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Severe sub-valvular Aortic Stenosis in Late Pregnancy: a multidisciplinary approach

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Introduction

The number of pregnant women presenting with preexisting valvular heart disease has increased during the past few decades. Heart disease complicates 1% of pregnancies (1); however, when present, it significantly increases maternal and fetal adverse outcomes. The incidence of left ventricular outflow tract (LVOT) obstruction in pregnancy is low (2). However, maternal mortality rates for these patients remain alarmingly high, ranging from 5% to 20% for aortic stenosis (3).

Fixed obstruction of LVOT accounts for 8 to 10 percent of all cases of congenital aortic stenosis (4), which, in turn, represents approximately 5.5 percent of all congenital cardiac defects (5). Two types of fixed, discrete obstruction of LVOT have been recognized. (6) The first, a more common type is a membranous diaphragm located at the cranial portion of the LVOT beneath the aortic cusps. The second type is usually described as a thick fibrous ring located more caudally and attached in part to the septal leaflet of the mitral valve. A moderate muscular or fibromuscular obstruction of the LVOT is frequently associated with this latter type of lesion. The left ventricular tunnel represents the extreme form of this malformation. Patients with severe LVOT obstruction are at risk of increased morbidity and mortality when undergoing anesthesia (7). This case report describes the multidisciplinary approach used in the clinical management of a woman with severe subvalvular aortic stenosis who presented with congestive heart failure at 32 weeks of gestation.

Case discussion

We present the case of a 36-year-old female, who presented at 32 weeks of gestation with chief complaints of shortness of breath and chest tightness. She had recently emigrated from Ghana, having received no prenatal care in Ghana or in the US. Further history revealed that she had exertional dyspnea since age thirteen, orthopnea which had worsened during her previous pregnancy, and decreased exercise tolerance. Her previous physician told her that she had a cardiac murmur.

Past medical history included malaria 5 years ago, and there was no significant surgical or family history or cardiovascular disease. She had two previous uneventful pregnancies. The most recent pregnancy was eleven years prior. She was not taking any medications on presentation.

On examination, her blood pressure was 94/67 mm Hg, heart rate 130 beats/minute, and respiratory rate 20/minute. She was afebrile and oxygen saturation 92% on room air. Cardiovascular exam was significant for a 4/6 holosystolic murmur at second intercostal space along midclavicular line, radiating to the apex. There were no bruits and no gallop. The respiratory exam was normal. There was bilateral lower extremity pitting edema. There were no signs of labor.

Laboratory work-up showed anemia, but was otherwise unremarkable. EKG revealed sinus tachycardia and left ventricular hypertrophy. Chest radiograph showed mild pulmonary congestion.

She was admitted to antepartum unit for observation and management of heart failure.

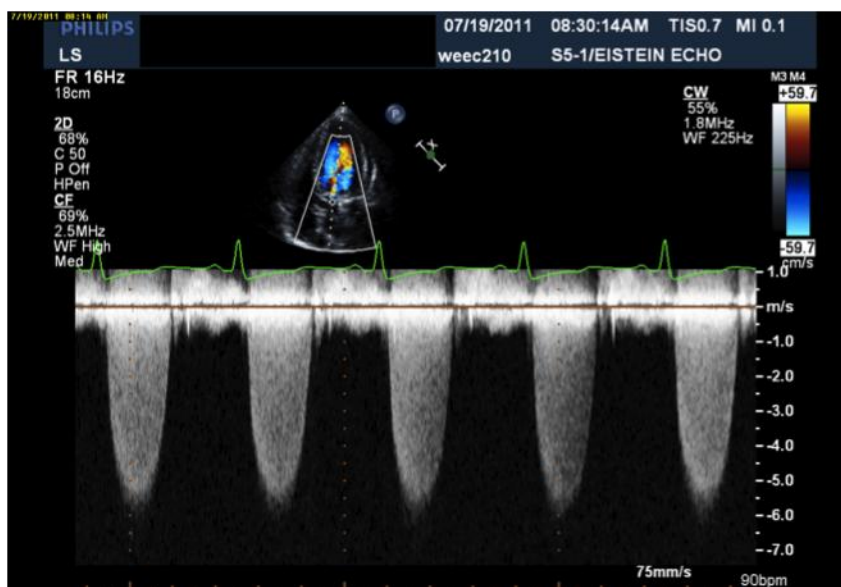
A two dimensional transthoracic echocardiography demonstrated a discrete subaortic membrane (figure 1, 2). There was severe subvalvular LVOT obstruction with a peak instantaneous gradient of 103 mmHg (Figure 3). There was moderate concentric left ventricular hypertrophy with normal left ventricular ejection fraction. Despite normal left ventricular ejection fraction, there was significant decrease in both radial and longitudinal strain on 2D speckle tracking (Figure 4,5).



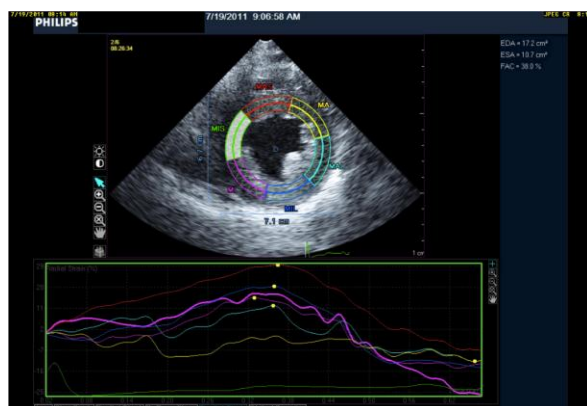
(1)



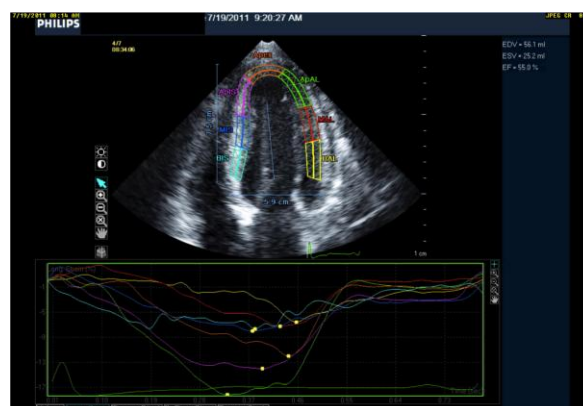
(2)



(3)



(4)



(5)

Corticosteroids were not offered as the benefit to fetus would be minimal, but there would be risk of worsening fluid overload in the mother. Because the patient was relatively unstable with heart failure symptoms, which may progress going further into pregnancy, the team decided that she should deliver the baby as soon as possible. Vaginal delivery is usually safe and C-section is indicated for obstetric reasons. Given patient's multiparous history, it was anticipated that the duration of delivery might be rapid. Under the care of a multidisciplinary team (including high risk obstetrics, anesthesia, cardiology, critical care and CT surgery on the periphery), the patient was induced and had an uneventful delivery. Careful epidural anesthesia and pain control with Demerol was given, avoiding vasodilatory agents because of the Left ventricular outflow tract obstruction. Patients with LVOT obstruction are at high risk for hypotension and reduced coronary perfusion with vasodilatation, as they depend on appropriate venous return (preload).

Other drugs that would increase heart rate and decrease Mean Arterial Pressure such as meperidine and terbutaline were avoided. No endocarditis prophylaxis was given. Both mother and baby did well. Back up for emergency Caesarean section was available. After delivery, she was observed in the CCU. Post-delivery, a repeat echo did not show significant change in the outflow gradients, thus proving that the gradients observed were real and not just pregnancy related changes.

Postpartum, she wanted to go back to Ghana with no intention to follow up. She underwent surgery with successful resection of the discrete sub-aortic membrane. Post op course was uncomplicated and she was discharged from the hospital 4 days after surgery.

She was seen in cardiology clinic a month after delivery, where was found to be doing well. She was then lost to follow up.

Discussion

Hemodynamic changes that occur during pregnancy challenge the functional adaptability of the cardiovascular system in patients with pre-existing cardiomyopathies. The greater metabolic needs of pregnancy are met by changes in blood volume, peripheral vascular resistance, and myocardial function. In women with underlying cardiac disease, however, the demands of pregnancy pose additional stressors that can lead to decompensation, arrhythmias, and rarely, maternal death. Although heart disease is present in 0.5% to 1% of all pregnant women, data from the United Kingdom suggest that a cardiac etiology is the most common cause of death among pregnant women in the developed world ([8])

Women with cardiac disease require a complete pre-conception evaluation and counseling to risk-stratify the maternal and fetal risks of pregnancy without putting the fetus at risk. A detailed history and physical examination, assessment of functional capacity and New York Heart Association (NYHA) functional class, and a 12-lead electrocardiogram are essential. Echocardiography is indicated in most women with a history of valvular or congenital heart disease prior to conception and delivery

A risk score has been developed by Siu et al. to identify predictors associated with the development of unfavorable cardiac events in pregnant women with heart disease and might be used to allow for the establishment of a plan of management for the antepartum, peripartum, and post-partum periods. If a woman has any 1 of the aforementioned poor prognostic factors, the estimated/expected risk is 27%, whereas if she has >1 risk factor, the risk rises to 75%. Poor functional class and left heart obstruction are negative predictors for neonatal outcome. Particularly, a subaortic ventricular outflow tract gradient more than 30 mmHg is a multivariate predictor for adverse neonatal events (1).

Women at elevated risk for adverse events should be managed by a multidisciplinary team at a tertiary care center equipped with the expertise to handle high-risk pregnancies. As part of a

complete risk assessment, plans for monitoring, type of delivery, and anesthetic concerns should be addressed.

This case was remarkable for patient's late presentation and advanced level of obstruction that was previously undiagnosed. These posed significant challenges for clinical decision making on the timing of delivery, mode of delivery, type of anesthesia and monitoring approaches. It is interesting to note that, despite normal left ventricular ejection fraction on echo, there was reduced radial and longitudinal strain on speckle tracking imaging, suggesting the presence of myocardial impairment second to severe pressure overload (9). As its name implies, this approach is based on tracking the motion of small bright spots in the myocardium (speckles) on the gray-scale image as they move during the cardiac cycle. Global and regional longitudinal strain measured with two-dimensional speckle tracking is associated with the global and regional (transmural) extent of scar tissue on contrast-enhanced MRI, allowing for earlier detection of Myocardial dysfunction compared to standard Echocardiographic techniques. (10) The advantages of speckle tracking over Doppler tissue velocities are simpler data acquisition, lack of angle dependence, and direct measurement of strain

Despite that, with a team of high risk obstetrics, cardiology, anesthesiology, cardiothoracic surgery and social services, she did well and had a successful pregnancy outcome. This highlights the importance of a multidisciplinary approach in such cases.

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