Case Report

Successful outcome of pregnancy in RHD with severe MS, severe pulmonary artery hypertension, moderate MR/TR/AR and mild AR

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ABSTRACT
Rheumatic heart disease with severe Pulmonary Arterial Hypertension (PAH) in pregnancy is a grave situation, present with high maternal morbidity and mortality. In this case report, we describe our successful management of such a case which was even more difficult in combination with severe mitral stenosis, severe pulmonary artery hypertension and mild to moderate MR/TR. This patient got her diagnosis late in pregnancy, beyond the time at which a therapeutic termination could not have been performed.

Keywords: RHD, Pregnancy, PAH, MS

INTRODUCTION
Rheumatic heart disease with severe Pulmonary Arterial Hypertension (PAH) in pregnancy is a grave situation, present with high maternal morbidity and mortality. In this case report, we describe our successful management of such a case which was even more difficult in combination with severe mitral stenosis, severe pulmonary artery hypertension and mild to moderate MR/TR. This patient got her diagnosis late in pregnancy, beyond the time at which a therapeutic termination could not have been performed.

CASE REPORT
Patient was 23 years old unbooked female, primigravida with height of 156 cm and weight of 56 kg, presented with eight and half month amenorrhea. She gave history of hemoptysis, cough and dyspnoea of NYHA grade 2, belongs low socio-economic class. On examination, her pulse 80/min regular BP-110/70 mm of Hg with 1+/1+ peripheral oedema. There is loud S1 with diastolic murmur with grade (2/6) on cardiovascular examination. There is some evidence of pulmonary oedema in chest X-ray. Per abdomen examination showed 34 week size of uterus, relaxed, cephalic and presence of regular F.H.S 140.

The report of 2D-ECHO showed rheumatic heart disease, severe mitral stenosis with severe pulmonary artery hypertension, moderate TR/MR, mild PR and moderate AR (Figure 1, Figure 2) with systolic pulmonary hypertension of 75 mmHg and mitral valve area 1.2 cm² with mobility 2+, thickening 3+, calcification 1+, subvalvular tension 1+ (Wilkin’s Score 1-7/16). Patient was managed conservatively with medication as inj. benzathin penicillin, tab lanoxin, frusemide and injectable antibiotics. After 2 weeks elective LSCS was done with multidisciplinary team (anaesthetics, cardiologist and obstetrician) under epidural analgesia. Patient was monitored during labour and delivery with oximetry and arterial and central venous pressure line. Single live female baby weight 2.5 kg born with APGAR score 9 at the 1st and 5th minute. On post op day 3 patient was dyspotic, but she was managed by our multidisciplinary
team (anaesthetics, cardiologist and obstetrician). She discharged on post op day 14.

**Figure 1:** The report of 2D-ECHO showed rheumatic heart disease, severe mitral stenosis with severe pulmonary artery hypertension, moderate TR/MR, mild PR and moderate AR with systolic pulmonary hypertension of 75 mmHg and mitral valve area.

**DISCUSSION**

Cardiac disease was the most common cause of indirect maternal deaths and the most common cause of death overall. It may be present with cardiovascular decompensation during pregnancy at time of delivery or immediate postpartum period. The estimated incidence of PAH in pregnancy is 1.1 per 100000 pregnancies.\(^5\) PAH is considered a contra-indication to pregnancy, with an associated 30-50% risk of mortality.\(^4,5\) Pulmonary Hypertension (PH) is defined as a hemodynamic and pathophysiological condition with an elevated mean pulmonary arterial pressure (Pap ≥25 mm of Hg) at rest as assessed by right heart catheterisation.\(^6\) The physiological hemodynamic adaptations in pregnancy are poorly tolerated in PAH patients, with termination of pregnancy discussed and advised. The early introduction of advanced therapies and personalized planned delivery appears to confer improved outcomes.\(^5,6\) Women should review every month until 28 weeks gestation, fortnightly until 32 weeks and weekly thereafter until planned delivery. Elective caesareans were undertaken under regional anaesthesia with frequent, regular post-partum follow-up for six months. Advanced therapy options prescribed in pregnancy include calcium channel blockers, intravenous prostacyclin, prostacyclin analogues (nebulised iloprost) and phosphodiesterase inhibitors (sildenafil). Bosentan (an endothelin-1 receptor antagonist) has been shown to be teratogenic in animal studies and is therefore avoided. Anticoagulation with low molecular weight heparin or unfractionated heparin should also be considered. Fluid restriction, a low sodium diet and the use of diuretics in the third trimester have been implemented to reduce RV distension to minimize the risk of acute decompensation.\(^3\) Adequate cardiovascular invasive monitoring is essential and should be administered and maintained in the postpartum period with the same criteria that reduce morbidity and mortality in cardiac patients undergoing general surgery.\(^7,8\)

**CONCLUSION**

A successful outcome of pregnancy in PAH patients is possible but require careful monitoring, prompts treatment, and individualized clear management plans. This should cover the mode of delivery, anaesthesia techniques, and protocols for the management of PAH crises. Guidance on the escalation of pulmonary hypertension therapy should also be made available. These patients should certainly be managed in a tertiary centre environment by a specialist multi-disciplinary team.

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**REFERENCES**


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