A case of rheumatic heart disease diagnosed in the third trimester

Amanda Manorot,¹ Nina N. Moreira, MD²

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Abstract

Migration from areas with a high incidence of rheumatic heart disease has led to an increase in pregnancy complicated by rheumatic heart disease in high-income countries. We present a case of rheumatic heart disease diagnosed in a 33-year-old G2P0010 French-speaking Congolese woman at 32 weeks gestation. She was initially hospitalized with respiratory syncytial virus (RSV) bronchiolitis at 24 weeks gestation and established care in our clinic. Mitral valve stenosis was identified at 32 weeks gestation after she presented with severe edema and was hospitalized for acute on chronic heart failure complicated by urosepsis and cellulitis. She was managed in the cardiovascular intensive care unit with a subsequent emergent cesarean delivery at 33 weeks gestation for nonreassuring fetal status. Postoperatively, pulmonary artery pressures were 40 mm Hg and left ventricular ejection fraction was 35%. Her condition stabilized and she was discharged home with outpatient cardiology management on postoperative day 10 with baby in the NICU. This case illustrates the importance of a high threshold of suspicion for women at risk for complications of heart disease in pregnancy. A triad of cardiovascular risk screening, patient education and multidisciplinary team planning with maternal-fetal medicine, cardiology, and anesthesiology has been shown to optimize outcomes in women with known cardiovascular disease.

¹University of Iowa Carver College of Medicine, Iowa City, Iowa
²Department of Obstetrics and Gynecology, University of Iowa Carver College of Medicine, Iowa City, Iowa

Introduction

In the United States, 26.5% of pregnancy-related deaths are due to cardiovascular disease, making it the leading cause of death in pregnancy during the postpartum period.¹ As the incidence of pregnancy in women with congenital and acquired heart disease continues to rise, it is imperative to have a high threshold of suspicion for women at risk for maternal heart disease and pregnancy-related mortality and morbidity.² Furthermore, there exists a disparity in pregnancy outcomes complicated by cardiovascular disease...
Rheumatic heart disease in pregnancy

among lower income women. Barriers include lack of pre-pregnancy cardiovascular disease assessment, failure to identify cardiovascular disease risk factors during prenatal care, gaps in high-risk intrapartum care and delay in recognizing cardiovascular disease symptoms in the postpartum period.\(^1\)

In low-income countries, rheumatic heart disease accounts for 90% of all cardiovascular disease in pregnant women.\(^3\) Increased migration from areas with high incidence of rheumatic heart disease has led to an increase in pregnancy complicated by rheumatic heart disease in high-income countries.\(^4\)

We present a case of rheumatic heart disease diagnosed in the third trimester of pregnancy at the University of Iowa Hospital and Clinics.

Case History

A 33-year-old G2P0010 Congolese woman at 24 weeks gestation with type 2 diabetes mellitus and uterine fibroids was transferred from her local intensive care unit 1.5 hours away to our medical intensive care unit for hypoxemic respiratory failure secondary to respiratory syncytial virus (RSV) bronchiolitis. She was managed by internal medicine with oral prednisone and bronchodilators and received antibiotics for a urinary tract infection. After discharge, her prenatal care was transferred to our high-risk obstetrics clinic at 29 weeks gestation.

At 32 weeks gestation, she presented with bilateral lower extremity swelling and tachycardia. She reported 2 weeks of worsening edema that became painful and weeping. Given prior outside echocardiogram showing mitral valve stenosis, she was admitted for acute or chronic heart failure and concern for cellulitis. Transthoracic echocardiogram revealed 45% - 50% ejection fraction, mildly enlarged right atrial chamber, right ventricle, and left atrium. Cardiology identified mitral valve stenosis and moderate tricuspid regurgitation consistent with rheumatic heart disease with elevated right-sided pressures. The mitral valve mean gradient was 7 mm Hg (normal <5 mm Hg). She was medically managed in the cardiac intensive care unit and received antibiotics for cellulitis and E.coli urosepsis. On hospital day 7, fetal heart rate monitoring became nonreassuring and she underwent an emergent cesarean delivery at 33 weeks gestation.

The infant was delivered at 2.73 kg with Apgar scores of 5 at one minute and 9 at five minutes. Postoperatively, she remained intubated on vasopressors with pulmonary artery pressures at 40 mm Hg and LVEF 35%. On postoperative day 3 her ejection fraction returned to 45%. She required 2 units of blood for symptomatic anemia and was discharged in stable condition on postoperative day 10. She denied any knowledge of rheumatic fever as a child or history of heart disease. Her care was complicated by a French Lingala language barrier requiring phone interpretation services.

Discussion

The incidence of rheumatic fever in high-income countries has decreased substantially due to rapid diagnosis and treatment of streptococcal pharyngitis.\(^4\) Rheumatic heart disease may occur following acute rheumatic fever where an immunological response leads to antibodies formed against M proteins of the bacteria. The antibodies cross react
with glycoprotein antigens in the heart, joints, and other tissues. These antibodies lead to inflammation of the heart resulting in progressive fibrosis of the mitral valve. The most common cardiac sequelae of rheumatic heart disease is mitral stenosis.

In low-income countries, rheumatic heart disease comprises 50-88% of heart disease during pregnancy. This is highest among populations in Sub-Saharan Africa occurring in approximately 13 per 1,000,000 pregnancies. Mitral stenosis is usually of rheumatic origin and severity of mitral stenosis is correlated with increase maternal risk for congestive heart failure, pulmonary edema, arrhythmia, and maternal death. Stenotic valves also limit increase in stroke volume reducing uteroplacental perfusion and leading to intrauterine growth restriction, preterm delivery, and stillbirth. Physiologic changes to the cardiovascular system in pregnancy result in increased preload on the heart due to increased blood volume and cardiac output. In the postpartum period, cardiac output remains elevated due to decreased caval compression, autotransfusion from the uterus, and resorption of extracellular fluid into the intravascular compartment. These hemodynamic changes place women at greatest risk and necessitates increased surveillance during pregnancy and the puerperium.

It is important to recognize the disparity that exists in cardiovascular outcomes in pregnancy among lower-income women given our patient has Congolese heritage, primarily spoke French Lingala, and had a significant travel barrier to our tertiary care center. U.S. data shows non-Hispanic black women have a 3.4 increased risk of mortality from cardiovascular pregnancy complications. This disparity can be partially attributed to structural and systemic barriers that prevent the provision of health care. Furthermore, physician implicit and explicit bias can play a role in missed diagnosis. In our case, the language barrier played a significant role in healthcare provision.

The Modified World Health Organization Pregnancy Risk Classification for Women with Pre-existing Cardiovascular Disease classifies women with moderate mitral stenosis as Class III with a significantly increased risk of maternal mortality or severe morbidity. There is a 20-27% maternal cardiac event rate, and the WHO recommends follow up with cardiology every 1-2 months. Women with severe mitral stenosis are classified as WHO Class IV with an extremely high risk of maternal mortality and severe morbidity. The maternal cardiac event rate is >27% and pregnancy is contraindicated. Our patient qualified as WHO Class III risk and due to delay in diagnosis was unable to be established with cardiology per WHO recommendations.

Prior to conception, women with congestive heart failure, NYHA Class > II and moderate or severe mitral stenosis defined as a mitral valve area < 1.5 cm² are candidates for percutaneous balloon mitral valvuloplasty. Asymptomatic women with moderate to severe mitral stenosis should be evaluated for exercise capacity and exercise induced pulmonary hypertension. Women with excellent exercise capacity and normal pulmonary artery pressure may not need surgical intervention. Barriers to timely antenatal care or urgent percutaneous valvuloplasty should be considered when evaluating an asymptomatic woman with
moderate to severe mitral stenosis for percutaneous intervention prior to pregnancy. However, the 2014 American Heart Association / American College of Cardiology recommends preconception percutaneous mitral balloon commissurotomy for asymptomatic patients with a mitral valve area < 1.5 cm².8

Women with mitral stenosis in pregnancy can be managed medically with beta blockers and diuretics with monthly or bimonthly follow up for moderate to severe mitral stenosis per the 2011 European Society of Cardiology guidelines.9 Percutaneous balloon mitral valvuloplasty is an established treatment option in pregnancy with class III and IV heart failure refractory to medical management.10 Surgical intervention is recommended after 20 weeks gestation, but prior to late in the third trimester as the gravid uterus can interfere with femoral access. Time for intervention involves balancing risk of emergent delivery during the procedure as neonatal outcome improves with advanced gestational age versus increased maternal decompensation from severe mitral stenosis leading to emergency delivery compromising both maternal and fetal survival.

In conclusion, it is important to recognize the social determinants of health in cardiovascular disease outcomes during pregnancy and have a high threshold of suspicion for women at risk for complications of heart disease in pregnancy. A triad of cardiovascular risk screening, patient education and multidisciplinary team planning with maternal fetal medicine, cardiology, and anesthesiology has been shown to optimize outcomes in women with known cardiovascular disease.11

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Rheumatic heart disease in pregnancy