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Case Report

Paediatrics Section

Co-occurrence of Severe Pulmonary Stenosis and Pelvi-ureteric Junction Obstruction in a Neonate: A Case Report

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ABSTRACT

Severe Pulmonary Stenosis (PS) in neonates is a critical congenital cardiac condition that may initially present with subtle or no clinical signs. When coexisting with antenatal hydronephrosis, the clinical picture becomes more complex and may obscure the underlying pathophysiology. The present report describes a full-term male neonate born to consanguineous parents who was antenatally diagnosed with left-sided hydronephrosis and postnatally found to have a grade 3 pansystolic murmur. Echocardiography revealed severe PS with a pressure gradient of 89 mmHg. A successful Balloon Pulmonary Valvotomy (BPV) was performed, and the hydronephrosis was managed conservatively. The present case is unique as it highlights the uncommon co-occurrence of dual-organ anomalies-critical congenital heart disease and renal outflow obstruction-in a stable neonate with no overt symptoms at birth. The consanguineous background raised suspicion of a possible genetic association, necessitating genetic counselling for the family. Management required timely multidisciplinary intervention involving neonatology, paediatric cardiology, and nephrology, ensuring a favourable outcome. Vigilant neonatal examination, early recognition of subtle signs, and coordinated care are essential in managing infants with multiple congenital anomalies. The present case underscores the importance of considering genetic evaluation, long-term follow-up, and counselling in similar scenarios.

Keywords: Balloon valvotomy, Congenital heart disease, Hydronephrosis, Genetic counselling, Neonatal anomalies

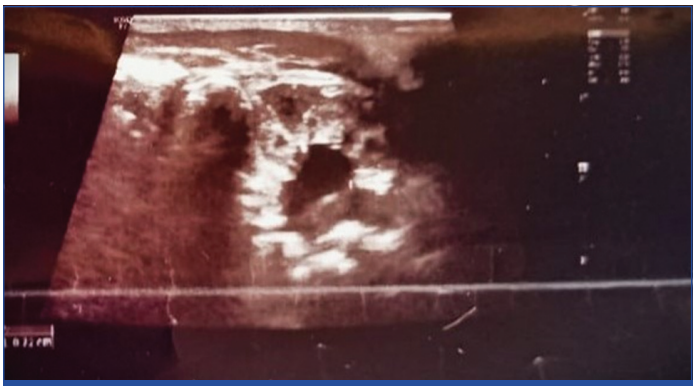
CASE REPORT

A male neonate presented to the Department of Paediatrics for routine postnatal examination on day 3 of life, when a cardiac murmur was detected. The neonate was born at 39 weeks of gestation via lower-segment caesarean section due to non-progressive labour. His birth weight was 2.3 kg, and APGAR {Appearance (skin color), Pulse (heart rate), Grimace (reflex irritability/response), Activity (muscle tone), and Respiration (breathing effort)} scores were 8 and 9 at one and five minutes, respectively. The baby cried immediately after birth and transitioned well.

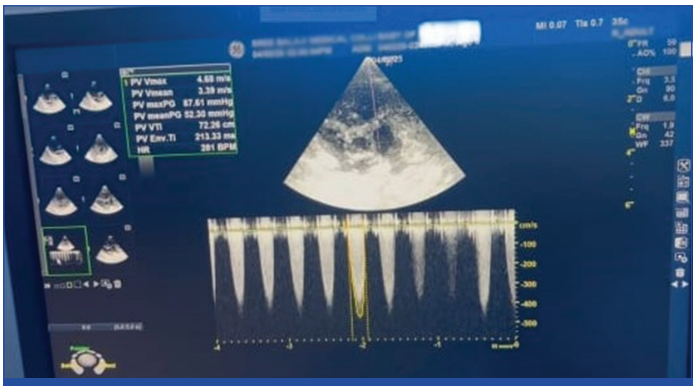
The parents were third-degree consanguineous, increasing the risk of autosomal recessive genetic conditions. The mother developed pregnancy-induced hypertension in the third trimester, which was managed conservatively. There were no other antenatal complications such as infections, teratogenic exposures, or gestational diabetes. Routine antenatal ultrasonography revealed mild left-sided hydronephrosis detected in the third trimester, with no other associated anomalies.

At birth, the neonate appeared clinically stable, with normal feeding, urine output, and passage of meconium. There were no dysmorphic features. The umbilical cord had a normal three-vessel configuration. On systemic examination on day 3 of life, vital parameters were stable (heart rate 142/min, respiratory rate 42/min, SpO_2 98% on room air). A grade 3 pansystolic murmur was detected on cardiovascular examination. Other systemic examinations were unremarkable, with no cyanosis, hepatomegaly, or respiratory distress.

An abdominal ultrasound confirmed the antenatal finding of mild left-sided hydronephrosis. The renal pelvis measured 7.2 mm, with abrupt narrowing at the Pelvi-Ureteric Junction (PUJ), consistent with PUJ obstruction [Table/Fig-1]. Echocardiography revealed severe valvular PS with a peak pressure gradient of 89 mmHg, associated pulmonary hypertension, and mild tricuspid regurgitation [Table/Fig-2,3]. Despite the absence of overt clinical symptoms, the severity of stenosis warranted urgent cardiology referral.

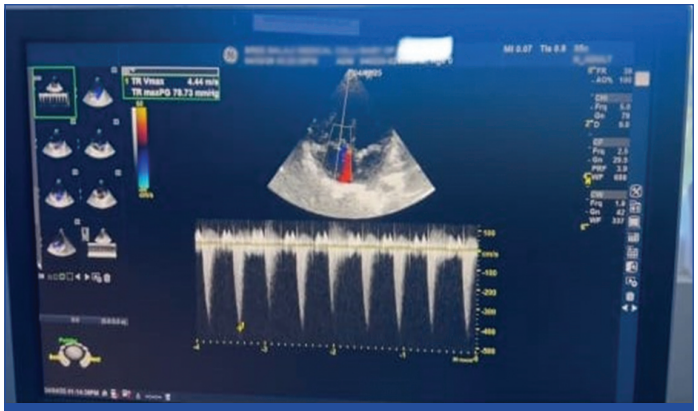


[Table/Fig-1]: Ultrasonography image showing the renal pelvis measured 7.2 mm, with an abrupt narrowing at the PUJ, consistent with PUJ obstruction.



[Table/Fig-2]: Transthoracic echocardiographic image showing severe pulmonary valve stenosis with a high-pressure gradient across the valve.

Preanaesthetic evaluation showed stable haemodynamics and normal baseline laboratory parameters. The infant underwent Balloon Pulmonary Valvotomy (BPV) under fluoroscopic guidance. A balloon catheter was introduced via the femoral vein and inflated across the pulmonary valve, relieving the obstruction within minutes. Following the intervention, the murmur resolved, and the infant remained clinically stable.



[Table/Fig-3]: Doppler study demonstrating a pressure gradient of 89 mmHg across the pulmonary valve, confirming the severity of the stenosis.

The PUJ obstruction was managed conservatively under paediatric nephrology care, with regular imaging to monitor renal function and pelvic dilatation. No surgical intervention was required during the neonatal period, as the hydronephrosis remained mild and non-progressive.

The infant was discharged on day 12 of life in stable condition, with follow-up planned for both cardiology and nephrology. At the two-month follow-up, echocardiography demonstrated sustained relief of PS, and renal function remained stable.

DISCUSSION

The present case highlights the rare co-occurrence of severe PS and left-sided PUJ obstruction in a full-term neonate, emphasising the need for clinical vigilance and multidisciplinary care.

Congenital heart disease has a reported prevalence of 8-10 per 1,000 live births, with PS accounting for approximately 5-10% of cases [1]. Severe PS in neonates may result in cyanosis, right ventricular hypertrophy, and reduced pulmonary perfusion; however, early clinical signs may be subtle. In the present case, a cardiac murmur was the first indication of disease and was detected only during routine postnatal examination. BPV is the first-line treatment for critical valvular PS in neonates and infants. Studies by Rao PS and McCrindle BW et al., have demonstrated high success rates (>90%) with low complication risks [2,3]. In patient of the present study, BPV resulted in immediate haemodynamic improvement.

Hydronephrosis occurs in approximately 1-5% of pregnancies and is most commonly caused by PUJ obstruction [4]. In neonates with mild hydronephrosis, conservative management with serial imaging is recommended, as deterioration is uncommon [5]. Several case reports have documented associations between antenatal hydronephrosis and congenital heart disease. Atić N et al., and Alp EK et al., described neonates presenting with both cardiac and renal anomalies, often creating diagnostic challenges and necessitating coordinated multidisciplinary management [6,7].

When renal and cardiac anomalies coexist, differential diagnoses include syndromic conditions such as Noonan syndrome, Alagille syndrome, and congenital obstructive uropathy syndromes [8]. In the present case, these conditions were considered but ruled out, as the neonate exhibited no dysmorphic features, systemic involvement, or biochemical abnormalities.

Given the consanguineous parental background, an underlying genetic aetiology remains possible, although no definitive diagnosis was established. Genetic testing and counselling were advised to guide future family planning and sibling screening. There was no family history of similar conditions. The hydronephrosis was detected antenatally in late gestation and remained mild postnatally. Existing literature suggests that antenatal hydronephrosis, even when mild, may occasionally serve as a marker for associated cardiac defects [9].

Consanguinity increases the likelihood of autosomal recessive inheritance. Genetic counselling was provided regarding recurrence risk, sibling evaluation, and consideration of genetic screening in future pregnancies. At short-term follow-up, both cardiac and renal outcomes were favourable. Long-term surveillance remains essential, as PUJ obstruction may progress over time and pulmonary valve restenosis can occur following intervention.

CONCLUSION(S)

The present case underscores the importance of thorough neonatal evaluation in identifying coexisting congenital anomalies. Early diagnosis and timely intervention for both severe PS and PUJ obstruction resulted in a favourable clinical outcome. Multidisciplinary coordination, genetic counselling, and long-term follow-up are crucial for optimising neonatal care and supporting informed family planning in similar clinical scenarios.

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