Primary Presentation of Pulmonary Hypertension in the Peripartum
Preparing for Patients With Eisenmenger Physiology

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ABSTRACT

A Bosnian woman at 20 weeks gestation presented with dyspnea and hypoxia. She was diagnosed with Eisenmenger physiology with severe pulmonary hypertension, ventricular septal defect, and patent ductus arteriosus. Given high maternal mortality, coordination of care with a multidisciplinary team approach may allow for best possible outcomes. (Level of Difficulty: Intermediate.) (J Am Coll Cardiol Case Rep 2020;2:125–30) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 28-year-old G1P0 immigrant woman from Bosnia without prior medical history presented to her obstetrician’s office at 20 weeks’ gestation with progressive dyspnea. On physical examination, blood pressure was 104/74 mm Hg, heart rate 68 beats/min, and oxygen saturation 85% on room air, prompting transfer to the emergency room for additional evaluation. Physical examination was further notable for cyanosis, clubbing, a right ventricle (RV) heave, loud S2, and a II/VI pansystolic murmur loudest at the left sternal border. Laboratory tests showed a hematocrit of 46% and creatinine of 0.5 mg/dl.

DIFFERENTIAL DIAGNOSIS

The differential for dyspnea and hypoxia during pregnancy includes congestive heart failure, pulmonary embolism, and pulmonary hypertension. The RV heave and loud S2 suggest pulmonary hypertension, and the pansystolic murmur suggests a ventricular septal defect (VSD), in combination concerning for a hypoxic shunt due to previously undiagnosed congenital heart disease. A transthoracic echocardiogram would assess for the presence of any structural cardiac abnormalities and estimate pulmonary pressures. It also would be essential to rule out a pulmonary embolism with a computed tomography angiogram.

INVESTIGATIONS

Transthoracic echocardiogram revealed severe RV dilation and hypertrophy (Figure 1, Video 1) with an S’ of 9 cm/s and tricuspid annular plane systolic excursion of 2.5 cm, severe pulmonary hypertension with an estimated pulmonary artery (PA) systolic pressure of 90 mm Hg, a patent ductus arteriosus (Figure 2, Video 2),
and a membranous VSD (Videos 3 and 4). A computed tomography angiogram of the chest showed a dilated PA, but no pulmonary embolism (Figure 3). Fetal ultrasound demonstrated severe intrauterine fetal growth restriction, oligohydramnios, and fetal decelerations.

Right heart catheterization demonstrated severe pulmonary hypertension with normal right- and left-sided filling pressures: right atrium pressure 4 mm Hg, RV 111/4 mm Hg, PA 110/54 mm Hg (mean 76 mm Hg), pulmonary capillary wedge pressure 2 mm Hg, a calculated pulmonary vascular resistance of 12 WU, and a thermal cardiac output of 5.3 l/min. Systemic blood pressure was 100/70 mm Hg. On shunt run, oxygen saturation was as follows: superior vena cava 64%, right atrium 67%, RV 67%, PA 71%, and femoral artery 88% (Figure 4).

MANAGEMENT

Given the severity of PA hypertension and significant right to left shunting in this patient with complex congenital heart disease (World Health Organization class III), she was started on intravenous epoprostenol to lower her pulmonary vascular resistance. Recognizing that epoprostenol is generally contraindicated in pregnancy, we prioritized maternal stabilization, as discussions for therapeutic abortion were ongoing. Intracardiac monitoring proved difficult as her PA catheter repeatedly prolapsed into the RV and ultimately necessitated removal. Shortly thereafter, there was spontaneous intrauterine fetal demise. With concern for placental abruption, bleeding, and disseminated intravascular coagulation, venous thromboembolism prophylaxis was initially held, but restarted on day 5 of admission.

A multidisciplinary team, including cardiac intensivists, advanced heart failure specialists, maternal fetal medicine, family planning gynecologists, cardiac anesthesiologists, and cardiac surgeons discussed options for delivery. Although possible to support patients with Eisenmenger syndrome through delivery, special care needs to be taken when considering anesthetic options. Anesthetic agents, such as propofol and etomidate, cause myocardial depression and reduction in systemic vascular resistance. This, in conjunction with positive pressure ventilation, can reduce venous return, increase pulmonary vascular resistance, and exacerbate right to left shunting. Slow-onset epidural anesthesia may be safe (1); however, the risk of reducing preload persists.
The most dangerous time periods for mothers are during delivery and early postpartum (2). With the increase in blood return, potential blood loss, as well as hypercoagulable state, postpartum complications include worsening shunt, hypovolemia, thromboembolism, and preeclampsia (3).

A collaborative decision was made to proceed with dilation and evacuation in the operating room with light conscious sedation and local cervical anesthetic. Importantly, a Bosnian translator was requested for the
procedure to facilitate communication, lessen the patient’s anxiety, and reduce sedation requirements. In anticipation of decline in systemic vascular resistance, low-dose vasopressin would be instituted prophylactically with target parameters of both systemic pressure and maintenance of baseline oxygen saturation. Plans were made for extracorporeal membrane oxygenation (ECMO) place holders for emergent cannulation should the patient develop hemodynamic compromise.

The patient initially tolerated the dilation and evacuation well with only 50 ml of blood loss. Immediately following the procedure, while still in the operating room, she became acutely hypotensive and hypoxic, which progressed to pulseless electrical activity arrest requiring approximately 10 min of cardiopulmonary resuscitation. She was intubated and rapidly cannulated for veno-arterial ECMO support. On emergent transesophageal echocardiogram, she was found to have a thrombus in the inferior vena cava extending into the RV and across the VSD into the aortic root, consistent with a clot in transit. She was stabilized in the operating room with administration of bolus epinephrine and epinephrine drip, vasopressin drip, bicarb, and calcium chloride. She also received aggressive fluid resuscitation, including 2 l of plasmalyte, 4 U of packed red blood cells, fresh frozen plasma, and platelets. She was then transferred to the cardiothoracic intensive care unit. Within 24 h, she exhibited intact neurologic function and was communicating with family.

DISCUSSION

Management of Eisenmenger syndrome during pregnancy is challenging, carrying a high risk of fetal demise and maternal mortality (4) and is an absolute contraindication to pregnancy (5). Because of a fixed and elevated pulmonary vascular resistance, normal physiologic changes during pregnancy, such as the increase in blood volume and decrease in systemic vascular resistance, are not well tolerated. These changes may cause right to left shunt in patients with previously compensated congenital heart abnormalities, resulting in signs of Eisenmenger syndrome, including hypoxia, cyanosis, and right heart failure. During labor, uterine contractions and Valsalva maneuvers contribute to potentially dangerous fluctuations in preload, afterload, and cardiac output, necessitating cesarean delivery for those with severe pulmonary hypertension. In patients with Eisenmenger syndrome who do become pregnant, termination of the pregnancy is recommended.

Treatment for Eisenmenger syndrome during pregnancy focuses on balancing shunting to optimize blood flow and oxygenation. Diuretics can be used with caution to relieve right heart failure. Pulmonary vasodilators, such as nitric oxide, prostanoids, endothelin receptor antagonists, or phosphodiesterase-5 inhibitors, may improve oxygenation, improve maternal stability until delivery, and decrease mortality (6). Although it is not
standard of care, prophylactic anticoagulation in the peripartum period may be indicated given the increased risk of venous thromboembolism. Anticoagulation should be used cautiously in patients with risk of bleeding, thrombocytopenia, or coagulopathy (2,7). Case reports of both maternal and fetal survival highlight various management strategies, including ECMO, to provide hemodynamic support through delivery (8).

In our case, collaboration across different specialties was essential in coordinating our patient’s care. Through multidisciplinary discussions, we developed a management plan that optimized her outcome to the best of our ability. We anticipated the need for hemodynamic support during her dilation and evacuation procedure and had positioned ECMO placeholders before the procedure. The patient’s intraoperative hemodynamic collapse was likely multifactorial, but at least in part due to reduced systemic cardiac output from the thrombus preventing flow across her VSD. The patient’s pregnant state, immobility, and lack of consistent venous thromboembolism prophylaxis during her admission significantly increased her risk to develop a venous thromboembolism. Although amniotic fluid embolus would generally have been in the differential diagnosis, this appeared very unlikely given the marked oligohydramnios noted on fetal ultrasound. Initiating ECMO allowed us to bridge the patient beyond the immediate postpartum period.

**FOLLOW-UP**

With epoprostenol titration, the patient weaned from veno-arterial ECMO to veno-venous ECMO. She then was extubated and subsequently decannulated from veno-venous ECMO 2.5 weeks after her procedure. Despite titration of epoprostenol, she had persistently high oxygen requirements and was unable to leave the cardiothoracic intensive care unit. The advanced heart failure team considered heart/lung transplantation as the only definitive therapy; however, she was deemed not to be a candidate because of continued functional decline as well as other concerning social factors. She ultimately transitioned to hospice care and died shortly thereafter.
CONCLUSIONS

Eisenmenger syndrome carries a high fetal and maternal mortality. Coordination of care with a multidisciplinary team approach in combination with vasodilators and ECMO support may allow for best possible outcomes.

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KEY WORDS anesthesia, congenital heart defect, hemodynamics, pregnancy, pulmonary hypertension

APPENDIX For supplemental videos, please see the online version of this paper.