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PERIPARTUM CARDIOVASCULAR DISEASE MINI-FOCUS ISSUE

BEGINNER

CASE REPORT: CLINICAL CASE

A Cardio-Obstetric Approach to Management of the Complex Pregnant Cardiac Patient

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ABSTRACT

A 23-year-old female presented at 28.5 weeks gestation with symptomatic heart failure due to severe mitral stenosis and severe pulmonary arterial hypertension. After multidisciplinary planning, she underwent caesarean delivery with mitral valve replacement 48 h postpartum. Cardio-obstetric teams provide expert coordinated care for complex cardiovascular disease in pregnancy. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2020;2:86–90)

HISTORY OF PRESENTATION

A 23-year-old G2P0101 female presented at 28.5 weeks gestation with dyspnea, edema, orthopnea, and syncope. Her heart rate was 115 beats/min, respirations were 25/min, blood pressure was 122/83 mm Hg, and oxygen saturation was 98%. She was tachycardic, with 2/6 blowing holosystolic murmur loudest at the apex, soft diastolic rumble heard at the apex, jugular venous pressure of 12 cm, bibasilar crackles, gravid uterus, 2+ bilateral lower extremity pitting edema, and cool extremities.

LEARNING OBJECTIVES

- To recognize the unique challenges in managing cardiovascular disease in the setting of pregnancy.
- To understand the benefit of a cardio-obstetric team approach to management of the pregnant cardiac patient.

PAST MEDICAL HISTORY

Pregnancy 4 years prior was complicated by postpartum dyspnea. Transthoracic echocardiography showed severe mitral stenosis (MS) and moderate mitral regurgitation (MR) secondary to parachute mitral valve (MV). She was subsequently lost to follow-up.

DIFFERENTIAL DIAGNOSIS AND INVESTIGATIONS

Initial concern was for symptomatic severe MS, given presentation at peak hemodynamic load of pregnancy. Laboratory studies are shown in **Table 1**.

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Informed consent was obtained for this case.

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Electrocardiogram showed sinus tachycardia, biatrial enlargement, and rightward axis. Transthoracic echocardiography indicated a normal left ventricular size (ejection fraction 55%), moderate right ventricle enlargement with mild dysfunction, biatrial enlargement, parachute MV with severe MS (mean gradient 15 mm Hg, MV area 0.6 cm^2) and moderate MR, severe tricuspid regurgitation with dilated inferior vena cava, and an estimated pulmonary artery systolic pressure of >100 mm Hg (**Figure 1**). Right heart catheterization showed systemic pulmonary artery pressures, elevated right and left filling pressures, and reduced cardiac output (**Table 2**).

MANAGEMENT

The patient presented with a challenging combination of heart failure due to valvular heart disease and severe pulmonary arterial hypertension (PAH). The modified World Health Organization classification of maternal cardiovascular risk considers both severe MS and severe PAH as class IV risks, or extremely high risk of maternal morbidity and mortality, such that pregnancy is contraindicated (1). Severe PAH independently carries a risk as high as 30% to 50% for maternal mortality (2).

A multidisciplinary approach was taken involving cardiology, maternal-fetal medicine, pulmonary hypertension, interventional cardiology, cardiothoracic surgery, obstetric and cardiothoracic anesthesiology, neonatology, and clinical pharmacology teams. Each team of specialists evaluated the patient independently, communicating urgent findings with the primary team. A comprehensive treatment plan was developed through multidisciplinary face-to-face meetings to optimize maternal health before delivery and maximize maternal and fetal outcomes given the high risk of mortality and morbidity for both (**Figure 2**). Diuretics were administered to reduce cardiopulmonary congestion. Beta-blockers, although the mainstay of treatment for mitral stenosis, were avoided due to acute decompensated right heart failure. Instead, digoxin was given to improve inotropy without exacerbating tachycardia. Three management strategies were contemplated.

The first was pre-delivery valvular intervention (valvuloplasty or percutaneous mitral valve replacement [MVR]). This would urgently improve hemodynamics, permitting a lower risk delivery at term. Percutaneous balloon valvuloplasty has been performed in children with parachute MV (3). However this was not deemed feasible with her anatomy and concomitant MR, necessitating a surgical approach. Though successful pre-delivery surgery potentially would allow her to carry to term, there would be increased risk of perioperative fetal demise and spontaneous preterm labor in the setting of cardiopulmonary bypass (CPB).

The second strategy was preterm delivery by caesarean section followed by immediate MVR. This would eliminate intraoperative fetal risks due to CPB at the expense of prematurity. Immediate MVR could reduce the risk of postpartum hemodynamic decompensation due to anticipated fluid shifts. General anesthesia would be required for delivery to optimize pulmonary and systemic artery pressures, thereby reducing the likelihood of hemodynamic collapse. Additional concerns included risks of prolonged anesthesia on right ventricular dysfunction, ventilation, and post-caesarean bleeding with CPB.

The last strategy considered was preterm delivery by caesarean followed by urgent MVR. MVR would occur within 48 h of delivery, as postpartum fluid shifts begin to occur 48 to 72 h postpartum. This would reduce the duration of general anesthesia and the risk of bleeding. In the event of decompensation during delivery, prophylactic venous and arterial access would be obtained to allow for rapid ECMO cannulation, with cardiac surgery on standby in the operating room.

Given severe pulmonary hypertension and refractory acute heart failure, caesarean delivery was advised, consistent with American Heart Association and European Society of Cardiology guidelines (1,4). She underwent delivery and bilateral tubal ligation under general anesthesia in the cardiac operating room at 29 weeks' gestation followed by bioprosthetic MVR (per patient preference despite medical

ABBREVIATIONS AND ACRONYMS

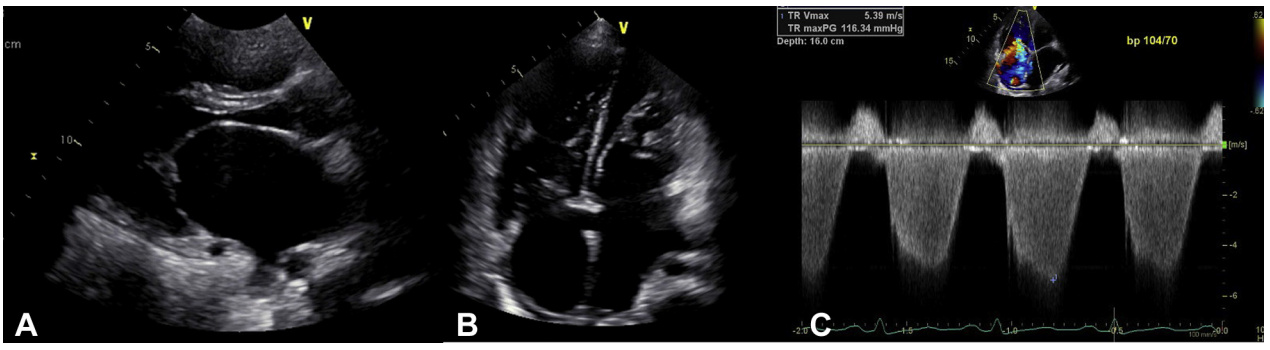
CPB = cardiopulmonary bypass
MS = mitral stenosis
MV = mitral valve
MVR = mitral valve replacement
PA = pulmonary artery
PAH = pulmonary arterial hypertension

TABLE 1 Admission Laboratory Studies

Lab	Result	Reference Range
Sodium, mmol/l	135	135–145
Potassium, mmol/l	4.2	3.3–4.9
BUN, mg/dl	10	8–25
Creatinine, mg/dl	0.67	0.60–1.10
Hemoglobin, mg/dl	11.2	11.9–15.5
Hematocrit, %	34.6	35.6–45.5
WBC, k/mm^3	11.6	3.8–9.9
Platelets, k/mm^3	253	150–400
Total bilirubin, mg/dl	1.1	0.1–1.2
AST, U/l	137	10–45
ALT, U/l	121	7–45
NT-proBNP, pg/ml	1,540	≤300
Troponin I, ng/ml	<0.03	0.00–0.03

ALT = alanine aminotransferase; AST = aspartate aminotransferase; BUN = blood urea nitrogen; NT-proBNP = N-terminal pro-B-type natriuretic peptide; WBC = white blood cells.

FIGURE 1 Selected Frames From the Patient's Echocardiogram



(A) Parasternal long-axis view illustrating her parachute mitral valve. **(B)** Apical 4-chamber view illustrating her parachute mitral valve and right ventricular enlargement. **(C)** Continuous-wave Doppler across the tricuspid valve illustrating suprasystemic estimated pulmonary artery pressures.

recommendation for mechanical MVR) 48 h later without complication. Postoperatively, pulmonary pressures improved but remained elevated, consistent with mixed World Health Organization class I and II PAH. Sildenafil was initiated. She was discharged on postpartum day 12. On follow-up 8 weeks postpartum, she reported NYHA functional class I symptoms and her infant was healthy.

DISCUSSION

Pregnancy-related deaths in the United States are rising, with reported 17.2 deaths per 100,000 live births in 2015 (5). Cardiovascular disease is the leading cause of maternal mortality, accounting for >25% of deaths (6). There is increasing recognition of acquired cardiovascular conditions that impact patients pre-pregnancy, during pregnancy, postpartum, and lifelong (6).

TABLE 2 Right Heart Catheterization Data

End-expiratory pressure, mm Hg	
Right atrium (mean)	18
Right ventricle	112/20
Pulmonary artery	114/77, mean 87
Pulmonary capillary wedge	30
Saturation, %	
Pulmonary arterial	53
Systemic arterial	96
Outputs and resistances	
Fick cardiac output, l/min	3.31
Fick cardiac index, l/min/m^2	2.00
Pulmonary vascular resistance, WU	17.1
Systemic vascular resistance, WU	21.6

Patients with pre-existing or acquired heart disease should be managed by an integrated group with expertise in cardiovascular disease from preconception through the postpartum periods (2,6). This multidisciplinary team-based approach has been integrated into other areas of cardiology (cardio-oncology, valvular disease, advanced heart failure and transplantation) (7-9). Our institution's team-based approach provides comprehensive preconception counseling, pregnancy and delivery management, and postpartum care. Our core team includes cardiologists with expertise in cardio-obstetrics, maternal-fetal medicine specialists, and obstetric and cardiac anesthesiologists. Our comprehensive team includes designated members from the cardiac surgery, pulmonary hypertension, interventional cardiology, family planning, neonatology, psychiatry, and clinical pharmacology divisions, who participate in the patient care team depending on each patient's unique needs. Pregnancy management and delivery plans are developed in monthly multidisciplinary meetings. Mode of delivery is particularly highlighted during planning to reduce the likelihood of unnecessary caesarean delivery (reserved for obstetric indications or high-risk cardiac pathology), to reduce maternal and fetal complications. Preconception plans are also developed (e.g., ensuring appropriate type of valve is chosen) to minimize risk for future desired pregnancies. Post-delivery management includes counseling about future pregnancies and contraception. As we learn about complications of pregnancy such as preeclampsia increasing future risk of premature cardiovascular disease, our integrated approach ensures early patient engagement for prevention and maintenance of long-term health.

FIGURE 2 **Multidisciplinary, Cardio-Obstetric Approach**

Specialty teams' independent goals for this patient's care that were taken into consideration to manage the patient in a "cardio-obstetrics approach." MVR = mitral valve replacement; RV = right ventricle.

CONCLUSIONS

This case demonstrates the complexity of managing maternal cardiovascular disease and the importance of a multidisciplinary approach. Current management guidelines for cardiovascular disease in the nonpregnant patient do not apply in totality, as one must consider both maternal and fetal health and physiologic changes that occur with pregnancy, delivery, and the postpartum period.

Consistent with the Presidential Advisory from the American Heart Association and the American College of Obstetrics and Gynecologists, we urge development of the cardio-obstetric team—a team in which we "bridge the disciplines of cardiology and obstetrics and gynecology, including standardized protocols and enhanced cardiac screening" such that "care can be coordinated to minimize cardiovascular morbidity and mortality and to improve outcomes…by providing a platform for comprehensive well-woman care, primary prevention, and early intervention" (6).

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KEY WORDS **acute heart failure, congenital heart defect, mitral valve, pregnancy, pulmonary hypertension, treatment**