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Posterior Urethral Valve in Triamniotic Trichorionic Triplets, a Case Report

Noor Alfradan, MD[1*], Omar Taso MD[2], Areeg Besharat MD[3], Yusuf Yusuf MD[3]

[1]Medical Intern, Department of Obstetrics and Gynecology, King Hussein Medical Center Amman 11831 Jordan
[2]Fetal medicine specialist, Department of Obstetrics and Gynecology, King Hussein Medical Center Amman 11831 Jordan
[3]Neontology specialist, Department of Pediatrics, King Hussein Medical Center Amman 11831 Jordan

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***Corresponding author:** Noor Alfradan, MD
Medical Intern, Department of Obstetrics and Gynecology, King Hussein Medical Center Amman 11831 Jordan

Abstract **Case Report**

Posterior urethral valves (PUV) incresingly identified before birth, displays a range of severity levels. The extent of obstruction resulting from this anomaly hinges on the specific structure of the obstructive membrane valve within the urethra. While posterior urethral valves are congenital, their exact embryological origin provoke ongoing discussion and lacks consensus. In fetuses diagnosed with PUV early in pregnancy with evidence of reduced amniotic fluid volume along with declining renal function due to urine aspiration, antenatal intervention such as vesicoamniotic shunting can improve postnatal renal function. We present a case involving triamniotic trichorionic triplets conceived via in vitro fertilization (IVF) and preimplantation genetic testing for aneuploidy (PGT-A). Identified via antenatal ultrasound screening, these triplets demonstrate diverse degrees of severity and outcomes. This case highlights the significance of early diagnosis and prompt identifycation of such conditions to reduce complexity and enhance outcomes.

Keywords: Posterior urethral valve, PUV, triamniotic, trichorionic, triplets, IVF.

INTRODUCATION

Posterior urethral valves (PUV) represent a congenital blockage within the posterior urethra. It forms when the Wolffian ducts fail to integrate properly into the urethra, leading to the development of a membrane that obstructs the lower urinary tract. The development of the male urethra typically is completed by week 14 of gestation [1]. As this condition arises early in gestation, it subjects the bladder and upper urinary tract to increased pressure and backflow reflux during development, consequently inducing significant alterations in bladder function and resulting in hydronephrosis and renal injury eventually. Low urine output during fetal development can lead to oligohydramnios in which fetus may experience compression of the chest cavity, limiting lung expansion and development. This compression can result in pulmonary hypoplasia.

The specific incidence of posterior urethral valve (PUV) obstruction among triplets is not readily available in the literature. Incidence rates for PUV itself vary, with estimates typically provided for singleton pregnancies and among siblings rather than multiples. In most cases, suspicions of posterior urethral valves (PUVs) arise from antenatal ultrasound examinations, with the possibility of detecting prenatal signs of PUVs as early as the 13th week of gestation [2]. Close monitoring through antenatal ultrasound screenings can help identify signs of PUV obstruction early on and guide subsequent management decisions to decrease mortality and morbidity.

We report a detailed and comprehensive description of a unique case to the collective knowledge of posterior urethral obstruction in triamniotic trichornionic tripltes, a product of IVF and sex selection. This manuscript was prepared following the CARE guidelines (https://www.care-statement.org).

CASE PRESENTATION

At 23 weeks of gestation, the 36-year-old G4P300 woman was referred to the unit for shunting after confirmed diagnosis of PUV has been made during a routine antenatal visit in a private clinic. Further detailed ultrasonic imaging revealed findings consistent with complete posterior urethral valve obstruction in two of the three fetuses and incomplete PUV in the third fetus. Family history was negative for any urogenital anomolies or renal diseases and the mother had no

complaints during this fourth preganancy. Karyotyping showed no abnormalities.

Diagnostic Assessment

Ultrasound scanning showed Triplet 1, with a maternal right lower posterior placenta, is male and exhibits normal growth with no liquor. However, severe bilateral hydronephrosis with hydroureter on the left side was observed. Additionally, there is a large bladder with a thick wall and a keyhole shape, indicating the presence of posterior urethral valve and lower urinary tract obstruction (LUTO). Triplet 2, positioned in the middle upper with an anterior placenta, is a male and exhibits moderate bilateral hydronephrosis. There were no hydroureters observed, but an enlarged bladder without a keyhole shape suggesting partial obstruction. Adequate liquor was present. Triplet 3, positioned in the maternal left lower quadrant, is a male and the leading one. He had a posterior placenta and showed normal growth, but presents with anhydramnios and severe lower urinary tract obstruction (LUTO) similar to Triplet 1.

The cervical length measures 23 mm, and the mother was experiencing mild lower abdominal pain. Consequently, the option of vesicoamniotic shunting or any other invasive procedure was deemed impractical due to the risk of precipitating preterm labor. Termination of pregnancy was proposed due to suspected poor outcome, however it was refused by the parents due to religious and personal believes.

The pregnancy was closely monitored until the 28th week and 5 days, at which point the mother went into labor. An emergency cesarean section was performed, resulting in the birth of three live babies. The triplets required mechanical ventilation for pulmonary hypoplasia. Unfortunately, triplets 1 and 3 passed away 2 and 4 hours after birth, while triplet 2 survived.

Follow-up and Outcomes

The live fetus was born with features of pulmonary hypoplasia, he needed mechanical ventilation for two days. He made a smooth recovery and was discharged after one month of monitory in the NICU. Renal ultrasound, serum chemstry and voiding cystourethrography done during this period deemed satisfactory. Following discharge, the patient's management plan included following up with serial renal ultrasound every 6 months and prophylaxis antibiotics.

DISCUSSION

The prevelance of PUV is estimated to be 1 in 5000 to 8000 male births [3]. Yet the actual incidence of PUVs is not precisely known, partly due to the variable pathological spectrum and the possibility of asymptomatic cases [4]. And as litereture describes familial occurrence and multiple case reports of PUVs in monozygotic twins and siblings suggest a multifactorial etiology, where both genetic predisposition and environmental factors play a role, the complex phenotypic presentation and discordance in triamniotic trichorionic tripltes validates this theory [5].

CONCLUSION

The case of triamniotic trichorionic triplets with posterior urethral valve (PUV) obstruction presents a complex clinical scenario involving multiple fetuses affected by varying degrees of urinary tract obstruction. In this case, detailed ultrasonographic imaging revealed complete PUV obstruction in two of the triplets and incomplete obstruction in the third. This case highlights the complexity and challenges associated with this condition. While familial occurrence and genetic predisposition suggest a partial genetic basis, the variable phenotypic expression and discordance in non-identical triplets underscore the multifactorial nature of PUVs.

Based on animal models and the new understanding of the effect of obstruction on nephrogenesis, it is likely that renal damage in congenital bladder outlet obstruction occurs as early as 14 weeks [6]. Moreover renal damage is significant and irreversible in as little as days [7]. Maintaining a high index of suspicion during this crucial period to achieve early detection and prompt intervention is challenging, particularly considering that current screening recommendations commence four to six weeks after this developmental stage [6].

Decreasing the morbidity and mortality of conegintal posterior urethral valves continues to pose significant challenges in fetal medicine. Because of the high complications rates of available antenatal interventions and the poor long term outcome of pateints surviving PUV, further research is warranted to enhance our understanding and management of this condition.

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