ACTA SCIENTIFIC CLINICAL CASE REPORTS

Volume 6 Issue 4 April 2025

Case Report

Severe Pulmonary Stenosis with Dysfunctional Single Kidney – A Successful High Risk Pregnancy

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Received: February 28, 2025

Published: March 27, 2025

Abstract

A high-risk pregnancy is a pregnancy where there is increased risk to both maternal and fetal health due to the pregnancy itself, or from the mother's preexisting condition. They need specialized care in all phases of pregnancy i.e. antenatal, intrapartum and post-partum period. Pulmonary stenosis and unilateral renal agenesis are two such high risk preexisting conditions in the mother that need close monitoring. Our case report shows the journey of a mother who despite all odds had a successful outcome.

Keywords: Severe Pulmonary Stenosis; High Risk Pregnancy; Unilateral Renal Agenesis; Cardio Obstetric Care; Nephro Obstetric Care

Introduction

Pulmonary stenosis (PS) is the narrowing of pulmonary valve, a rare congenital heart disease accounting for 7 to 12% of all cases of heart disease [1]. Its severity is determined by the peak gradient across the pulmonic valve estimated using echocardiography. Severe PS is defined as peak gradient of >64 mmHg. It comes under Class III of Modified World Health Organization (mWHO) Classification of Maternal Cardiovascular Risk, and is associated with increased risk of maternal mortality or morbidity, varying from 5 to 15%. Unilateral renal agenesis is yet another rare condition seen in roughly 0.1% of adults associated independently with higher risk of adverse outcomes in pregnancy like preeclampsia and preterm delivery [2]. In this case report, we present a pregnancy involving both these high-risk conditions in the mother and how this was managed successfully.

Case Report

A 27 years old primigravida with sickle cell trait first reported to antenatal OPD of All India Institute of Medical Sciences (AIIMS), Raipur, at 21+6 weeks period of gestation complaining of shortness of breath on exertion and palpitation for past 2 years, with recent worsening of symptoms for 15 days. She was symptomatic even at rest, coming under NYHA (New York Heart Association) Grade IV. On examination she had tachycardia with delayed peaking ejection systolic murmur grade IV/VI in pulmonary area. Her ECG showed right axis deviation, 'P' pulmonale and right ventricular hypertrophy with strain pattern. 2D ECHO was done and she was diagnosed with severe pulmonary stenosis with significant sub valvular dynamic obstruction. After complete evaluation and initial stabilization, she underwent Balloon Pulmonary Valvotomy with TYSHAK II Balloon at 23+1 weeks gestation at AIIMS Raipur. However, there was no significant change in Right Ventricular Outflow Tract (RVOT) gradient post procedure. She was put on Tab Metoprolol (extended-release tablets) 75mg BD. Her exertional spO2 dropped to 60% and thus termination of pregnancy was planned considering the maternal risk but the patient and her attenders refused and were lost to follow up.

With a telephonic call explaining all the risks and threat to her life, she was convinced to get admitted in the institute for safe confinement under the supervision of cardio-obstetric care. Strict bed rest with BP, PR and spO2 monitoring was done. At 28+3 weeks her

doppler scan showed early onset Stage 1 Fetal Growth Restriction (FGR). Hence, dexamethasone and MgSo4 coverage were given for lung maturation and neuroprotection of the baby respectively. Daily Fetal Movement Count (DFMC) charting was checked every day with Non-Stress Test (NST) being done twice daily for fetal monitoring.

Her creatinine values showed rising trend with elevated urine Albumin Creatinine Ratio (ACR) and 24-hour urinary protein. BP was however normal. Hence, USG KUB was done and she was incidentally found to have right renal agenesis. She was started on Tab sodium bicarbonate 500mg BD and Tab Alpha Ketoanalogue after taking Nephrology consultation with strict urine output monitoring. Weekly Cardiology consultation was done and patient was found to have improved symptomatically (NYHA Grade II) though her RVOT gradient remained unchanged. Dose of Metoprolol was increased to 100mg BD due to PR> 90/min persistently. Twice weekly USG Obst Doppler was done. Her serial growth scans showed poor fetal growth (EFW-0%) with pathological umbilical artery PI (>99%.). Rescue Dexamethasone dose was given at 31 weeks.

At 32+1 weeks there was Absent End Diastolic Flow (AEDF) in the umbilical artery. Hence, she was taken for Elective Caesarean section at 32 + 2 weeks in view of Stage 2 FGR with severe pulmonary stenosis under the supervision of cardiac anesthesia and Cardio Thoracic Vascular Surgery (CTVS) team. She had a preterm alive male of 1.26kg with Appearance Pulse Grimace Activity Respiration (APGAR) Score of 7/10 at 1 minute of delivery. Baby cried immediately after birth and was kept in Neonatal Intensive Care Unit (NICU) because of prematurity for 39 days. There were no intraoperative or postoperative adverse cardiac events. Both mother and baby are healthy at present. Baby is under Kangaroo Mother Care (KMC) and doing well. Mother was discharged with a plan of RVOT muscle resection at a later date and follow up in Nephrology OPD for single dysfunctional kidney.

Discussion

Pulmonary valvular stenosis accounts for 7% to 12% of congenital heart disease. Mild and moderate PS is usually asymptomatic. Severe PS causes RV hypertrophy and is related to poor long-term outcomes unless treated with valvotomy or balloon valvuloplasty. Intervention is recommended in patients with severe PS, regardless of the patient's symptom status. It comes under Class III of WHO Classification of Maternal Cardiovascular Risk, significantly increasing risk of maternal mortality or morbidity (5-15%). Studies have found that such patients achieve a decrease in pulmonary gradient with successful balloon pulmonary valvulotomy, improving the likelihood of an optimal pregnancy outcome [3].

In our case, however, such target could not be achieved and the RVOT gradient post procedure remained 170 mm Hg. Uncorrected severe PS is associated with a number of serious complications to the fetus, including preterm birth in 17% of patients and a high offspring mortality of 4.8%. As per our literature search, no cases of uncorrected severe PS at 32 weeks period of gestation could be found, indicating the rarity of a pregnancy being prolonged to such gestation under such high risk.

Moreover, the single kidney bearing the load of rising creatinine and persistent proteinuria combined with the risk of acute kidney injury was also worrisome. In general, it is advisable to terminate such pregnancy considering the maternal risk. But the mother was willing to risk her own health to realize her dream of motherhood which became a challenge for us. We managed to save the pregnancy with joint efforts of a cardiology team a nephrology team and a vigilant obstetric team pro towards saving both the mother and baby. Hence cardio obstetric and nephro obstetric clinics are to be recommended for follow up of such high-risk cases.

Conclusion

Severe pulmonary stenosis and unilateral renal agenesis are high risk pregnancy conditions which need proper counselling of the patient and family explaining both maternal and fetal risks. Such pregnancies need intensive specialist cardiac, nephrological and obstetric monitoring throughout pregnancy, childbirth and puerperium. Thus, with the help of early intervention and multidisciplinary approach, a high-risk pregnancy, combining two rare conditions in one, gave a healthy outcome of two joyful lives.

Bibliography

1. Mancebo A and Wanner A. "Lung tumor in a patient with congenital unilateral hypoplasia of the pulmonary artery". *Chest* 68.6 (1975): 846-847.

2. Ito M., *et al*. "Unilateral absence of the left pulmonary artery accompanied by right lung cancer". *Annals of Thoracic Surgery* 90.1 (2010): 6-8.

3. Makdisi G., *et al*. "Pulmonary Artery Agenesis Associated With Emphysema and Multiple Invasive NonSmall Cell Lung Cancers". *Annals of Thoracic Surgery* 99.6 (2015): 2192-2195.

4. Ping Wang., *et al*. "Isolated unilateral absence of pulmonary artery in adulthood: a clinical analysis of 65 cases from a case series and systematic review". *Journal of Thoracic Disease* 9.12 (2017): 4988-4996.

5. Zhang N., *et al*. "Clinical characteristics and prognosis of pulmonary inflammatory myofibroblastic tumor: An over 10-year retrospective analysis". *Pediatric Investigation* 4.3 (2020): 192-197.

6. Li X., *et al*. "A case report of tracheal inflammatory myofibroblastic tumor in a 34-week pregnant woman misdiagnosed with asthma". *Medicine (Baltimore)* 96.33 (2017): e7872.

7. Schweigert M., *et al*. "Use of extracorporeal membrane oxygenation in non-elective major thoracic surgery for infectious lung abscesses". *European Journal of Cardio-Thoracic Surgery* 25 (2022): ezac116.

8. Liang-Ze Zhang., *et al*. "Unilateral absence of pulmonary artery associated with contralateral lung cancer". *Journal of Thoracic Disease* 8.9 (2019): E942-E946.