ORIGINAL RESEARCH ARTICLE

Diponegoro International Medical Journal 2026 July, Vol 7, No.1: 31-34
e-ISSN: 2745-5815

Prenatal Diagnosis of Fetal Lower Urinary Tract Obstruction with Posterior Urethral Valve: A Case Report

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Keywords:

Fetal lower urinary tract obstruction (LUTO)
Posterior urethral valve (PUV)
Prenatal ultrasound
Megacystis
Keyhole sign
Anhydramnios Fetal urology

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Article history:

Received 23-09-2025
Accepted 22 -12-2025
Available online 30-01-2026

ABSTRACT

Background: Fetal lower urinary tract obstruction (LUTO) is an uncommon but serious congenital anomaly that may result in progressive renal damage, pulmonary hypoplasia, and increased perinatal mortality. Posterior urethral valve (PUV) is the most common cause of LUTO in male fetuses. Prenatal ultrasonography plays a pivotal role in the early recognition of characteristic imaging findings, enabling timely counseling and management planning.
Case Presentation: A 26-year-old gravida 2 para 1 woman at 24–25 weeks' gestation was referred for evaluation of a suspected fetal urinary tract anomaly. Prenatal ultrasound demonstrated a single breech fetus with marked bladder distension (megacystis), bilateral hydronephrosis, severe anhydramnios, and a characteristic keyhole sign, findings highly suggestive of LUTO secondary to posterior urethral valve. After multidisciplinary evaluation, conservative management was selected considering the gestational age and overall clinical condition, and the patient received counseling regarding the anticipated prognosis and planned postnatal evaluation.
Conclusion: This case highlights the pivotal role of prenatal ultrasonography in recognizing fetal LUTO and identifying imaging features highly suggestive of posterior urethral valve. Early prenatal diagnosis facilitates appropriate parental counseling, individualized perinatal management, multidisciplinary planning, and preparation for postnatal evaluation and treatment.
DIMJ, 2026, 7(1), 31-34 DOI: https://doi.org/10.14710/dimj.v7i1.27429

1. Introduction

Fetal lower urinary tract obstruction (LUTO) comprises a heterogeneous group of congenital disorders characterized by partial or complete obstruction of the fetal urethra, resulting in impaired urinary outflow. Persistent obstruction may lead to progressive bladder distension, bilateral hydroureteronephrosis, renal dysplasia, oligohydramnios or anhydramnios, pulmonary hypoplasia, and ultimately increased perinatal morbidity and mortality. Reported incidence ranges from approximately 1 in 5,000 to 1 in 25,000 pregnancies; however, the true incidence is likely underestimated because severe cases may result in spontaneous intrauterine fetal demise or elective termination following prenatal diagnosis, thereby limitingZaccurate population-based estimates.[1–3]

Posterior urethral valve (PUV) represents the most common cause of fetal LUTO and occurs almost exclusively in male fetuses. The obstructing membranous folds within the posterior urethra increase intravesical pressure, resulting in progressive bladder enlargement, upper urinary tract dilatation, and impaired renal development. The severity of renal injury depends on both the degree and duration of obstruction, making early prenatal diagnosis crucial for prognostic assessment and clinical decision-making.[2–5]

Prenatal ultrasonography remains the first-line imaging modality for evaluating suspected fetal LUTO because of its widespread availability, real-time capability, and excellent depiction of fetal urinary tract anatomy. Typical sonographic findings include megacystis, bilateral hydronephrosis, hydroureter, bladder wall thickening, oligohydramnios or anhydramnios, and the characteristic keyhole sign caused by dilatation of the bladder neck and proximal urethra.

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Although the keyhole sign is highly suggestive of posterior urethral valve, it is not pathognomonic and may also be encountered in urethral atresia, urethral stenosis, or megacystis–microcolon–intestinal hypoperistalsis syndrome (MMIHS). Therefore, comprehensive sonographic evaluation and careful differential diagnosis remain essential.[2,4,6]

Fetal magnetic resonance imaging (MRI) may serve as a complementary imaging modality in selected cases, particularly when ultrasonographic evaluation is limited by severe oligohydramnios, maternal obesity, or unfavorable fetal position. In addition, prenatal assessment aims not only to establish the diagnosis but also to identify fetuses who may benefit from fetal interventions such as vesicoamniotic shunting or fetal cystoscopy. Nevertheless, patient selection remains highly individualized because the benefits of prenatal intervention depend largely on gestational age, residual renal function, and overall fetal prognosis.[6,7] Herein, we report a prenatally diagnosed case of fetal LUTO highly suggestive of posterior urethral valve, characterized by megacystis, bilateral hydronephrosis, severe anhydramnios, and the keyhole sign. This report highlights the importance of systematic prenatal imaging, appropriate differential diagnosis, multidisciplinary counseling, and individualized management in optimizing prenatal and postnatal care.

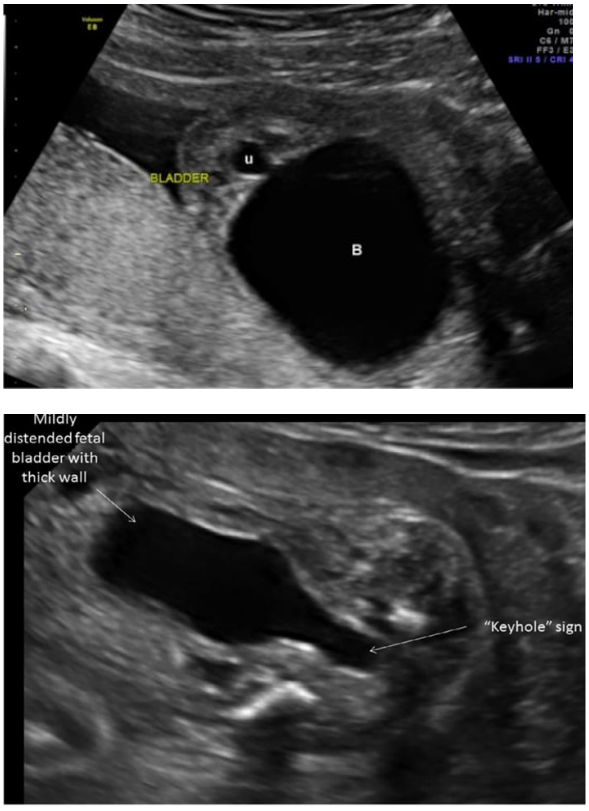


Figure 1. Ultrasound imaging of the posterior urethra typically reveals a 'keyhole' sign.[2,8]

2. Case Presentation

A 26-year-old gravida 2 para 1 woman at 24–25 weeks of gestation was referred from a secondary referral hospital for further evaluation of a suspected fetal urinary tract anomaly detected on routine antenatal ultrasonography. Her previous pregnancy had been delivered by cesarean section in 2023 because of cephalopelvic disproportion and premature rupture of membranes. She had no history of chronic medical illness, congenital anomalies, teratogenic exposure, or hereditary renal disease. Antenatal care had been performed regularly at primary healthcare facilities and by an obstetrician without previous reports of fetal abnormalities.

The estimated date of delivery was 11 August 2025, calculated from the last menstrual period on 11 November 2024. Maternal physical examination revealed stable vital signs, and fetal heart rate was 149 beats/minute. Laboratory investigations were unremarkable except for mild anemia with a hemoglobin concentration of 10.5 g/dL.

A detailed prenatal ultrasonographic examination performed on 23 April 2025 demonstrated a singleton fetus in breech presentation with an estimated fetal weight of 573 g, corresponding to 24 weeks and 2 days of gestation. Ultrasonography demonstrated marked bladder distension (megacystis) measuring approximately 5.1 × 2.3 cm, associated with bilateral hydronephrosis and severe reduction of amniotic fluid (amniotic fluid index 1.43 cm), consistent with anhydramnios (Figure 2). In addition, dilatation of the bladder neck and proximal urethra produced the characteristic keyhole sign, a finding highly suggestive of posterior urethral valve (Figure 3).

Detailed assessment of bladder wall thickness, renal cortical echogenicity, corticomedullary differentiation, and hydronephrosis grading according to the Society for Fetal Urology (SFU) or Urinary Tract Dilation (UTD) classification was not routinely documented during the examination, representing a limitation of this case report.

Based on the combination of megacystis, bilateral hydronephrosis, severe anhydramnios, and the keyhole sign, a prenatal diagnosis of fetal lower urinary tract obstruction highly suggestive of posterior urethral valve was established.

Because the ultrasonographic findings were considered sufficiently characteristic to establish the presumptive diagnosis, additional fetal magnetic resonance imaging (MRI) was not expected to provide information that would alter immediate clinical management.

After multidisciplinary consultation involving maternal-fetal medicine specialists and radiologists, expectant (conservative) management was selected. The decision was based on the gestational age at presentation, severe anhydramnios indicating advanced urinary tract obstruction, and careful consideration of the anticipated benefits and limitations of prenatal intervention. The patient received comprehensive counseling regarding fetal prognosis, the possibility of pulmonary hypoplasia and renal impairment,

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and the need for delivery at a tertiary referral center with access to neonatal intensive care and pediatric urology services.

Following delivery, the planned neonatal evaluation included renal ultrasonography, serum renal function assessment, voiding cystourethrography (VCUG), and consultation with a pediatric urologist to confirm the diagnosis and determine the need for definitive surgical management.

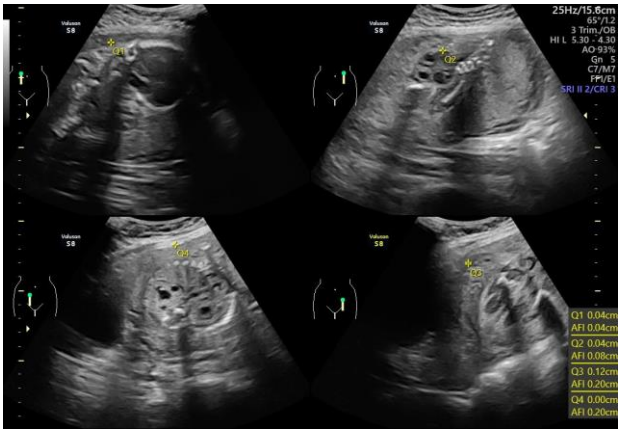


Figure 2. Ultrasound images demonstrating bilateral hydronephrosis



Figure 3. Megacystis with "Key hole sign".

3. Discuscion

Fetal lower urinary tract obstruction (LUTO) represents a spectrum of congenital abnormalities that impede fetal urinary outflow and may result in progressive bladder distension, hydroureteronephrosis, renal dysplasia, oligohydramnios, pulmonary hypoplasia, and ultimately chronic kidney disease or perinatal death. Posterior urethral valve (PUV) is the most common cause of LUTO and occurs almost exclusively in male fetuses. In the present case, the combination of a male fetus, megacystis, bilateral hydronephrosis, severe anhydramnios, and the keyhole sign strongly supported the prenatal diagnosis of LUTO secondary to PUV.[1–3]

The pathophysiological consequences of PUV depend on both the severity and duration of urinary tract obstruction. Progressive elevation of intravesical pressure causes bladder wall hypertrophy and impaired bladder compliance, while retrograde pressure transmission to the upper urinary tract leads to hydronephrosis, renal tubular injury, nephron loss, and renal dysplasia. Reduced fetal urine production subsequently causes oligohydramnios or anhydramnios, increasing the risk of pulmonary hypoplasia, which remains one of the principal causes of neonatal mortality in severe LUTO.[2,4]

Prenatal ultrasonography remains the primary imaging modality for evaluating fetal urinary tract obstruction because it provides real-time assessment of fetal anatomy, urinary tract morphology, and amniotic fluid volume. In this patient, ultrasonography demonstrated megacystis, bilateral hydronephrosis, severe anhydramnios, and the keyhole sign. Although the keyhole sign is classically associated with PUV, current evidence indicates that it should be regarded as highly suggestive rather than pathognomonic, as similar appearances may occur in urethral atresia, urethral stenosis, and megacystis–microcolon–intestinal hypoperistalsis syndrome (MMIHS). Therefore, diagnosis should always integrate the complete sonographic findings and clinical context rather than relying on a single imaging sign.2,5

The principal differential diagnoses in this case included urethral atresia, prune belly syndrome, and MMIHS. Urethral atresia usually presents with severe bladder distension but often lacks the characteristic proximal urethral dilatation seen in PUV. Prune belly syndrome is characterized by deficient abdominal wall musculature, bilateral cryptorchidism, and urinary tract abnormalities, findings that were not identified prenatally in this fetus. MMIHS predominantly affects female fetuses and is associated with bowel dilatation and intestinal hypoperistalsis, features that were absent in the present case. Collectively, these considerations made PUV the most likely diagnosis.

Several prenatal ultrasonographic parameters have prognostic value in fetuses with LUTO, including the degree of hydronephrosis, bladder wall thickening, renal cortical echogenicity, corticomedullary differentiation, renal cortical cysts, and amniotic fluid volume. Severe anhydramnios in the present case suggested longstanding urinary tract obstruction and raised concern for impaired renal function and pulmonary hypoplasia. Although bladder wall thickness, renal cortical echogenicity, and standardized hydronephrosis grading were not systematically documented, this limitation did not preclude recognition of the characteristic constellation of findings supporting the diagnosis.

Fetal magnetic resonance imaging (MRI) may provide complementary anatomical information when ultrasonographic evaluation is limited by maternal obesity, fetal position, or severe oligohydramnios. MRI also facilitates assessment of associated anomalies and may assist in planning fetal intervention. However, in the present case, ultrasonography adequately demonstrated the characteristic features of LUTO, and additional MRI was unlikely to alter either the prenatal diagnosis or the immediate management strategy.

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risks, and their long-term renal benefits remain uncertain. Appropriate candidate selection depends on gestational age, estimated residual renal function, associated anomalies, and multidisciplinary evaluation. In the present case, expectant management was selected because the fetus presented at 24–25 weeks' gestation with severe anhydramnios suggestive of advanced urinary tract obstruction, making the anticipated benefit of prenatal intervention limited.

Prenatal counseling constitutes an essential component of management. Parents should be informed regarding the potential risks of pulmonary hypoplasia, neonatal respiratory insufficiency, chronic kidney disease, bladder dysfunction, and the possible need for long-term pediatric urological follow-up. Following delivery, postnatal confirmation with renal ultrasonography, voiding cystourethrography (VCUG), renal function assessment, and pediatric urology consultation is recommended to establish the definitive diagnosis and determine the optimal timing of valve ablation. The principal strength of this report lies in the clear demonstration of the characteristic prenatal ultrasonographic findings of LUTO secondary to suspected PUV. However, several limitations should be acknowledged. First, detailed prenatal assessment of bladder wall thickness, renal cortical echogenicity, and standardized hydronephrosis grading was unavailable. Second, postnatal imaging confirmation and long-term renal outcomes were not yet available at the time of manuscript preparation. These limitations should be considered when interpreting the findings.

This case emphasizes the importance of systematic prenatal ultrasonography, comprehensive differential diagnosis, multidisciplinary counseling, and individualized management in fetuses with suspected LUTO. Early recognition of characteristic imaging findings facilitates appropriate parental counseling, delivery planning, and timely postnatal evaluation, all of which are essential for optimizing neonatal outcomes.

4. Conclusion

Fetal lower urinary tract obstruction (LUTO), particularly when caused by posterior urethral valve (PUV), is a serious congenital condition associated with significant perinatal morbidity and mortality. Prenatal ultrasonography plays a pivotal role in the early recognition of characteristic findings, including megacystis, bilateral hydronephrosis, severe anhydramnios, and the keyhole sign. However, because the keyhole sign is highly suggestive rather than pathognomonic, careful evaluation of differential diagnoses and comprehensive sonographic assessment remain essential.

Early prenatal diagnosis facilitates timely parental counseling, multidisciplinary decision-making, individualized management, and appropriate preparation for postnatal evaluation and definitive treatment. This case highlights the importance of integrating characteristic prenatal imaging findings with clinical judgment to optimize both prenatal care and neonatal outcomes.

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