Managing asymptomatic severe rheumatic mitral stenosis in pregnancy: a case report

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Background

Rheumatic heart disease (RHD) is a disease of disparity most prevalent in developing countries and among immigrant populations. Mitral stenosis (MS) is a common sequela of RHD and affects females disproportionately more than males. Rheumatic MS remains a significant management challenge as severe MS is usually poorly tolerated in pregnancy due to haemodynamic changes and increased cardiovascular demands of progressing pregnancy. Pregnancy remains contraindicated in current management guidelines based on expert consensus, due to a paucity of evidence-based literature.

Case summary

A 28-year-old aboriginal woman with known severe MS was found to be pregnant during routine health review, despite contraceptive efforts. Echocardiography demonstrated mean mitral valve (MV) gradient 14 mmHg; stress echocardiography demonstrated increased MV gradient 28–32 mmHg at peak exercise and post-exercise pulmonary artery pressure 56 + 3 mmHg with marked dynamic D-shaped septal flattening. Left ventricular systolic function remained preserved. She remained remarkably asymptomatic and underwent successful elective induction of labour at 34 weeks. Postpartum, she remained euvoalaemic despite worsening MV gradients and new atrial fibrillation (AF). She subsequently underwent balloon mitral valvuloplasty with good result.

Discussion

Severe rheumatic MS in pregnancy carries significant morbidity and mortality, due to an already fragile predisposition towards heart failure development compounded by altered haemodynamics. Pregnancy avoidance and valvular intervention prior to conception or in the second trimester remain the mainstay of MS management; however, we present an encouraging case of successful near-term pregnancy with minimal complications in a medically managed asymptomatic patient with critical MS, who subsequently underwent valvular intervention post-partum.

Keywords

Rheumatic heart disease • Mitral stenosis • Pregnancy • Balloon valvuloplasty • Heart failure • Pulmonary hypertension • Case report

Learning points

• There is limited information regarding the management of severe mitral stenosis (MS) in women of childbearing age or who are pregnant, particularly if they remain asymptomatic.
• Serial echocardiography combined with stress testing and physiological assessment for surveillance of mitral valve gradients, symptoms, and overall function during pregnancy may be utilized in asymptomatic patients to guide optimization of medical therapy, and timing of percutaneous balloon mitral valvuloplasty (BMV).
• Percutaneous BMV may be safely deferred until postpartum in women with severe MS who remain asymptomatic, to minimize procedural risks for mother and developing foetus.
**Introduction**

Left-sided stenotic valvulopathies, including mitral stenosis (MS), remain a feared entity in pregnancy due to the effects of altered physiology including increased intravascular volume, cardiac output and heart rate, and decreased systemic vascular resistance and systolic blood pressure\(^1\) which increases MS complication risks. Maternal complications include pulmonary oedema, arrhythmias, and increased mortality, whilst foetal complications include preterm delivery, intrauterine growth restriction, and foetal death.\(^1\)

The risk of developing heart failure corresponds with MS severity.\(^2\) Severe MS is usually poorly tolerated in pregnancy, with 67% of women developing peripartum heart failure even if previously asymptomatic.\(^3-4\) This is contributed to by increased heart rates antepartum, resulting in shortened diastolic filling time, increased left atrial pressure, and pulmonary venous pressures predisposing to heart failure.\(^4\) Pregnancy is contraindicated in current guidelines, which recommend percutaneous valvular intervention in women with moderate or severe MS contemplating pregnancy\(^7\) (Figure 1).

We present our management of a remarkably asymptomatic patient with critical MS, who presented 13 weeks antepartum despite contraceptive efforts and without preconception valve intervention. The valvular intervention occurred postpartum, with good outcomes for both mother and child.

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**Timeline**

**Preparation**

- **Antagonist**
  - February 2020: Presented to remote cardiologist for antifibrinolytic inpatient and found to be 33 weeks pregnant.
  - Full-cardiac TTE (33 weeks): severe MS (4/6 mitral), pulmonary hypertension (PAP 41 mmHg), normal LV function.
  - Review by Cardiologist at 33 weeks antepartum.
  - 18 March 2020: TTE (33 weeks antepartum): severe mitral stenosis (mitral valve area 0.6 cm\(^2\) orifice area).
  - Normal LV size and function.
  - Normal LA size and function.

- **Anticoagulation**
  - 23 March 2020: TTE (20 weeks ante-partum): Atrial fibrillation.

- **Percutaneous baloon mitral valvoplasty**
  - 6 May 2020: Ancillary echocardiogram revealed left atrial thrombus.
  - 25 May 2020: Referral to tertiary hospital for further evaluation.

- **Surgical valve repair / replacement**
  - July 2020: TTE (14 weeks antepartum): severe MS (mitral valve area 0.5 cm\(^2\)).

- **Post-partum care**
  - July 2020: Presented to remote cardiologist for antifibrinolytic inpatient and found to be 33 weeks pregnant.
  - Full-cardiac TTE (33 weeks): severe MS (4/6 mitral), pulmonary hypertension (PAP 41 mmHg), normal LV function.

**Figure 1** Managing severe rheumatic mitral stenosis in pregnancy.

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\(1\) Increased mortality, whilst foetal complications include preterm delivery, intrauterine growth restriction, and foetal death.

\(2\) Severe MS is usually poorly tolerated in pregnancy, with 67% of women developing peripartum heart failure even if previously asymptomatic.

\(3\) This is contributed to by increased heart rates antepartum, resulting in shortened diastolic filling time, increased left atrial pressure, and pulmonary venous pressures predisposing to heart failure.

\(4\) Pregnancy is contraindicated in current guidelines, which recommend percutaneous valvular intervention in women with moderate or severe MS contemplating pregnancy.

\(7\) We present our management of a remarkably asymptomatic patient with critical MS, who presented 13 weeks antepartum despite contraceptive efforts and without preconception valve intervention. The valvular intervention occurred postpartum, with good outcomes for both mother and child.
Case presentation

A 28-year-old aboriginal Australian woman from a remote community with known severe MS was found to be 13 weeks pregnant despite contraceptive depot injections, after presenting for routine review at the local community clinic.

She had a previous episode of acute rheumatic fever aged 10. She had been adherent with standard secondary prophylaxis regimen since but subsequently developed severe mitral stenosis necessitating balloon mitral valvuloplasty (BMV) aged 14. Prior to pregnancy, her last echocardiogram in 2018 demonstrated severe rheumatic MS with mean pressure gradient (mPG) 17 mmHg, mitral valve area (MVA) 0.9 cm², and mild pulmonary hypertension with estimated pulmonary artery pressure (EPAP) 30 mmHg + right atrial pressure (RAP) 3 mmHg. Left ventricular systolic function was preserved. She remained clinically asymptomatic and was therefore managed conservatively. Additionally, she received pregnancy-avoidance counselling and agreed to commence 3-monthly contraceptive depot injections. She did not have other medical conditions or regular medications.

Due to the cultural significance of pregnancy within her community, she elected to continue her pregnancy fully cognisant of the increased risks of adverse outcomes secondary to her valvulopathy. She was subsequently referred for Cardiology Specialist input.

She remained remarkably asymptomatic despite critical MS. Physical examination demonstrated dual heart sounds with an additional soft diastolic murmur. There was no evidence of pulmonary congestion of peripheral oedema. Her NTproBNP level was 136 ng/L. She had mild normocytic anaemia (Hb 106 g/L; mean cell volume 85.8 fL); electrolytes and creatinine levels were normal (sodium 135 mmol/L, potassium 3.8 mmol/L, creatinine 54 μmol/L, estimated glomerular filtrate rate >90 mL/min/1.73 m²). Transthoracic echocardiography (TTE) at 13 weeks antepartum demonstrated a doming hockey stick appearance of the anterior mitral valve leaflet, thickened immobile posterior mitral leaflet with markedly restricted excursion resulting in severe

Figure 2 Exercise stress echocardiogram demonstrating (A) left ventricular at rest and (B) marked septal D-shape flattening with peak exercise.

Figure 3 Transthoracic echocardiogram demonstrating (A) severe mitral stenosis and (B) pulmonary hypertension.
mitral stenosis (mPG 14 mmHg), mitral valve area 0.4 cm², severely dilated left atrium (indexed volume 50 mL/m²), mild pulmonary hypertension (EPAP 34 ± 3 mmHg). Stress echocardiogram performed at 20 weeks antepartum demonstrated mitral valve (MV) mPG 28–32 mmHg at peak exercise, and post-exercise EPAP 56 ± 3 mmHg with development of D-shaped septal flattening (Figure 2). She did not desaturate during this test. Given her minimal symptoms, she was successfully managed in a culturally sensitive manner in her remote community until the early third trimester, where she was electively admitted to a local tertiary hospital before transfer to a tertiary centre with cardiothoracic services for valve intervention prior to delivery.

Serial TTEs during antepartum demonstrated critical but stable MV gradients (mPG 19–21 mmHg, Figure 3). She was also commenced on beta-blocker therapy to minimize maternal tachycardia. She remained clinically and haemodynamically stable until the planned induction of labour at 34 weeks. This was subsequently
converted to caesarean section due to abnormal foetal cardiotocography, with the delivery of a healthy male baby.

Beta-blocker therapy was held for 24 h post-partum. The reason for this was unclear however was re-initiated together with digoxin due to worsening MV gradients (mP 29 mmHg), EPAP 57 ± 3 mmHg (Figure 4), and development of palpitations secondary to new atrial fibrillation (AF) Day 2 postpartum. She remained clinically euovoltaemic, and following consultation with her treating teams, she underwent elective BMV Day 8 postpartum with improved MV gradients and MVA (Figure 5). She remained well and was discharged Day 10 post-partum post-contraceptive implantation. She remained well on telephone review Day 2 post-discharge, prior to return to her local community.

Discussion

Whilst not common in developed countries, MS accounts for 9.5% of all valvular heart disease in Europe. Rheumatic heart disease (RHD) remains the most common aetiology. In Australia, RHD is a striking disease of disparity, where 89% of affected Australians identify as Aboriginal or Torres Strait Islander, representing a rate 6.6 times higher than non-Indigenous Australians. 47% of Aboriginal or Torres Strait Islanders affected are aged under 20 years. Women account for 61% of the total indigenous RHD burden, attracting the greatest risk of poor outcomes, especially during pregnancy.

Pregnancy remains a World Health Organisation (WHO) Class IV contraindication in severe MS, due to maternal and foetal morbidity and mortality; however, there is a paucity of information regarding the management of severe MS in women of childbearing age or who are pregnant, particularly if they remain asymptomatic. The current guidelines are based on expert consensus which strongly recommends pregnancy avoidance or consideration of pregnancy termination, and do not address or acknowledge cultural sensitivities, varying levels of medical literacy or socioeconomic constraints of patient populations where RHD is likely more prevalent. Management of an already pregnant patient focuses on a combination of pharmacological agents including beta-blockers and diuretics, and clinical monitoring to identify the development of intervention indications. Percutaneous valvular intervention is recommended preconception. The intrapartum intervention has been reported but only in persistently symptomatic patients between second and third trimesters to consider organogenesis of the foetus. This does not take into account the hyperactive mammary tissue exposed to radiation, or the longer-term risk of childhood malignancy. Current guidelines do not address or acknowledge cultural sensitivities, varying levels of medical literacy or socioeconomic constraints of patient populations where RHD is likely more prevalent. Management of an already pregnant patient focuses on a combination of pharmacological agents including beta-blockers and diuretics, and clinical monitoring to identify the development of intervention indications. 5,8 Percutaneous valvular intervention is recommended preconception. The intrapartum intervention has been reported but only in persistently symptomatic patients between second and third trimesters to consider organogenesis of the foetus. This does not take into account the hyperactive mammary tissue exposed to radiation, or the longer-term risk of childhood malignancy. Current guidelines do not address or acknowledge cultural sensitivities, varying levels of medical literacy or socioeconomic constraints of patient populations where RHD is likely more prevalent. Management of an already pregnant patient focuses on a combination of pharmacological agents including beta-blockers and diuretics, and clinical monitoring to identify the development of intervention indications. 5,8 Percutaneous valvular intervention is recommended preconception. The intrapartum intervention has been reported but only in persistently symptomatic patients between second and third trimesters to consider organogenesis of the foetus. This does not take into account the hyperactive mammary tissue exposed to radiation, or the longer-term risk of childhood malignancy. Current guidelines do not address or acknowledge cultural sensitivities, varying levels of medical literacy or socioeconomic constraints of patient populations where RHD is likely more prevalent. Management of an already pregnant patient focuses on a combination of pharmacological agents including beta-blockers and diuretics, and clinical monitoring to identify the development of intervention indications.

This case demonstrates a number of important points. Achievement of pregnancy avoidance may not always be achieved despite its prescription. Serial echocardiography in combination with stress testing and physiological assessment for the surveillance of MV gradients, symptoms, and overall function during pregnancy may be utilized in asymptomatic patients to guide optimization of medical therapy, as well as the timing of percutaneous BMV. We have also demonstrated the safe and successful deferral of percutaneous BMV until post-partum in asymptomatic patients, protecting both the developing foetus and also maternal mammary tissue which remains increasingly active postpartum from potentially harmful radiation.

Our case highlights the importance of patient-specific care and that further understanding of severe MS management in women of childbearing age and intrapartum is required to improve maternal and foetal outcomes.

Lead author biography

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Supplementary material

Supplementary material is available at European Heart Journal - Case Reports online.

Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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