Renographic Evaluation with ^{99m}Tc-DTPA in Bilateral Nephrolithiasis with Cystinuria: A Rare Case Report

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ABSTRACT

Cystinuria is a rare inherited disorder leading to recurrent nephrolithiasis and potential renal impairment. This report presents the case of a 9-year-old female with elevated urinary cystine, with normal serum biochemical parameters. Ultrasound showed mild hydronephrosis due to bilateral staghorn calculi while intravenous urography demonstrated preserved excretory function. A 99mTc DTPA renogram revealed pelvicalyceal dilatation without obstruction and mild functional impairment of the left kidney, with normal right renal function. The patient underwent left open pyelolithotomy with DJ stent placement. This case highlights the importance of DTPA renogram in detecting early renal dysfunction, differentiating obstruction, and guiding management in pediatric cystinuria to preserve renal function.

Keywords: Cystinuria, pediatric nephrolithiasis, DTPA renogram, renal function.

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INTRODUCTION

Cystinuria is a rare autosomal recessive metabolic disorder caused by defective reabsorption of cystine and dibasic amino acids (lysine, ornithine, and arginine) in the renal proximal tubules. It accounts for 6–8% of pediatric urolithiasis and often presents with recurrent nephrolithiasis. In many cases, stones are detected during adolescence, with increased risk of hypertension, recurrent obstructive stones, and chronic kidney disease (CKD). Cystinuria leads to recurrent stone formation and over time, it can compromise the renal function. The 99mTc DTPA renogram is an indispensable tool in medical diagnostics which can provide quantitative data about the glomerular filtration rate (GFR), assess renal function, detect obstruction, evaluate perfusion, assist with surgical planning, and monitor long-term kidney health.

CASE SUMMARY

The reported case presented with bilateral nephrolithiasis in a 9-year-old female with early-onset cystinuria. She was referred to the Institute of Nuclear Medicine and Allied Sciences (INMAS), Mohakhali, in November 2024 for renal function evaluation using a 99mTc DTPA renogram. She reported a two-year history of recurrent fever and loin pain, with a positive family history for recurrent nephrolithiasis. Her general and per abdominal examination was unremarkable. Ultrasound showed bilateral staghorn calculi with mild hydronephrosis in the left kidney (figure 1). Intravenous urography demonstrated normal excretory function of both kidneys. Serum creatinine, calcium, and parathyroid hormone levels were within normal limits (S. creatinine - 0.66 mg /dl, S. calcium - 10.1 mg/dl, Spot urine calcium- 1.1 mg/dl, S. PTH -32 Pg/mL). Urine microscopy showed pus cells(6-10/HPF) and some RBCs, with no bacterial growth on culture. Urinary cystine levels were elevated (189 mg/dl in 24-hour urine collection). The DTPA renogram indicated pelvicalyceal dilatation without obstruction and mild parenchymal impairment in the left kidney, with normal function in the right kidney (figure 2). The patient underwent left open pyelolithotomy with DJ stent placement.

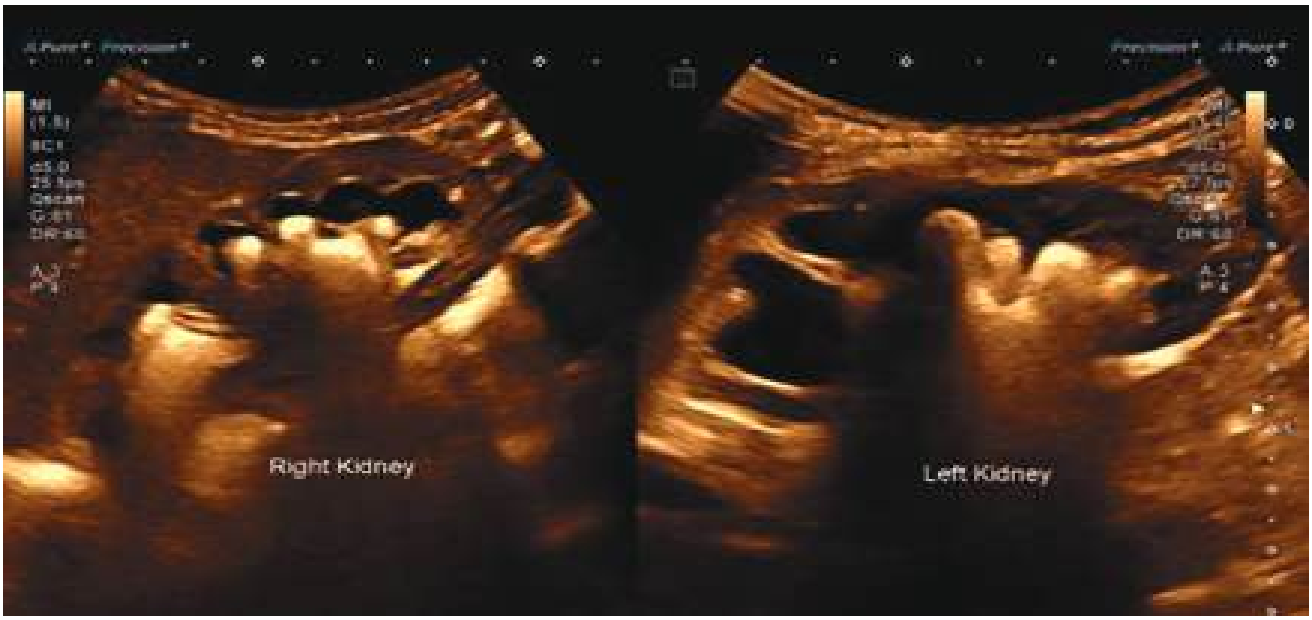

Figure 1: Ultrasound image showing bilateral staghorn calculi.

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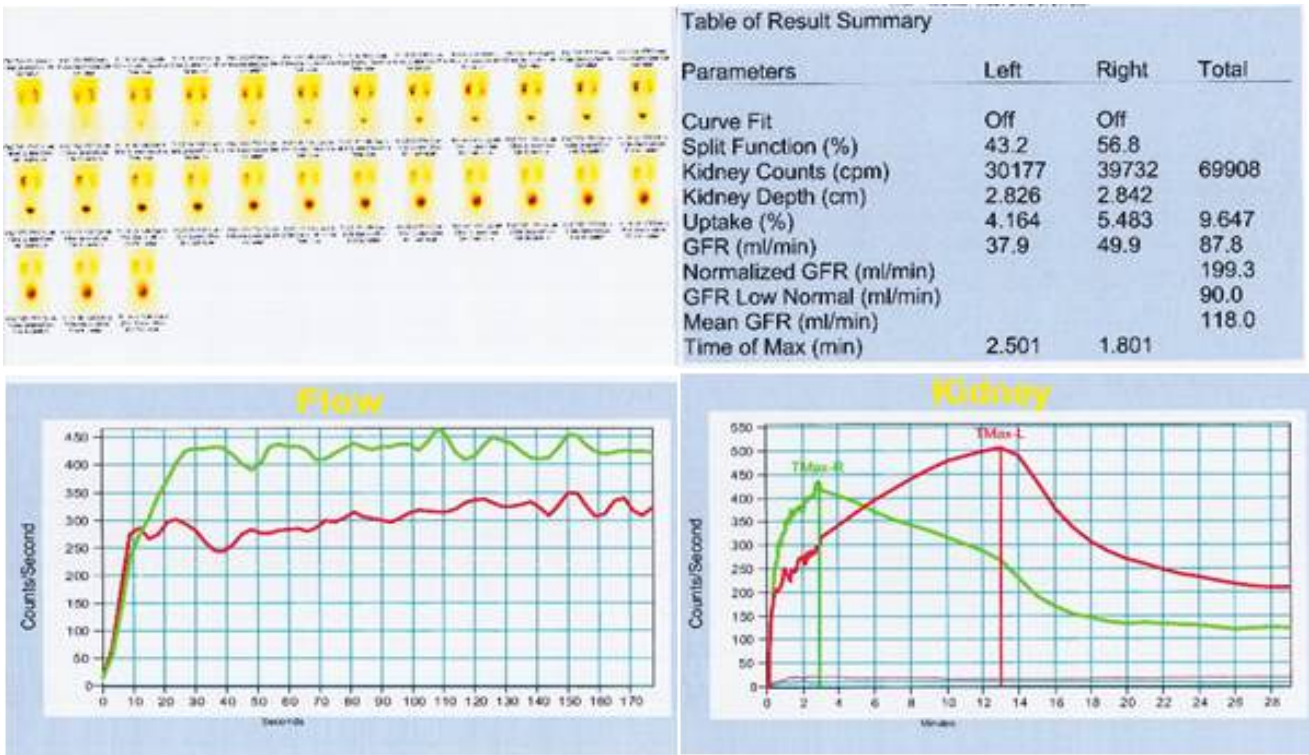

Table of Result Summary

Parameters	Left	Right	Total
Curve Fit	Off	Off	
Split Function (%)	43.2	56.8	
Kidney Counts (cpm)	30177	39732	69908
Kidney Depth (cm)	2.826	2.842	
Uptake (%)	4.164	5.483	9.647
GFR (ml/min)	37.9	49.9	87.8
Normalized GFR (ml/min)			199.3
GFR Low Normal (ml/min)			90.0
Mean GFR (ml/min)			118.0
Time of Max (min)	2.501	1.801	

Figure 2: Graphs and images of 99mTc DTPA renogram of the same patient showing non-obstructive dilatation in left kidney with mild parenchymal impairment. Time activity curve and quantitative parameters of the right kidney are within normal limit.

Figure 3: Cystine stone after pyelolithotomy

DISCUSSION

Cystinuria is a rare autosomal recessive disorder characterized by defective renal tubular reabsorption of cystine and dibasic amino acids, leading to persistent hyperexcretion of cystine and recurrent nephrolithiasis. Although it accounts for a small proportion of overall urinary stone disease, it represents a significant cause of pediatric urolithiasis, particularly in cases with early onset and recurrent stone formation (1). The present case illustrates a typical but clinically significant presentation of cystinuria in a child, complicated by bilateral staghorn calculi and early functional renal changes.

The early age of presentation in this patient is consistent with previous studies, which indicate that cystinuria often manifests during childhood or adolescence with recurrent flank pain, urinary symptoms, and stone recurrence (2). The positive family history in this case further supports the inherited nature of the disorder, commonly associated with mutations in the SLC3A1 and SLC7A9 genes (3). Early identification of such cases is crucial, as recurrent stone formation can lead to progressive renal damage, hypertension, and eventual chronic kidney disease if not appropriately managed (4).

A striking feature of this case is the presence of bilateral staghorn calculi, which are relatively uncommon in pediatric patients but may occur in severe or untreated cystinuria. These large, branching calculi can occupy the renal pelvis and calyces, predisposing patients to obstruction, infection, and progressive loss of renal function (5). Despite the significant stone burden, this patient maintained normal serum creatinine and preserved excretory function on intravenous urography, suggesting that biochemical markers alone may not adequately reflect early functional impairment.

In this context, the role of ^{99m}Tc-DTPA renogram becomes particularly critical. Unlike conventional imaging modalities that primarily assess anatomical abnormalities, DTPA renogram provides dynamic and quantitative evaluation of renal perfusion, function, and drainage. In the present case, the renogram revealed pelvicalyceal dilatation without significant obstruction and mild parenchymal impairment in the left kidney, while the right kidney demonstrated preserved function. This functional insight is of paramount importance, as it allows detection of early renal compromise that may not be apparent on structural imaging or routine biochemical tests (6,7).

The discrepancy observed between ultrasound findings (hydronephrosis) and renography results (non-obstructive drainage) highlights the complementary role of functional imaging. Hydronephrosis does not always indicate true obstruction; DTPA renography helps differentiate between obstructive and non-obstructive dilatation, thereby preventing unnecessary or inappropriate interventions (8). In this patient, the absence of functional obstruction guided a more targeted surgical approach rather than urgent decompression, demonstrating how renography directly influences clinical decision-making.

Furthermore, DTPA renogram provides valuable information on split renal function, which is essential in planning surgical management, especially in bilateral disease. Preservation of adequate function in the affected kidney supports nephron-sparing procedures, as performed in this case with left open pyelolithotomy and DJ stent placement. In addition, baseline renography assessment allows for longitudinal monitoring of renal function, enabling early detection of deterioration and guiding long-term management strategies (9).

Biochemically, the markedly elevated urinary cystine level confirms the diagnosis of cystinuria, while normal serum calcium and parathyroid hormone levels exclude other metabolic causes of nephrolithiasis. The presence of sterile pyuria and microscopic hematuria is consistent with chronic mucosal irritation from stones rather than infection, a common finding in cystine stone disease (10).

Management of cystinuria is multifaceted, involving high fluid intake, urinary alkalinization, and pharmacological therapy to reduce cystine concentration. However, in cases with large or complex stones, surgical intervention becomes necessary. In such scenarios, functional imaging with DTPA renogram plays a central role not only in preoperative assessment but also in postoperative follow-up, ensuring preservation of renal function and early identification of complications (11).

Overall, this case underscores the importance of a comprehensive diagnostic approach in pediatric cystinuria, integrating biochemical evaluation, anatomical imaging, and functional assessment. Among these, 99mTc DTPA renography emerges as an indispensable tool, providing critical information on renal function, drainage, and parenchymal integrity. Its role is particularly vital in guiding management decisions, optimizing surgical planning, and preventing long-term renal sequelae in children with complex urolithiasis.

CONCLUSION

This case highlights the importance of ^{99m}Tc DTPA renogram in pediatric cystinuria with complex nephrolithiasis. It enables early detection of renal functional impairment, differentiates obstructive from non-obstructive dilatation, and guides appropriate management. Incorporating functional imaging alongside routine evaluation is essential for preserving renal function and improving outcomes.

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