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CASE REPORT
Other

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 sequalae 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 euvolaemic 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.

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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.[2–4] 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[5] (*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.

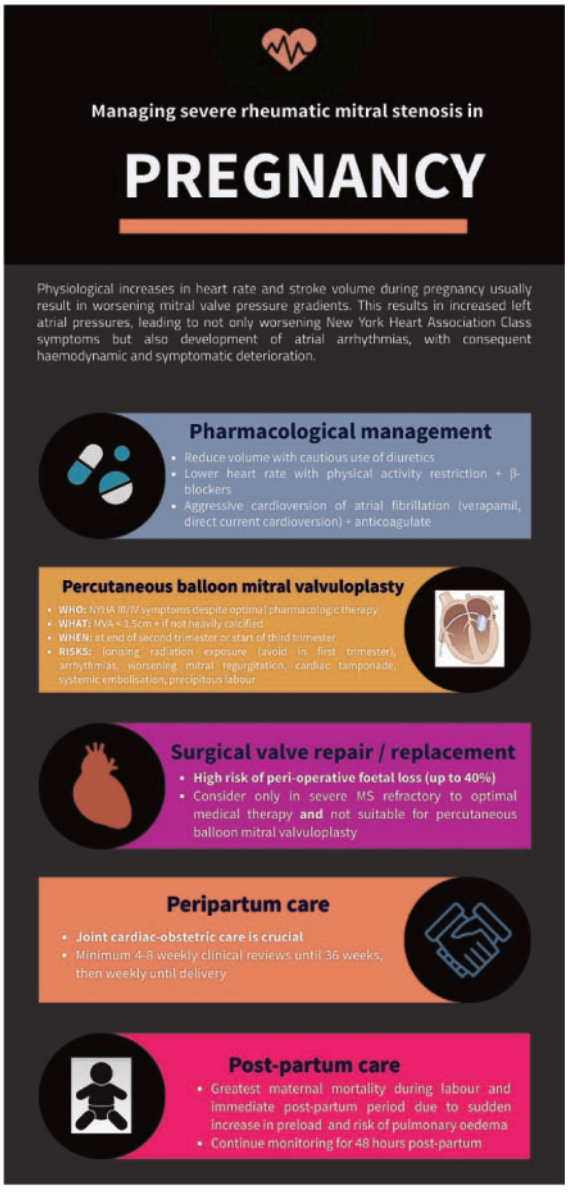

Figure 1 Managing severe rheumatic mitral stenosis in pregnancy.

Timeline

Timeline	Description
Prepartum	
	Acute rheumatic fever first diagnosed in 2002 (aged 10). Progression to severe MS necessitating BMV in 2006 (aged 14). Adherent with standard secondary prophylaxis regimen (LA bicillin). Subsequent recurrence of severe MS however no further valve intervention as asymptomatic Pregnancy-avoidance counselling and commenced on 3-monthly contraceptive depot injections
Antepartum	
February 2020	Presented to remote community clinic for contraceptive depot injection and found to be 13 weeks pregnant.
	First antepartum TTE (13 weeks) - severe MS (mPG 14 mmHg) + pulmonary hypertension (EPAP 37 mmHg). Normal LV function.
	Reviewed by Cardiologist at 15 weeks antepartum.
19 March 2020	TTE (19 weeks antepartum) - rheumatic severe MS (mPG 18 mmHg, MVA 0,7 cm^2 planimetry). Normal RV size and function. Normal LV size and function.
23 March 2020	ESE (20 weeks antepartum) Achieved 7,5 minutes on Bruce protocol, reaching 10 METS and 75% of maximal age-predicted heart rate. Test ceased due to non-cardiac symptoms (patient did not have breakfast prior). Severe MS (mPG 16-18 mmHg) at rest, increasing to 28-32 mmHg at peak exercise. Post-exercise pulmonary artery pressure 56 + 3 mmHg with marked D-shaped septal flattening. Preserved LV function throughout ESE.
	Local multidisciplinary team meeting involving Cardiology, Obstetrics and Anaesthetics regarding pregnancy management. Subsequent discussion with interstate tertiary hospital Heart Team - BMV should ideally be performed in first trimester however given patient currently 20 weeks antepartum, BMV to be delayed for as long as possible until at least 30 weeks antepartum.
6 May 2020	6-minute walk test - achieved 250 – 368m and HR 95-134. O_2 94-97% on room air.
21 May 2020	Patient electively transferred to tertiary hospital for further observation until planned vaginal delivery.
26 May 2020	TTE (24 weeks antepartum) - severe MS (mPG 22 mmHg; MVA 1,1 cm^2). Mild MR. EPAP 43 + 3 mmHg. Normal LV function – EF 65%.
29 May 2020	Initiated on labetalol 50mg BD
1 June 2020	TTE (29 weeks) - similar to previous. Labetalol changed to metoprolol 25mg BD
9–26 June 2020	Weekly TTEs between 29 - 33 weeks antepartum – similar findings to previous.
30 June 2020	Planned induction of labour at 34 weeks antepartum. Subsequently converted to lower-section caesarean section due to abnormal foetal cardiotocography. Successful delivery with no immediate postpartum complications.
Postpartum	
1 July 2020	TTE - severe MS (mPG 29 mmHg, MVA 0,5 cm^2). Normal LV function – EF 50-55%. EPAP 57 + 3 mmHg. Normal IVC size which collapses with inspiration. Normal RV size and function. Metoprolol withheld (unclear reason)
2-4 July 2020	First postpartum Cardiology review - for 6-minute walk test to assess symptoms and guide potential valve intervention prior to discharge. Subsequently developed palpitations in context of new-onset AF RVR. Recommenced on metoprolol and initiated on digoxin for further rate control. Reverted to sinus rhythm 3/7/2020 before further paroxysmal AF RVR 4/7/2020. Metoprolol increased to 50mg BD
7 July 2020	Reviewed by Interventional Cardiologist and planned for elective BMV.
8 July 2020	Underwent elective BMV. Pre-BMV TOE: severe MS (mPG 10 mmHg, MVA 0,6 cm^2 planimetry) and marked spontaneous echo contrast. Post-BMV TOE: severe MS (mPG 5 mmHg, MVA 1,2 cm^2 planimetry) and reduction in spontaneous echo contrast. No pericardial effusion. Reverted to sinus rhythm.
9 July 2020	Post-BMV TTE: Moderate MS (mPG 6 mmHg, MVA 1,3 cm^2 planimetry). Normal LV size and systolic function – EF 59%. Mild to moderate MR. EPAP 31-36 mmHg + RAP 3-8 mmHg. Initiated on warfarin in context of valvular AF with CHA_2DS_2-VASC score 1.
10 July 2020	Implanon contraception inserted. Discharged from hospital on metoprolol 25mg BD and warfarin with bridging clexane.

TTE = transthoracic echocardiogram, TOE = transoesophageal echocardiogram, ESE = exercise stress echocardiogram, AF RVR = atrial fibrillation with rapid ventricular response, HR = heart rate, MV = mitral valve, BMV = balloon mitral valvuloplasty, mPG = mean pressure gradient (mmHg), EPAP = estimated pulmonary artery pressure, MS = mitral stenosis, MR = mitral regurgitation, IVC = inferior vena cava, LV = left ventricle, RV = right ventricle, EF = ejection fraction

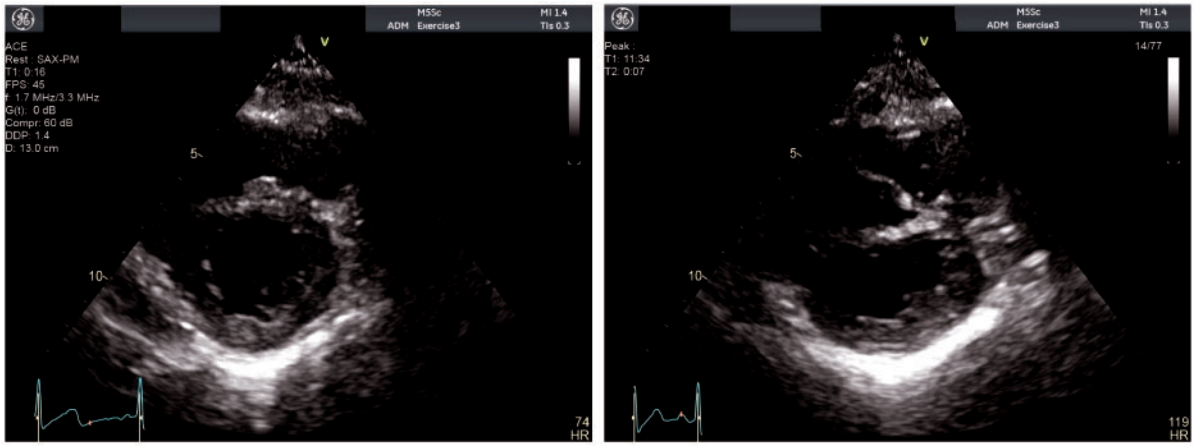

Figure 2 Exercise stress echocardiogram demonstrating (*A*) left ventricular at rest and (*B*) marked septal D-shape flattening with peak exercise.

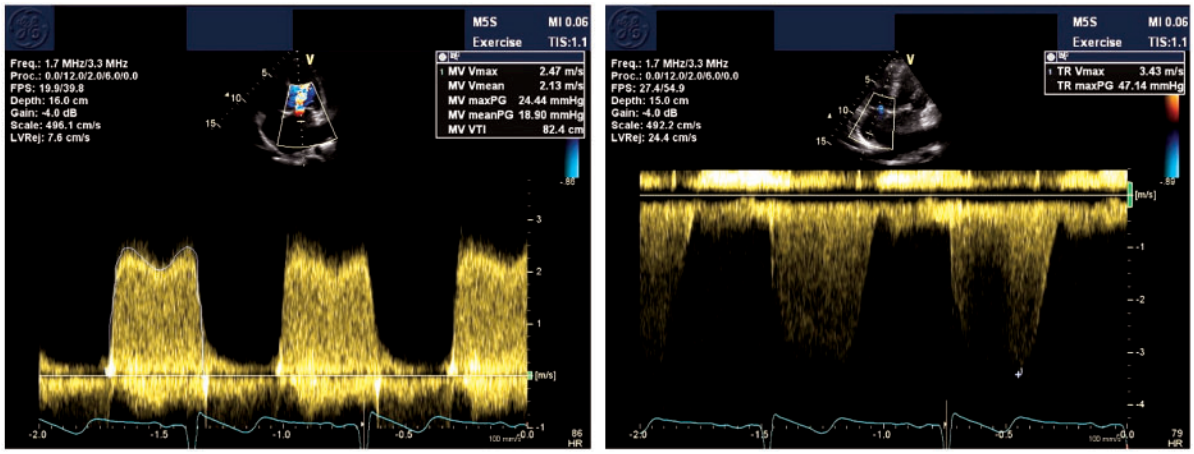

Figure 3 Transthoracic echocardiogram demonstrating (*A*) severe mitral stenosis and (*B*) pulmonary hypertension.

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 cm^2, 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 m^2). 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

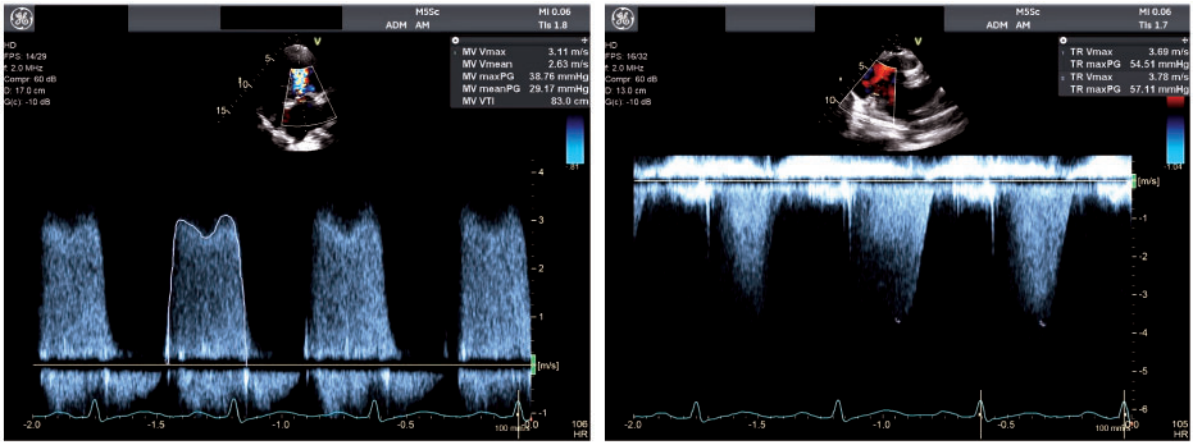

Figure 4 First postpartum transthoracic echocardiogram demonstrating (*A*) critical mitral stenosis and (*B*) pulmonary hypertension.

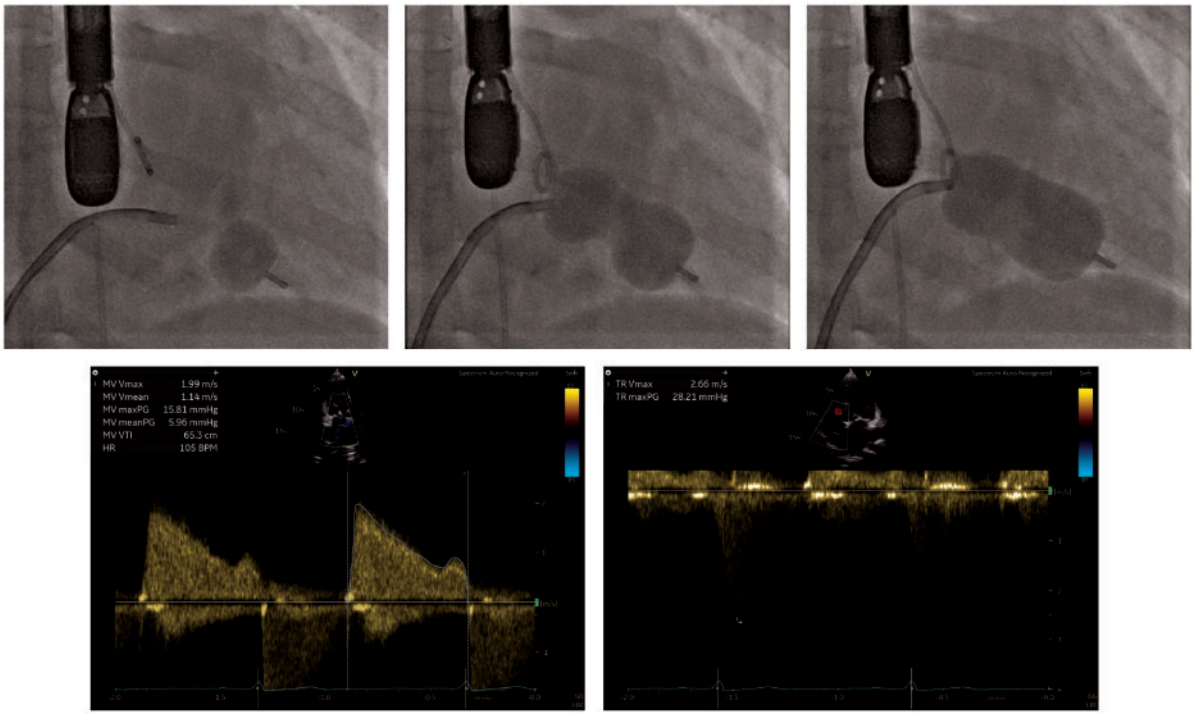

Figure 5 Balloon inflation during balloon mitral valvuloplasty. (*A*) Distal balloon inflation. (*B*) Inflation of proximal and middle balloon, with waist in mid-portion of balloon demonstrating mitral stenosis. (*C*) Full balloon inflation. Post-balloon mitral valvuloplasty transthoracic echocardiogram demonstrating (*D*) moderate mitral stenosis and (*E*) mild pulmonary hypertension.

mitral stenosis (mPG 14 mmHg), mitral valve area 0.4 cm^2, severely dilated left atrium (indexed volume 50 mL/m^2), 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 (mPG 29 mmHg), EPAP 57 ± 3 mmHg (*Figure 4*), and development of palpitations secondary to new atrial fibrillation (AF) Day 2 postpartum. She remained clinically euvolaemic, 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.[6] 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.[7] 47% of Aboriginal or Torres Strait Islanders affected are aged under 20 years.[7] Women account for 61% of the total indigenous RHD burden,[7] 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;[5] 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.[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[9] 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 account for many of these issues, or the timing and utility of serial echocardiographic assessments to guide percutaneous intervention as pregnancy develops.

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

Joanne M. H. Eng-Frost is a Cardiology Advanced Trainee at Flinders Medical Centre in Adelaide, Australia. She completed a Bachelor of Science (Biomedical Science) degree with Honours in Physiology at the University of Adelaide, Australia before earning a Doctor of Medicine (MD) degree at Flinders University, Australia. Her clinical interests include structural heart disease and interventional cardiology.

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.

Conflict of interest: None declared.

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