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# Case Report
# An Interesting Case of Neonatal Kidney Biopsy

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## ABSTRACT

**Background and Aim:** Kidney biopsies are seldom performed these days in neonates, especially after the advent of advanced antenatal imaging and the widespread availability of genetic studies.

**Case Presentation:** We described a newborn with kidney failure, for whom a biopsy was conducted, leading to a diagnosis of glomerulo cystic kidney disease.

**Conclusion:** Sporadic glomerulocystic kidney disease can manifest as renal failure during the newborn period. Renal biopsy may be used to diagnose glomerulocystic kidney disease in this setting.

**Keywords:** Neonate, Kidney failure, Kidney biopsy, Glomerulocystic kidney disease

## Introduction

Glomerulocystic kidney disease is a heterogeneous group of disorders with different underlying etiologies. It is usually diagnosed by radiology and rarely by histopathology.

## Case Presentation

A male newborn was referred at 48 hours of life with complaints of anuria noted since birth. The baby was born at 36 weeks to an elderly (40-year-old) primi mother and a 45-year-old father from a non-consanguineous marriage.

The mother had pregestational hypertension that was not well controlled. There was no history of diabetes mellitus or any renal illness. She had no prior antenatal checkups or first and second trimester ultrasound imaging. At 34 weeks of gestational age, she was found to have oligohydramnios, with an amniotic fluid index of 4 (AFI-4) and intrauterine growth restriction, and the baby was delivered by cesarean section two weeks later. The baby weighed 2.3 kg and cried soon after birth. However, there was respiratory distress, necessitating oxygen administration and continuous positive airway pressure (CPAP).

Upon examination, the baby had no dysmorphic features, bony anomalies, neurocutaneous markers, or Potter's facies. His vital signs showed a heart rate of 150 beats per minute, a respiratory rate of 55 breaths per minute, and a blood pressure of 106/64 mm Hg. The baby's head circumference was 33 cm, and his length was 48 cm. He had no palpable kidneys or urinary bladder, and his external genitalia were of normal male type. The systemic examination was within normal limits. He presented with mild facial and presacral edema upon admission. His renal function tests revealed a blood urea

J Ped Nephrol
Journal of Pediatric Nephrology

level of 106 mg/dl and a serum creatinine level of 5.6 mg/dl. Ultrasound imaging revealed normally sized kidneys bilaterally for his gestational age, albeit with altered echogenicity. The corticomedullary differentiation was partially preserved, and no cysts or hydronephrosis were noted. The urinary bladder was nearly empty. There was no hepatomegaly or altered liver echogenicity. His cardiac echocardiography appeared normal, and his ophthalmology evaluation was normal (no coloboma or retinitis pigmentosa).

He passed a small amount of urine during catheterisation; however, he remained oliguric and developed hyperkalemia and metabolic acidosis. His blood counts were normal, and there was no anemia. Urine analysis indicated 2+ albuminuria with 10-15 white blood cells per high power field (hpf) and 5-10 red blood cells per hpf. He was started on acute peritoneal dialysis. There was uncertainty regarding whether the mother had taken angiotensin-converting enzyme inhibitors (ACEi) for pregestational hypertension, but we could not confirm or rule it out based on any records.

The probable differentials considered at this point were: 1) Bilateral hypodysplastic kidneys; 2) Nephronophthisis and related ciliopathies; 3) Renal tubular dysgenesis related to antenatal ACE inhibitor exposure; and 4. Hepatocyte nuclear factor β (HNF β)/uromodulin/renin (REN/UMOD)-related autosomal dominant tubulointerstitial kidney disease (ADTKD).

There was no family history of renal illness. Both parents underwent evaluation through ultrasound imaging of the kidneys, which revealed normal results. Neither parent had hyperuricemia, hypomagnesemia, or diabetes mellitus. The family was counseled about a genetic study, but they were unwilling to proceed. The baby remained on peritoneal dialysis for the next five days, but there was no improvement in urine output. The family was counseled about the insertion of a continuous ambulatory peritoneal dialysis (CAPD) catheter for the baby, but they wanted to know the etiology and prognosis of the patient. At this point, the option for a high-risk renal biopsy was discussed with them, and they were willing to proceed. We used an 18-gauge needle for the renal biopsy, and only one core was taken due to a small perinephric hematoma post-biopsy. The hematoma did not progress and did not require any further interventions.

### Histopathology

There were ten glomeruli observed in the light microscopy. The glomeruli appeared immature, showing prominent cystic dilatation of the Bowman's space, up to three times the normal size (Figure 1). The tubules also appeared immature, with focal cystic dilatation present. A diagnosis of glomerulocystic disease with renal tubular dysgenesis was considered.

The irreversible and likely genetic nature of the disease was conveyed to the parents; unfortunately, they discontinued the treatment, and the baby was discharged against medical advice.

## Discussion

Fetal and early neonatal renal failure requiring the initiation of kidney replacement therapy is rare. Most of the time, the etiology is determined through good ultrasound imaging or genetic analysis studies. Unfortunately, in our patient, we lacked adequate antenatal imaging, and postnatal imaging was inconclusive. There was no relevant family history indicating any specific etiology.

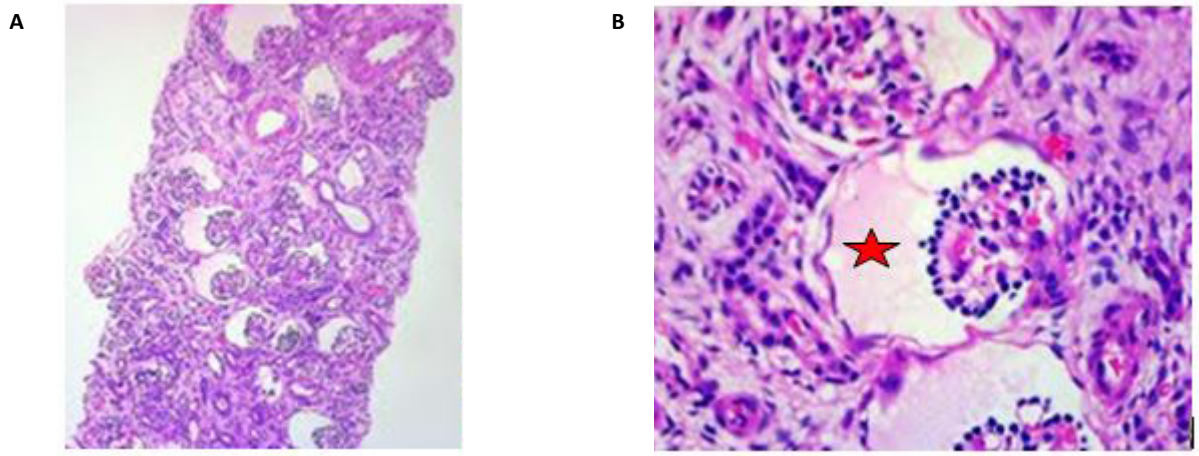


**Figure 1.** Prominent cystic dilatation of the Bowman's spaces

GCKD is a pathologically descriptive term that has been recognized for a long time. Bernstein defined GCKD as the dilatation of Bowman's space, up to two to three times in at least 5% of the glomeruli, and classified it into three groups [1]:

1) Non-syndromic inheritable and sporadic conditions such as polycystic kidney disease (PKD) (PKD1, PKD2); 2) Part of inheritable malformation syndromes (e.g. tuberous sclerosis, Meckel-Gruber syndrome); and 3. Dysplastic kidneys (part of congenital anomalies of the kidney and urinary tract).

After the advent of widespread genetic analysis, nearly 20 inherited genetic malformation syndromes and ciliopathy-related renal diseases have been found to be associated with GCKD [2].

Dysplastic and acquired varieties of GCKD secondary to urinary tract obstruction are also recognized and are generally diagnosed through effective ultrasound imaging. A recent addition to the etiology of GCKD is the spectrum of autosomal dominant tubulointerstitial kidney disease (ADTKD), particularly involving UMOD and HNF1-β mutations [3].

Unfortunately, we could not perform the genetic study in our patient. However, multiple case reports of sporadic glomerulocystic kidney disease from the Indian subcontinent have indicated that the genetic workup has been negative [4, 5].

## Conclusion

Sporadic glomerulocystic kidney disease can manifest as renal failure during the newborn period. In a resource-limited setting, a renal biopsy may be used to diagnose glomerulocystic kidney disease.

## Ethical Considerations

### Compliance with ethical guidelines

There were no ethical considerations to be considered in this research.

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### Authors' contributions

All authors contributed equally to the conception and design of the study, data collection and analysis, interception of the results and drafting of the manuscript. Each author approved the final version of the manuscript for submission.

### Conflict of interest

The authors declared no conflict of interest.

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