Percutaneous mitral commisurotomy during pregnancy – A report of two cases performed in a United Kingdom tertiary centre and a review of the literature

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Abstract

We report two cases of severe mitral stenosis where percutaneous mitral commisurotomy was performed within pregnancy. The first case involves an emergency procedure for a new diagnosis of severe mitral stenosis in a woman presenting with pulmonary oedema at 27 weeks' gestation. The second case is of a woman known to have mitral stenosis who underwent a semi-elective procedure for deterioration in symptoms. This procedure is not commonly performed in the United Kingdom because of low incidence of rheumatic heart disease. In addition, percutaneous mitral commisurotomy during pregnancy is rarely performed in the United Kingdom because of the improved healthcare system where majority of the women with moderate to severe mitral stenosis (even asymptomatic) will undergo planned interventions (percutaneous mitral commisurotomy or mitral valve surgery) before contemplating pregnancy. These cases highlight both the acute and chronic presentations of mitral stenosis and the impact pregnancy has on this condition. In addition, these cases show the importance of retaining skills in performing percutaneous mitral commisurotomy within our United Kingdom cardiologists.

Keywords

High-risk pregnancy, cardiology

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Introduction

Cardiovascular diseases remain not only the largest single cause of indirect maternal death in the United Kingdom but also the commonest overall cause of death according to the last Confidential Enquiry into Maternal Death across United Kingdom (UK) and Ireland.¹ Mitral stenosis (MS) is the commonest cause of acquired heart disease in pregnancy and accounts for 0–32% of maternal mortality worldwide, with outcomes varying largely by geographical region.^{2–7}

Marked haemodynamic changes occur during pregnancy, which can have profound effects on pre-existing heart disease. Hence, the management of patients with cardiovascular disease during pregnancy poses a challenge to obstetricians, cardiologists and anaesthetists. These changes are particularly problematic in pregnant woman with MS as the fixed valve struggles to cope with the increase in cardiac output and heart rate, resulting in an increase in transmitral pressure gradient. Decompensation of previously asymptomatic patients, with even moderate MS has been described.^{2–4} Such patients are at risk of developing acute pulmonary oedema and cardiogenic shock, along with thromboembolism and atrial fibrillation (AF). The risks are greatest during the second and third trimesters, although complications can also occur intra-partum or immediate postpartum. The prompt diagnosis and the swift initiation of the appropriate management regime remain the greatest challenge in the acutely decompensated patient.

We report two cases of severe MS where percutaneous mitral commisurotomy was performed within pregnancy. The first case involves an emergency procedure for a new diagnosis of severe MS in a woman presenting with pulmonary oedema at 27 weeks' gestation. The second case is of a woman known to have MS who underwent a semi-elective procedure for deterioration in symptoms. This procedure is not commonly performed in the UK because of low incidence of rheumatic heart disease. In 2013, 84 PTMC procedures were undertaken in 28 institutions. The median number of PTMC procedures per institution was 2. In addition, PTMC during pregnancy is rarely performed in the UK because of the improved healthcare system where majority of the

women with moderate to severe MS (even asymptomatic) will undergo planned interventions (PTMC or mitral valve surgery) before contemplating pregnancy.

Patient and observation

Case 1

A 34-year-old, otherwise fit and healthy, Asian woman was booked under midwife-led care during her third pregnancy. Her first two pregnancies ended in early losses. She presented at 27 weeks' gestation to Emergency Department with worsening breathlessness, pleuritic chest pain and productive cough over the last few days. She also gave a history of limited exercise tolerance pre-pregnancy.

On examination, she was tachycardic, hypotensive and found to be in respiratory distress. Her heart sounds revealed a diastolic murmur. Her jugular venous pressures were elevated. On lung auscultation, she had bilateral wheeze and crepitations.

An electrocardiogram (ECG) showed sinus tachycardia. Arterial blood gas (ABG) was suggestive of Type 1 respiratory failure. Her WCC and CRP were raised at 13.6 and 109, respectively.

Differential diagnoses included community-acquired pneumonia, aortic dissection or pulmonary embolus.

She was started on intravenous antibiotics and given 2.5l of crystalloid and colloids. She was started on non-invasive ventilation with a

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plan to transfer to Adult Intensive Care Unit for early intubation and ventilation, if necessary.

An urgent transthoracic echocardiogram (TTE) was undertaken as she deteriorated clinically, which showed severe MS, likely rheumatic in origin with mean gradient across mitral valve of 23 mm of Hg and pulmonary artery systolic pressure of 60 mm of Hg (Figure 1). This was confirmed on transoesophageal echocardiography (TOE), which also importantly excluded thrombus in the left atrium and clinically significant mitral regurgitation (MR).

A final diagnosis of pulmonary oedema secondary to moderate to severe MS with superimposed community-acquired pneumonia was made. She was intubated and ventilated, started on intravenous diuretics (furosemide) and metoprolol and transferred to the cardiac unit with a view to perform PTMC. Two doses of dexamethasone were given 24 h apart to promote fetal lung maturation. She was extubated the following day to allow us to obtain informed consent for PTMC and to have an informed discussion with her family. However, her clinical condition deteriorated significantly following extubation. She was reintubated and transferred to the cardiac catheterisation laboratory urgently that evening for emergency PTMC.

PTMC was undertaken under GA with TOE guidance. Right femoral vein access was achieved followed by trans-septal puncture to allow a 26 mm Inoue balloon to pass from the right atrium to the left atrium. After crossing the stenosed mitral valve, the balloon was then inflated within the mitral valve resulting in commissural splitting and expansion of the mitral orifice. The mitral valve was dilated serially from 22 mm to 26 mm in 1 mm increments under close TOE and fluoroscopic guidance to allow identification of any resultant mitral regurgitation at the earliest opportunity that would contraindicate further dilatation (Figure 2). PTMC carries a 5% risk of severe MR secondary to mitral valve disruption from oversizing of the balloon, which in this case may have resulted in decompensation, leading to possible foetal and maternal fatality. Special precautions were taken to reduce the radiation dose to both patient and foetus. These included minimising screening time, decreasing the frame rate and using TOE. The screening time was 17.6 min with a radiation dose of 1322.59 cGycm². Approximately 30 ml of Visipaque 320 contrast

agent was used. TOE guidance showed no increase in mitral regurgitation after the procedure, but a significant increase in valve area (1 cm² to 1.4 cm²) and reduction in the pressure gradient. She developed AF periprocedurally requiring DC Cardioversion with 100 J to restore sinus rhythm. She recovered well post-operatively and was discharged home after five days. Post-procedural echocardiogram confirmed improvement in mitral valve area with only mild MS and mild MR.

Figure 2. Fluoroscopy image of the PTMC with the Inoue balloon inflated across the mitral valve.

Figure 1. TOE image of the mitral valve with typical appearances of thickened tips and chordae.

She was followed up regularly in Antenatal Clinic with close input from cardiologists. Her breathlessness improved significantly following the above procedure and she tolerated the rest of the pregnancy very well. She developed gestational diabetes at 29 weeks' gestation, which was well controlled with diet. Regular fetal growth scans were normal. TTE was repeated at 34 weeks' gestation, which showed residual mild MS with a mean gradient of 5.5 mm of Hg and normal biventricular function. She went into spontaneous labour at 38 weeks' gestation and delivered vaginally a live male infant weighing 3200 g. She remained haemodynamically stable throughout the intrapartum and immediate postpartum period. She was discharged home after 48 h. Long-term follow up with the cardiology team has been planned.

Case 2

A 32-year-old Asian woman was booked consultant led care at 11 weeks of gestation during her fourth pregnancy as she was known to have moderate MS. In the past, she had three uncomplicated pregnancies and vaginal births.

She was from Pakistan and moved to United Kingdom two years ago and had very limited English. Her husband translated for the majority of the consultations, although telephone translation services were also used. She had rheumatic fever as a child and subsequently developed MS. She was thought to have had two mitral valvulotomies in the past in Pakistan. Symptomatically, she could walk on the flat with no real restrictions, but became short of breath when walking up an incline. Due to the language barrier it was difficult to elicit her true level of exercise tolerance, but was graded as New York Heart Association (NYHA) I-II. There was no history of presyncope or syncope. Echocardiogram done in early pregnancy showed severe MS with severely dilated left atrium but normal biventricular size and function.

She was admitted to hospital at around 27 weeks of gestation with fast AF and was started on IV Digoxin, which achieved cardioversion. She was discharged on 125 µg of Digoxin and low-molecular weight Heparin injections.

She was reviewed by the cardiology team at 31 weeks of gestation. She was quite comfortable at rest and had no specific complaints but on direct questioning she acknowledged chest discomfort, breathlessness and some light-headedness with physical effort such as walking or climbing stairs. In view of the difficulty in accurately establishing her symptoms, she was asked to demonstrate her exercise tolerance. She managed the stairs only extremely slowly and was not able to reach the next level without stopping due to light-headedness, chest discomfort and breathlessness. Her ECG confirmed sinus rhythm. Her repeat echocardiogram showed mitral valve was severely stenosed with mean gradient of 15 mm of Hg and having increased from 9.6 mm of Hg only three months earlier. Both atria were severely dilated (left > right). Good biventricular systolic function was present. In view of marked limitation of exercise tolerance and echocardiogram findings, an urgent PTMC was arranged. Two doses of dexamethasone injections were given 24 h apart to promote fetal lung maturity, in case the delivery is indicated during or immediately after the procedure.

At 33 weeks' gestation, she underwent PTMC under GA with TOE guidance. Her intra-atrial septum was rather tough, presumably on account of previous septal puncture and the right atria significantly enlarged, such that quite a considerable additional bend had to be made to the Brockenbrough needle to achieve satisfactory septal puncture in the fossa ovalis. The presence of TOE guidance was especially reassuring in light of this distorted anatomy. A 26 mm Inoue balloon was sequentially dilated from 22 mm to 26 mm resulting in a good improvement morphologically in valve motion and a halving of mitral gradient from 10 mm of Hg to 5 mm of Hg under general anaesthesia. She recovered well post-operatively and was discharged home the next day.

She was followed up every two weeks in Antenatal Clinic with close input from cardiologists. Her effort tolerance improved to some extent following the above procedure and she tolerated the rest of the

pregnancy well. She developed iron deficiency anaemia, which responded to oral iron supplementation. Regular fetal growth scans were normal. Her labour was induced at 39 weeks' gestation due to complex social circumstances. She had delivered vaginally a live female infant weighing 2600 g. She remained haemodynamically stable throughout the intrapartum and immediate postpartum period. She was discharged home after 48 h. Long-term follow up with the cardiology team has been planned.

Discussion

Rheumatic heart disease remains a major problem in developing countries and is still seen in western countries, especially within the immigrant population. An increasing number of women who are giving birth within the UK were born overseas. According to Office of National Statistics,⁸ 25.91% births in 2012 were to women who had not themselves been born in UK, and thus it is likely that we will continue to see these women within the obstetric setting. These cases highlight both the acute and chronic presentations of MS and the impact pregnancy has on this condition. In addition, these cases show the importance of retaining skills in performing PTMC within our UK cardiologists.

The first case emphasises the importance of the rapid diagnosis of a life threatening condition and the importance of remembering rarer causes of collapse in obstetric patients. Although the acute presentation of MS is frequently seen in the Asian subcontinent, it is rarely seen in the Western world; it is important to educate ourselves in the recognition of this condition to allow swift initiation of the appropriate management and referral to the appropriate speciality.

The second case highlights the difficulty in making adequate clinical assessments in migrant population due to language barrier, thereby hindering optimal management in a timely fashion.

MS is the most common cause of acquired heart disease during pregnancy and is predominantly caused by rheumatic heart disease,⁹ although less than 50% of patients with a heart lesion of rheumatic origin have a history of rheumatic fever.¹⁰

MS is a state of fixed cardiac output caused by left atrial outflow obstruction, with an increase in left atrial and pulmonary vasculature pressure resulting in pulmonary oedema. The mitral orifice normally is 4 to 6 cm² area by planimetry in mode 2D echocardiography short axis. Patients will not experience valve-related symptoms until the valve area is 2–2.5 cm² or less, at which point moderate exercise or tachycardia may result in exertional dyspnoea. Severe MS occurs with a valve area of less than 1 cm². In pregnancy, the risk of cardiac compromise is further aggravated by an increase in intravascular volume and maternal tachycardia. In cases of moderate to severe MS, approximately 60% of women will experience their first episode of pulmonary oedema at approximately 30 weeks' gestation.¹⁰ This risk continues during intrapartum and immediate postpartum period. The hyper coagulability associated with pregnancy along with an increase in left atrial pressure increases the risk of thrombus formation, AF and stroke. No maternal deaths were recorded in four large contemporary studies of pregnant women with MS from North America and Europe (total of 300 pregnancies).^{2–5} By contrast, in a sub-Saharan African study of 50 pregnancies in women with heart disease, most of whom had rheumatic MS, maternal mortality was high at 32%.⁷ Perinatal outcome depends on symptoms and pulmonary artery pressure (PAP) during pregnancy.³ The risk is higher in women in NYHA class III/IV during pregnancy with a mortality rate of up to 12%–31%.^{3,11} Prematurity rates are 20–30%, intrauterine growth retardation 5–20% and stillbirth 1–3%.^{2,4}

The goal of management of MS in pregnancy is to avoid cardiac decompensation. ESC guidance¹² on treatment of MS in pregnancy is outlined in Table 1. The management depends mostly on medical therapy. Invasive procedures like PTMC should be considered for pregnant women with acute presentation as in our first case or with significant functional deterioration or refractory pulmonary oedema despite

Table 1. ESC guidelines on management of pregnant woman with mitral stenosis.

- 1) All patients with moderate or severe MS (even when asymptomatic) should be counselled against pregnancy and intervention should be performed pre-pregnancy, favouring percutaneous interventions.
- 2) Clinical and echocardiographic follow-up is indicated monthly or bimonthly depending on haemodynamic tolerance. In mild MS, evaluation is recommended every trimester and prior to delivery.
- 3) When symptoms or pulmonary hypertension (echocardiographically estimated systolic PAP >50 mmHg) develop, activity should be restricted and β 1-selective blockers commenced.
- 4) Diuretics may be used if symptoms persist, avoiding high doses.
- 5) Therapeutic anticoagulation is recommended in the case of paroxysmal or permanent AF, left atrial thrombosis, or prior embolism. It should also be considered in women with moderate or severe MS and spontaneous echocardiographic contrast in the left atrium, large left atrium (≥ 40 mL/m²), low CO, or congestive heart failure because these women are at very high thrombo-embolic risk.
- 6) Percutaneous mitral commissurotomy is preferably performed after 20 weeks' gestation. It should only be considered in women with NYHA class III/IV and/or estimated systolic PAP >50 mmHg at echocardiography despite optimal medical treatment, in the absence of contraindications and if patient characteristics are suitable. It should be performed by an experienced operator. Abdominal lead shielding is recommended. The radiation dose should be minimized by keeping screening time as short as possible.
- 7) Closed commissurotomy remains an alternative in developing countries when percutaneous commissurotomy is not available.
- 8) Open-heart surgery should be reserved for cases in which all other measures fail and the mother's life is threatened.
- 9) Vaginal delivery should be considered in patients with mild MS and in patients with moderate or severe MS in NYHA class I/II without pulmonary hypertension.
- 10) Caesarean section is considered in patients with moderate or severe MS who are in NYHA class III/IV or has pulmonary hypertension despite medical therapy, in whom percutaneous mitral commissurotomy cannot be performed or has failed.

MS: mitral stenosis; PAP: pulmonary artery pressure; NYHA: New York Heart Association.

optimal medical management, provided the valve is suitable for the procedure.^{12–14} In our second case, elective PTMC was indicated as she would likely to decompensate at some point in the third trimester of pregnancy or during labour given her such marked limitation to her exercise tolerance. This was more likely as she developed AF. PTMC is most commonly performed using an Inoue balloon to split the mitral valve commissural fusion. For the percutaneous approach to have optimal outcome, it is ideal that valve is mobile, relatively thin and free of calcium. PTMC is preferred over other surgical treatments for severe MS in pregnancy because of its lower morbidity, short hospital stay, reduced cost and morbidity, avoids thoracotomy and prolonged general anaesthesia.^{15,16} Several studies have reported symptomatic improvement with favourable maternal (mortality <2%) and fetal outcomes (mortality <4%) in women managed with PTMC for severe MS during pregnancy.^{14–20} Jain et al. also showed women who underwent PTMC before pregnancy had a lower incidence of preterm labour compared to pregnant women with non-operated MS; however, there were no significant differences in mode of delivery, mean birth weight or neonatal complications.²¹ The radiation risk associated with the procedure is balanced against the risk of leaving the disease untreated. Specific measures to protect reduce radiation exposure include minimising fluoroscopy, the field of view and reducing the frame rate. Abdominal shielding was considered, but not used based on evidence that placing a lead apron over the pelvis of patients undergoing coronary angiography may actually increase the patient dose by up to two fold due to back-scatter.²² Follow-up upto 17 years has not identified any increase in developmental delay or adverse childhood outcomes following PTMC done during pregnancy.^{18–20} However, this procedure should not be performed in asymptomatic patients as there is a risk of severe mitral regurgitation needing an urgent mitral valve replacement (MVR), fatal arrhythmias and cardiac arrest.^{12,13} Closed commissurotomy remains an alternative in developing countries when PTMC is not available.^{12,13} Open-heart surgery should be reserved for cases with persistent decompensation unresponsive to all treatment modalities as this is associated with a 30% to 40% of fetal mortality rate and upto a 9% maternal mortality rate.^{12–14,23}

Conclusion

PTMC in pregnancy is safe and effective for mothers and preferable for the fetus, when compared with closed commissurotomy or open-heart

surgery. Our cases highlight the importance of maintaining the skills to perform PTMC amongst the UK cardiologists due to large influx of migrants from developing countries. It is likely that we will continue to see these women within the obstetric setting.

A clear understanding of the physiological changes and pathological consequences of MS during pregnancy is needed to allow optimal care and improve maternal and fetal outcome.

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Ethical approval

Written informed consent was obtained from the patients involved in these cases.

Guarantor

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Contributorship

All authors were involved in the care of the patient and the preparation of this manuscript.

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