Marfan Syndrome and Ascending Aortic Dilation During Pregnancy

Abstract Type: Case Report/Case Series
Jessica N. Rock, M.D.
Medical College of Wisconsin

Introduction: Although aortic dissection associated with pregnancy is rare, it can be catastrophic for mother and fetus. We present the management and course of a parturient with Marfan syndrome and preexisting aortic dilation, factors which increase the risk for aortic complications in pregnancy(1).

Case Report: A 33 year-old G5P1031 was referred for anesthesia consult by the high-risk OB service for a new diagnosis of Marfan syndrome with dilated ascending aorta (max diameter 5.1 cm) and mitral valve prolapsed with minimal regurgitation. Her first pregnancy was complicated only by fetal chromosomal abnormality. The patient had no other significant past medical history. Family history was positive for sudden death of her father in his 30s, cause unknown. Of note, the patient’s height is 79 inches (200 cm).

Cardiology suggested aortic root replacement after termination or during second trimester. However, the patient elected to continue the pregnancy without repair. She was initially placed on strict, outpatient bed rest and beta-blocker therapy to minimize increases in HR and BP, with frequent echocardiograms to follow aortic root diameter. At 29 weeks, she was admitted for inpatient monitoring and proximity to cardiothoracic surgical resources. At 31 weeks, echocardiogram showed slight progression of aortic root dilation, and decision was made to proceed with planned cesarean delivery at 32 weeks.

To avoid rapid changes in cardiovascular status, an epidural was planned. After placement of a radial arterial catheter to guide vasoactive drug administration, an epidural catheter was placed without difficulty. However, the level of surgical blockade was inadequate (T8) despite 45 mL bupivacaine 0.5% over the course of one hour, perhaps due to the patient’s height. After discussion with the surgeon and resolution of the epidural block, a CSE was performed with heavy bupivacaine 0.75% 2 mL, fentanyl, and morphine. Post-SAB hypotension, treated with crystalloid infusion and administration of a total of 300 mcg phenylephrine, lasted less than 5 minutes. The block was adequate and the procedure was uneventful.

The patient had an uneventful postpartum course. She had an uncomplicated aortic repair 5 ½ weeks after delivery.

Discussion: Pregnancy in a patient with Marfan syndrome can pose a management dilemma. Patients with aortic root diameter less than 45mm appear to have minimal or no increased risk of aortic dissection during pregnancy(2); dilation to greater than 50mm or progressive dilation place patients at higher risk. Repair during pregnancy has been successful, but carries a risk of fetal demise. In our patient, beta-blockade, bed rest, and careful monitoring allowed for a delivery that balanced dissection risk and fetal maturity.