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Pyelectasis dilatation associated with posterior urethral valves: A case report

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Abstract

Introduction: Posterior urethral valves (PUV) represent the most common cause of congenital lower urinary tract obstruction (LUTO) in male fetuses. Prenatal diagnosis is crucial for early prognostic evaluation and postnatal management planning. Pyelectasis and the "keyhole sign" are key sonographic features suggesting this anomaly.

Methods: We report the case of a male fetus diagnosed at 21 weeks of gestation with bilateral pyelectasis and signs of bladder outlet obstruction. Serial ultrasounds were performed, and postnatal confirmation was obtained via imaging and surgical intervention.

Results: Ultrasound at 21 weeks revealed bilateral renal pelvis dilatation, thickened bladder wall, and a keyhole sign. Follow-up showed mild oligohydramnios and increased renal echogenicity. Postnatal imaging confirmed PUV, and endoscopic valve ablation was successfully performed. Renal function stabilized with close follow-up.

Conclusion: This case underscores the importance of recognizing prenatal indicators of posterior urethral valves, such as pyelectasis and keyhole sign. Early detection enables appropriate counseling, neonatal preparation, and timely postnatal intervention to preserve renal function.

Keywords: Posterior urethral valves, congenital lower urinary tract obstruction, prenatal diagnosis

Introduction

Posterior urethral valves (PUV) are congenital obstructing membranous folds within the posterior urethra and are the most frequent cause of lower urinary tract obstruction (LUTO) in male fetuses. This condition affects approximately 1 in 5,000 to 8,000 live births and carries significant risk for long-term renal impairment or end-stage renal disease if left untreated.

Prenatal diagnosis relies primarily on ultrasound findings, including bladder distension, thickened bladder walls, bilateral hydronephrosis, dilated posterior urethra (keyhole sign), and in advanced cases, oligohydramnios. Early identification allows for prenatal surveillance, parental counseling, and planning for neonatal intervention.

We present a case of prenatally diagnosed PUV, initially detected by bilateral pyelectasis and keyhole sign, which led to timely postnatal management and a favorable outcome.

Case Presentation

A 29-year-old woman, gravida 2 para 1, was referred to our fetal medicine unit at 21 weeks of gestation following an anomaly scan that revealed bilateral renal pelvis dilatation. The initial ultrasound showed renal pelvis measurements of 0.37 mm on the right and 0.45 mm on the left, with normal kidney size and corticomedullary differentiation. The urinary bladder appeared significantly distended with a thickened wall, and a dilated posterior urethra created the classic "keyhole sign," highly suggestive of a lower urinary tract obstruction.

No other structural anomalies were noted. The amniotic fluid index was within normal limits, and fetal biometry was appropriate for gestational age. Fetal sex was identified as male.

A targeted fetal urinary tract ultrasound confirmed the findings: bilateral hydronephrosis, distended bladder, and a narrowed posterior urethra forming the keyhole configuration. The ureters were slightly dilated. The echogenicity of the kidneys was within normal limits at this stage, with preserved corticomedullary differentiation.

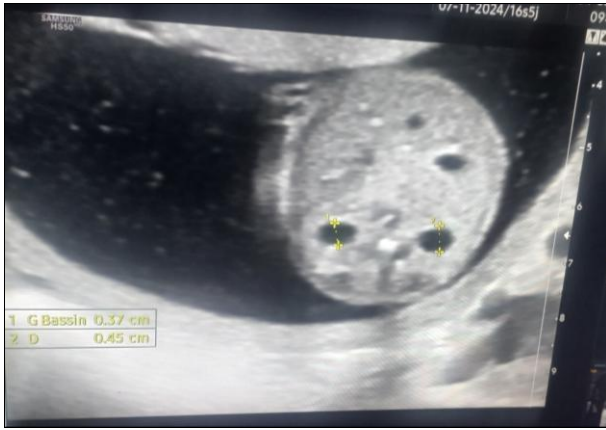


Fig 1: Prenatal ultrasound showing bilateral pelvic dilatation (pyelectasis) with preserved corticomedullary differentiation



Fig 2: Sagittal ultrasound view of the fetal pelvis showing a dilated posterior urethra forming the characteristic "keyhole sign", suggestive of posterior urethral valves

TORCH screening was negative, and there was no maternal history of diabetes or drug exposure. Given the suspected diagnosis of posterior urethral valves, serial ultrasounds were scheduled to monitor the progression of urinary tract dilatation and amniotic fluid levels.

At 28 weeks of gestation, mild oligohydramnios developed, and the renal parenchyma appeared slightly hyperechogenic, raising concerns for evolving renal injury. Despite this, the pregnancy remained stable, and no fetal interventions were performed. The pregnancy continued to term without complications.

The infant was delivered at 38 weeks via spontaneous vaginal delivery. Birth weight and Apgar scores were within normal limits. Soon after birth, the neonate developed lower abdominal distension and had difficulty urinating. Postnatal renal ultrasonography confirmed bilateral hydronephrosis, a thick-walled bladder, and posterior urethral dilatation. A voiding cystourethrogram (VCUG) demonstrated obstruction of the posterior urethra consistent with posterior urethral valves.

An endoscopic valve ablation was performed successfully within the first week of life. Initial serum creatinine was mildly elevated but gradually improved with good urinary output. The infant was discharged in stable condition under pediatric nephrology follow-up, with normalizing renal function over the first three months of life.

Discussion

Posterior urethral valves (PUV) represent the most common cause of congenital bladder outlet obstruction in male neonates, with an estimated incidence of 1 in 5,000–8,000 live births [1].

This condition arises from abnormal persistent membranous folds (Type I PUV, per Young's classification) in the posterior urethra, causing functional obstruction, elevated intravesical pressure, and secondary upper urinary tract dilation [2].

Prenatal Diagnosis & Sonographic Features

Prenatal ultrasound remains the cornerstone for early detection, with characteristic findings including:

- Bilateral hydronephrosis/pyelectasis (90% of cases) [3].
- Distended bladder with bladder wall thickening (>2 mm, due to detrusor hypertrophy) [4].
- Keyhole sign (dilation of the posterior urethra and bladder) – pathognomonic but not always present [5].
- Oligohydramnios (in severe cases, reflecting low fetal urine output and renal dysfunction) [6].

In this case, the second-trimester detection of progressive urinary tract dilation allowed for timely intervention. Notably, the absence of oligohydramnios suggested preserved renal function, a favorable prognostic marker [7].

Pathophysiological Consequences

The "valve-bladder" syndrome leads to:

1. Upper tract damage

- Hydroureteronephrosis from chronic reflux / obstruction.
- Renal dysplasia due to prolonged intrauterine pressure (evidenced by cortical cysts or echogenic kidneys) [8].

2. Pulmonary complications: Oligohydramnios-associated pulmonary hypoplasia (major cause of neonatal mortality) [9].

3. Bladder dysfunction: Poor compliance from detrusor hypertrophy, persisting post-ablation in 30–50% of cases [10].

Postnatal Management & Outcomes

- **First-line treatment:** Endoscopic valve ablation (fulguration), ideally within 48 hours of life to minimize renal injury [11].
- **Temporary measures:** Vesicostomy or upper tract diversion if endoscopic access is challenging [12].
- **Multidisciplinary care**
 - Pediatric urology (surgical correction).
 - Nephrology (monitoring for CKD, seen in 30–60% long-term) [13].
 - Neonatology (respiratory support if pulmonary hypoplasia exists) [14].

Prognostic Determinants

- **Favorable factors**
 - Normal amniotic fluid volume (indicating preserved renal function) [15].
 - Early detection + intervention (as in this case) [16].
- **Poor prognostic markers:** Oligohydramnios, renal cortical cysts, or serum creatinine >1.0 mg/dL at 1 month [17].

Long-term follow-up is critical, as 25–40% of PUV patients progress to ESRD by adolescence despite successful ablation [18]. Regular monitoring of renal function, bladder dynamics, and growth is essential [19].

Conclusion

This case highlights the importance of early prenatal detection of posterior urethral valves through careful ultrasound assessment. Recognition of bilateral pyelectasis and the keyhole sign should prompt close monitoring and postnatal planning. Prompt

diagnosis and surgical intervention after birth can prevent further renal deterioration and allow for normal growth and development. Prenatal ultrasound remains an invaluable tool in identifying such life-altering congenital anomalies and guiding perinatal decision-making.

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