

## CLINICAL VIGNETTE

# Postpartum Severe Mitral Regurgitation

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A 50-year-old G5 P3 Caucasian female was evaluated after developing worsening shortness of breath and lower extremity edema two days after delivery of a twin-gestation.

The patient had a history of short cervix and the current pregnancy was a result of in-vitro fertilization. She was admitted to the perinatal unit at 20 weeks of gestation for cervical incompetence and was placed on bedrest. Her hospital course was notable for development of pregnancy-related hypertension.

Prior to pregnancy, she had good exercise tolerance and was able to walk briskly for over an hour as a personal trainer. During this pregnancy, she had noticed a progressive dyspnea with exertion and lower extremity edema starting at around the second month of gestation; this was acutely worsened within 2 days after delivery.

On examination, the patient had a heart rate of 87, blood pressure of 131/78, and oxygen saturation of 95% on 3L nasal cannula. Cardiac examination revealed a JVP 11 cm of water, a holosystolic IV/VI blowing murmur best heard at the apex, and 3+ lower extremity edema bilaterally below the knee. Pulmonary exam revealed that she was mildly tachypneic and had crackles bilaterally 1/3 of the way up her lung fields.

EKG revealed sinus tachycardia. CT angiogram of chest showed pulmonary edema with no evidence of pulmonary embolism. Laboratory values were significant for marginally elevated troponin I of 0.12.

A trans-thoracic echocardiogram showed severe mitral regurgitation with moderate left ventricular dilation (LVDD 62 mm), severe left atrial dilation, moderate pulmonary hypertension with preserved LV systolic function (LVEF 60%). On trans-esophageal echocardiography, severe mitral regurgitation was again observed

with tethering of mitral valves and annular dilation without evidence of leaflet abnormality or papillary muscle or chordae tendinae rupture (figure 1 and 2).

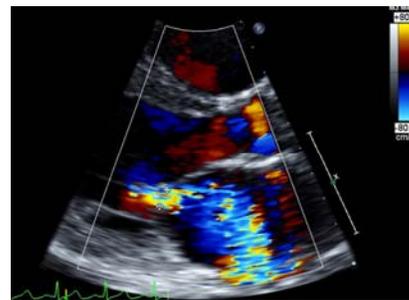


Figure 1

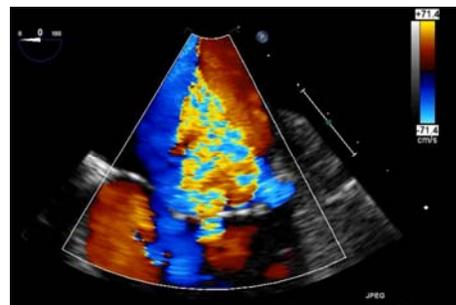


Figure 2

The patient was started on furosemide, an ACE inhibitor, as well as carvedilol once her fluid status improved. She had a dramatic symptomatic improvement and was discharged five days later. An echocardiogram one month later, while she remained asymptomatic, showed an LVEF of 45-50% and persistence of severe MR. At a two-month post-discharge interval, a follow-up echocardiogram revealed improvement of LVEF to 60% with improvement of mitral regurgitation (moderate) and left ventricular dimensions.

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### Mitral regurgitation and pregnancy

Etiologies of mitral regurgitation include 1) Leaflet abnormalities caused by variety of mechanisms including myxomatous degeneration and endocarditis, 2) Papillary muscle and chordae tendinae dysfunction and rupture, and 3) Functional regurgitation secondary to LV annular dilation due to LV dilation or ischemic-related papillary muscle displacement<sup>1</sup>. As expected, many of these etiologies can be seen and have been reported during pregnancy<sup>2,3</sup>.

The patient's TEE did not reveal any evidence of leaflet, papillary muscle or chordae tendinae abnormality; the presence of valve tethering coupled with annular and left ventricular dilation is suggestive of "functional" MR.

### Differential diagnosis of functional MR in pregnancy

#### *Pre-existing MR unmasked by pregnancy*

During a normal pregnancy, cardiac output increases substantially and there is a decrease in systemic vascular resistance, with a 5 to 10 percent increase in the ventricular ejection fraction<sup>4</sup>. There is also reversible cardiac remodeling that is associated with cardiovascular volume overload and that results in the gradual dilatation of all four cardiac chambers with associated functional valvular regurgitation<sup>5</sup>. In the presence of pre-existing asymptomatic (compensated) chronic mitral regurgitation, these changes may account for "unmasking" of the underlying valvular dysfunction<sup>6</sup>.

#### *Mitral regurgitation in the setting of peripartum cardiomyopathy*

Peripartum cardiomyopathy (PPCM) is a poorly understood disorder in which left ventricular dysfunction and symptoms of heart failure occur in the peripartum period. Incidence of PPCM in the United States is 1 in 3000 to 4000 births<sup>7</sup>. The diagnostic criteria are onset of heart failure in the last month of pregnancy or in first 5 months postpartum, absence of determinable cause for cardiac failure, and absence of demonstrable heart disease before the last month of pregnancy<sup>8</sup>. Echocardiographic criteria have been recommended (a left ventricular ejection fraction of less than 45 percent, fractional shortening of less than 30 percent)<sup>9</sup>. Risk factors for PPCM include advanced maternal age, multiparity, African race, twinning, gestational hypertension, and long-term tocolysis<sup>10</sup>.

The signs and symptoms of PPCM often mimic the normal physiologic findings of pregnancy and/or post-partum fatigue and sleep deprivation. Patients may experience pedal edema, dyspnea on exertion, paroxysmal nocturnal dyspnea, persistent cough, abdominal discomfort (from hepatic congestion), dizziness, chest pain, and palpitations<sup>6</sup>.

The outcome of patients with peripartum cardiomyopathy is variable. About 50% of patients with PPCM disorder are reported to recover baseline ventricular function within 6 months of delivery; the initial severity of the left ventricular systolic dysfunction or dilatation is not necessarily predictive of the long-term functional outcome<sup>11</sup>.

Classic criteria for the diagnosis of PPCM limit the diagnosis to the last gestational month and first 5 months after delivery and include the presence of LV systolic dysfunction (LVEF<45%). Our patient had the onset of her symptoms likely prior to the last month of gestation. Despite initially normal ejection fraction at the time evaluation, there was echocardiographic evidence of left ventricular and mitral annular dilation with LV systolic dysfunction noted within one month from diagnosis on repeat TTE. Additionally, several reports and a large database on patients with PPCM challenge the classic criteria by noting that clinical presentation and outcome of patients with pregnancy-associated cardiomyopathy diagnosed earlier than the last gestational month are similar to those of patients with traditional PPCM and suggest that these two conditions may represent a continuum of a spectrum of the same disease<sup>10,12</sup>.

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Submitted on December 10, 2012