignancy or obstruction.

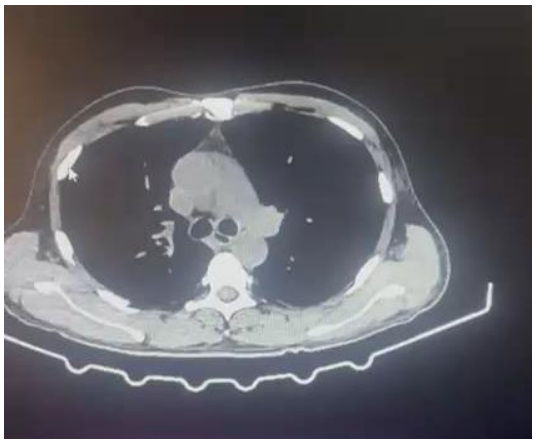

***Figure 2 : Preoperative abdominal imaging demonstrating massively enlarged polycystic kidneys in ADPKD.***

Given persistent pain, abdominal distension, and recurrent cyst hemorrhage, bilateral nephrectomy was performed via an open approach due to the massive size of the kidneys. Intraoperatively, both kidneys were markedly enlarged and extensively replaced by cysts. The procedure was technically challenging and required intraoperative blood transfusion but completed without major complications. The combined weight of the resected kidneys was approximately 17 kg.

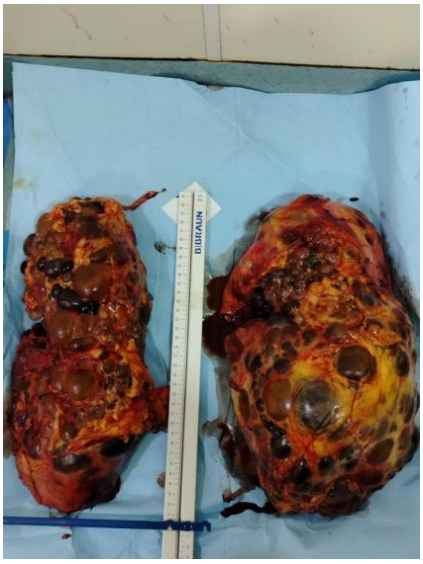

***Figure 3 : Comparative gross morphology of both kidneys following bilateral nephrectomy.***

Postoperatively, the patient was initially managed in the intensive care unit and then transferred to the general ward, with an uneventful recovery. His body weight decreased from 81 kg to 64 kg, reflecting removal of approximately 17 kg of renal tissue. Hemodialysis was resumed shortly after surgery. At discharge, flank pain had completely resolved, hematuria had ceased, and overall clinical condition had improved. He was scheduled for continued dialysis and further evaluation for renal transplantation.

**DISCUSSION**

This case illustrates the natural progression of autosomal dominant polycystic kidney disease (ADPKD) from an initially asymptomatic condition to advanced disease with significant morbidity. Progressive cyst enlargement leads to compression of normal renal parenchyma, culminating in loss of renal function and end-stage renal disease (ESRD). In addition to renal failure, complications such as chronic pain, cyst hemorrhage, and infection can markedly impair quality of life [2, 6].

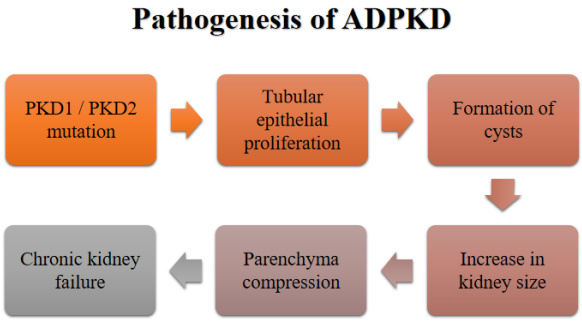


***Figure 4 : Schematic illustration of the pathogenesis of Autosomal Dominant Polycystic Kidney Disease (ADPKD).***

In this patient, persistent bilateral flank pain and recurrent hematuria were the primary indications for surgical intervention. Although nephrectomy is not routinely required in ADPKD, it becomes necessary in cases of severe, refractory symptoms. The decision for bilateral nephrectomy, rather than unilateral removal, was based on extensive involvement of both kidneys and bilateral symptom burden. While recognized, cases with extreme renal enlargement and significant mass effect remain uncommon and present unique surgical and perioperative challenges [7, 8, 9].

Bilateral nephrectomy is a major procedure with inherent risks; however, when performed electively in experienced centers, outcomes are generally favorable due to advances in perioperative care and surgical techniques [4]. Despite the growing adoption of minimally invasive approaches, open surgery remains necessary in cases of massive kidney enlargement, as seen in this patient.

ADPKD is the most common hereditary cystic kidney disorder but requires differentiation from other cystic diseases, particularly in atypical presentations. Integration of clinical, imaging, and genetic data is essential to distinguish it from conditions such as autosomal recessive polycystic kidney disease, nephronophthisis, and syndromic disorders including tuberous sclerosis complex and von Hippel–Lindau disease [1]. These distinctions are important due to differences in disease progression, extrarenal manifestations, and management.

**Table 1 summarizes key differential diagnoses relevant to our case.**

| Differential Diagnosis | Genetic Basis / Etiology | Age of Onset | Key Clinical Features | Imaging Findings | Distinguishing Features from ADPKD | Diagnostic Clues / Tests |
|---|---|---|---|---|---|---|
| **Autosomal Recessive Polycystic Kidney Disease (ARPKD)** | PKHD1 mutation | Neonatal / infancy | Enlarged kidneys, hepatic fibrosis, portal hypertension | Bilaterally enlarged echogenic kidneys with microcysts | Earlier presentation, liver involvement prominent | Genetic testing (PKHD1), prenatal ultrasound |
| **Simple Renal Cysts** | Age-related degeneration | >50 years | Usually asymptomatic | Few, unilateral or bilateral simple cysts | No progressive renal failure, no family history | Ultrasound/CT showing isolated cysts |
| **Acquired Cystic Kidney Disease (ACKD)** | Secondary to long-term dialysis | Adulthood (ESRD patients) | Often asymptomatic, risk of RCC | Small kidneys with multiple cysts | Occurs after ESRD onset, not hereditary | History of chronic dialysis |
| **Medullary Sponge Kidney** | Developmental abnormality | Early adulthood | Hematuria, nephrolithiasis | Cystic dilatation of collecting ducts | Medullary (not cortical) cysts, normal kidney size | IV urography (paintbrush appearance) |
| **Tuberous Sclerosis Complex (TSC)** | TSC1/TSC2 mutations | Childhood–adulthood | Seizures, skin lesions (adenoma sebaceum), angiomyolipomas | Renal cysts with angiomyolipomas | Extrarenal manifestations (brain, skin) | Genetic testing, MRI brain |
| **Von Hippel–Lindau (VHL) Disease** | VHL gene mutation | Early adulthood | Hemangioblastomas, RCC, pheochromocytoma | Multiple renal cysts and tumors | Strong association with malignancies | Genetic testing, screening for tumors |
| **Multicystic Dysplastic Kidney (MCDK)** | Developmental anomaly | Prenatal / infancy | Usually unilateral, non-functioning kidney | Multiple non-communicating cysts | Typically unilateral, contralateral kidney compensates | Prenatal USG, radionuclide scan |
| **Nephronophthisis** | NPHP gene mutations | Childhood–adolescence | Polyuria, polydipsia, anemia | Small or normal-sized kidneys with corticomedullary cysts | No kidney enlargement, early ESRD | Genetic testing, renal biopsy |
| **Glomerulocystic Kidney Disease** | Heterogeneous genetic causes | Variable | Hypertension, renal insufficiency | Predominantly cortical cysts | Cysts limited to Bowman's space | Histopathology |
| **Renal Cysts and Diabetes (RCAD syndrome)** | HNF1B mutation | Adolescence–adulthood | Renal cysts + early-onset diabetes | Small kidneys with cysts | Associated diabetes, genital tract anomalies | Genetic testing (HNF1B) |

***Table 1**: Differential Diagnosis of Cystic Kidney Diseases Mimicking ADPKD*

Contemporary evidence highlights ADPKD as a systemic disease with multisystem involvement, including hepatic cysts, intracranial aneurysms, and cardiovascular complications, all of which may influence management decisions [2]. The development of disease-modifying therapies, such as vasopressin receptor antagonists, underscores the importance of early and accurate diagnosis [3]. Nevertheless, many patients progress to ESRD, requiring renal replacement therapy and, in selected cases, surgical intervention.

The role of native nephrectomy in ADPKD, particularly in the context of ESRD and transplantation, remains debated. Current consensus recommends individualized decision-making based on severe clinical manifestations such as refractory pain, recurrent infection, hematuria, and mass effect [4]. Similarly, systematic evaluations suggest that routine pre-transplant nephrectomy is not universally indicated but should be reserved for symptomatic patients [5], which supports the approach taken in this case.

Recent surgical trends favor laparoscopic nephrectomy due to reduced morbidity and faster recovery; however, bilateral nephrectomy in ADPKD remains technically challenging due to massive organ size and distorted anatomy, often requiring open surgery and multidisciplinary planning [9]. Case-based reports also highlight rare findings such as incidental renal cell carcinoma following nephrectomy, emphasizing the importance of histopathological evaluation [6]. Retrospective

studies confirm that nephrectomy in ADPKD is most often performed for symptom control, with generally favorable outcomes when patients are appropriately selected [9].
The presence of hepatic cysts in this patient further underscores the systemic nature of ADPKD, polycystic liver disease, a common, usually asymptomatic, extrarenal manifestation [2]. Differential diagnosis of cystic renal disorders remains essential, particularly in atypical cases [1].

Following bilateral nephrectomy, patients become fully dependent on dialysis until transplantation; however, removal of significantly enlarged kidneys can provide substantial symptomatic relief, reduce abdominal distension, and eliminate sources of recurrent bleeding. In this case, the patient experienced complete resolution of pain and hematuria, along with marked improvement in overall well-being. This case also demonstrates the feasibility of managing advanced ADPKD with complex surgical intervention in a resource-limited setting.

**Limitations**
This report is limited by its single-patient design, which restricts generalizability. Additionally, long-term postoperative outcomes and transplant follow-up were not available at the time of reporting. Histopathological evaluation was not performed due to logistical constraints, representing a limitation, particularly given the known risk of occult malignancy in ADPKD. Further studies are needed to better define optimal timing and patient selection for bilateral nephrectomy in ADPKD.

## CONCLUSION
Autosomal dominant polycystic kidney disease can progress beyond renal failure to produce a significant systemic and mechanical disease burden due to massive cystic enlargement [2]. This case highlights that in patients with end-stage disease who develop debilitating symptoms such as intractable pain, recurrent hemorrhage, and marked abdominal distension, bilateral nephrectomy can serve as a definitive therapeutic intervention rather than merely a preparatory step for transplantation [4, 5].
The favorable postoperative outcome observed in this patient, including complete symptom resolution and improved functional status, reinforces the value of timely surgical decision-making in carefully selected individuals. Despite the inherent risks associated with major surgery, elective bilateral nephrectomy performed in a controlled setting can be both safe and highly effective [9].
To our knowledge, this is among the few reported cases of ADPKD requiring bilateral nephrectomy for such extreme renal mass (17 kg), particularly in a resource-limited setting. This case challenges the perception of nephrectomy as merely preparatory and supports its role as a definitive quality-of-life intervention in advanced ADPKD.
This case emphasizes the importance of a multidisciplinary approach in managing advanced ADPKD and supports consideration of bilateral nephrectomy as a means to improve quality of life and facilitate future renal transplantation in patients with severe symptomatic disease [4].

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