

Multidisciplinary Management of Peripartum Cardiomyopathy During Repeat Cesarean Delivery: A Case Report

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Peripartum cardiomyopathy is a potentially fatal form of heart failure associated with pregnancy. A 29-year-old African American woman, gravida 3, para 2, at 36 weeks' gestation had a history of cardiomyopathy, morbid obesity (body mass index > 70 kg/m²), uncontrolled hypertension, obstructive sleep apnea, and required a repeat cesarean delivery. The patient was admitted to the hospital several times throughout her pregnancy for congestive heart failure, pulmonary edema, and headaches. Two years previously the patient received a diagnosis of peripartum cardiomy-

opathy 3 weeks after the delivery of her second child.

This case report illustrates the recognition of peripartum cardiomyopathy and the risks early in pregnancy. It also describes the appropriate medical management, including transesophageal echocardiography and the need for collaboration of multiple medical specialists before and during delivery to provide the best possible outcome for both mother and infant.

Keywords: Awake intubation, cardiomyopathy, cesarean delivery, peripartum cardiomyopathy, pregnancy.

Peripartum cardiomyopathy (PPCM) is a potentially fatal form of heart failure associated with pregnancy. The worldwide incidence of PPCM is approximately 1 in 3,000 to 1 in 15,000 pregnancies.¹ The classic diagnostic criteria were established by Demakis and Rahimtoola² and Demakis et al,³ who limited the diagnosis to the last gestational month and the first 5 months after delivery. However, Hibbard et al⁴ describes PPCM earlier during pregnancy. Clinical presentation and outcome of patients with pregnancy-associated cardiomyopathy diagnosed early in pregnancy are similar to those of patients with traditional PPCM. These 2 conditions may represent a continuum of the same disease.⁵

Echocardiography, in addition to being an important diagnostic tool in the discovery and quantification of PPCM, may provide prognostic information with regard to recovery of cardiac function. The 25% to 50% mortality rate for PPCM and the long-term prognosis are directly related to left ventricular (LV) dilation and dysfunction.⁶

We present a case report of a patient with PPCM, managed by a multidisciplinary team consisting of a cardiologist, pulmonologist, perinatologist, neonatologist, obstetrician, and anesthesia providers. Collaborative treatment, including transesophageal echocardiography (TEE), reduced the risk of an emergency cesarean delivery and optimized maternal health preoperatively. The multiple specialists also coordinated her intraoperative and postoperative care. Equally important was the coordination of care for the fetus, thereby providing the safest possible outcome for this pregnancy.

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Case Report

A 29-year-old African-American woman (gravida 3, para 2) presented for obstetric care at 5 weeks' gestation. Her medical history consisted of poorly controlled hypertension, cardiomyopathy, congestive heart failure, morbid obesity (body mass index > 70 kg/m²), height of 160 cm, greatest weight of 189 kg, and obstructive sleep apnea. Her history also included previous tobacco use and resolved renal insufficiency. Although her first cesarean delivery was uneventful, her second cesarean delivery was complicated by congestive heart failure (CHF) 3 weeks postpartum, requiring prolonged hospital admission. The echocardiogram at that time revealed an ejection fraction (EF) less than 40%, concentric LV hypertrophy, mildly decreased LV contractility, left atrial enlargement, mild mitral valve regurgitation, trace aortic insufficiency, tricuspid regurgitation, pulmonary insufficiency, and normal right atrial and ventricular function. On the basis of the findings, a diagnosis of PPCM was made. Because cardiomyopathy accounts for an increasing proportion of reported pregnancy-related deaths and a 6-fold excess risk of death,⁷ another pregnancy was strongly discouraged. Subsequent pregnancies carry a high risk of LV deterioration and progressive heart failure. Therefore, the patient consented to a repeat cesarean delivery and bilateral tubal ligation.

At 32 weeks' gestation the patient complained of headaches and shortness of breath at rest, as well as

marked shortness of breath when her back was lowered from a sitting position to 45 degrees. She was admitted because of diuresis, hypertension, pulmonary edema, and CHF. Because the patient was initially unstable, with an oxygen saturation of 88% to 90% when breathing room air, she was admitted into the intensive care unit (ICU). In the ICU her medical condition was optimized by continuous positive airway pressure (CPAP) at 6 to 14 cm of H₂O, oxygen supplementation, and the following oral medications: furosemide, 80 mg daily; carvedilol, 12.5 mg twice a day; and hydralazine, 25 mg twice a day. The patient was evaluated by clinicians from the departments of cardiology, pulmonology, neurology (for her complaints of headaches), obstetrics, anesthesia, neonatology, and perinatology during her hospital stay. She also received a triple course of 12-mg betamethasone intramuscular injections each day. An amniocentesis at 34 weeks failed to show fetal pulmonary maturity, as the lecithin/sphingomyelin ratio (L/S ratio) was below 1.5 and the amounts of phosphatidyl glycerol were absent (mature = L/S ratio > 1.9 and phosphatidyl glycerol present).

She was hospitalized until 35 weeks, then discharged to home, at her request to be with her other children. At discharge, she had optimized cardiac and pulmonary status and improved echocardiographic findings, an EF of 55%, and lack of shortness of breath. Scheduled amniocentesis was performed every 10 to 14 days until fetal lung maturity was confirmed at 36 weeks with a surfactant-to-albumin assay of 55 mg/g, an L/S ratio greater than 1.9, and presence of phosphatidyl glycerol.

For 1 week the patient was stable at home, with a functional capacity described by the New York Heart Association Classification as class II/III. (Class II indicates patients with slight, mild limitation of activity; they are comfortable with rest or with mild exertion. Class III indicates patients with marked limitation of activity; they are comfortable only at rest.) A perinatologist who confirmed and documented reassuring fetal heart rate and electrocardiogram (ECG) tracings assessed her once a week in the clinic.

The goal of the multidisciplinary team was to find a therapeutic window when the patient was optimized for a cesarean delivery and to prevent an emergency cesarean delivery. On admission the day before her scheduled cesarean delivery, a preoperative current assessment by anesthesia and cardiology professionals revealed orthopnea requiring 3 pillows to breathe comfortably, 2+ peripheral edema, and breath sounds with crackles and rales noted at the bases and distant bilaterally. Hemoglobin saturation with oxygen ranged from 88% to 94% on room air and increased to 99% when CPAP and fraction of inspired oxygen (FiO₂) 30% oxygen were administered. It was determined that the patient had a class 3 Mallampati, 2-finger-width thyromental distance, large neck circumference, small mouth, large tongue, and ex-

cessive soft tissue. The preoperative echocardiogram was technically difficult to perform because of the patient's habitus, but did reveal hyperdynamic LV systolic function with EF 45% and concentric LV hypertrophy consistent with hypertensive heart disease. Cardiology consultants recommended continuation of CPAP and the following oral medications: hydralazine, 25 mg every 8 hours; carvedilol, 25 mg twice daily; and furosemide, 60 mg/d. Other oral medications included ferrous sulfate twice daily, 500 mg of vitamin C, and a prenatal multivitamin. Thrombosis prophylaxis was initiated using 7,500 U of heparin subcutaneously twice daily.

Frequent patient-staff discussions kept the patient highly motivated and compliant. An 18-g intravenous (IV) catheter was placed in the left external jugular vein the night before surgery. She had an elevated serum carbon dioxide level at 31 mmol/L (31 mEq/L) (normal range, 22-30 mmol/L) and a depressed serum sodium level at 132 mEq/L (132 mmol/L) (normal range, 137-145 mmol/L). All other electrolytes, the complete blood cell count, liver enzymes, and coagulation profile were normal. The patient was typed and crossed for 4 U of blood and had no food or water by mouth after midnight before the planned surgery.

The patient arrived in the operating suite holding area, where all questions, concerns, and alternatives were again fully discussed before she proceeded to the operating room. The patient consented to awake fiberoptic intubation, radial arterial line, an epidural block for postoperative pain relief, and a central line for central venous pressures. This would also allow a pulmonary artery catheter to be placed if the patient's condition decompensated during or after surgery. Citric acid and sodium citrate (Bicitra, 30 mL) was administered on transfer to the operating room.

In the operating room, the patient was monitored using ECG leads II and V, pulse oximetry, a properly fitting blood pressure cuff, and external fetal heart tone monitoring. The patient was prepared for fiberoptic intubation by the intramuscular administration of glycopyrrolate (0.4 mg) 30 minutes before the operation and by inhalation of nebulized 4% lidocaine. Additionally, the posterior pharynx was treated with 4% tetracaine. Oxygen was administered by face mask, along with midazolam, 0.5 mg, IV. While the patient was in the sitting position, a lumbar epidural catheter was placed, a test dose of 1.5% lidocaine with epinephrine at a 1:200,000 concentration was administered, and the catheter was secured. A left radial arterial line was placed; then the patient's trachea was intubated with a 7.5-mm endotracheal tube, over a fiberoptic bronchoscope, while she still was in a sitting position. Once tube placement was verified by repetitive and characteristic waveform of capnography in addition to auscultation of breath sounds bilaterally, propofol at 150 mg IV (based on ideal body

weight) was titrated to effect, and the patient's ventilation was controlled. Sevoflurane 1% end-tidal was administered for anesthesia. Once supine, the patient was placed in the left lateral uterine displacement position. The fetal heart rate was 120/min. The mother's vital signs were stable: blood pressures, 120 to 150 mm Hg systolic and 70 to 80 mm Hg diastolic; heart rate, 70 to 90/min, and oxygen saturation, 96% to 99%. The fetal heart tones sounded healthy, and the ECG tracing appeared normal.

A right internal jugular 8.5 French sheath was placed, secured, and transduced. To assist in monitoring the depth of anesthesia and to reduce the chance of recall, a bispectral index (BIS) monitor was placed on the patient's forehead. The pannus was elevated and taped on 2 right-angle posts secured at the head of the bed to allow better visualization of the suprapubic area where the incision was planned. While the abdomen was being prepared and draped, the anesthesia team placed a TEE probe and assessed the patient's cardiac function. The TEE revealed moderate LV hypertrophy, mild left and right arterial dilation, EF 40%, and moderate pulmonary hypertension. The TEE probe was left in place throughout the cesarean delivery. Furosemide, 40 mg, was administered IV to maintain the central venous pressure between 12 and 14 mm Hg. Total anesthesia time before incision was 45 minutes.

The neonatal resuscitation team consisting of a neonatologist and an advanced practice nurse had been informed that the newborn would deliver with significant depression by the inhaled anesthetic, so they were prepared for the expected need to ventilate the newborn. Twenty-five minutes after skin incision, a baby girl was delivered. Her Apgar scores of 4 at 1 minute and 9 at 5 minutes were consistent with the expected neonatal anesthetic effect. Once the placenta was delivered, 20 U of IV pitocin in 1,000 mL lactated Ringer's solution was administered at 100 mL/h, without much cardiac effect. The epidural catheter was dosed with 1.5% lidocaine with epinephrine at 1:200,000 (second test dose), then a 0.1% bupivacaine drip with fentanyl, 2 µg/mL, was started at 12 mL/h. The epidural was not used until the baby was delivered to prevent any drop in blood pressures. Total blood loss was 800 mL, and the patient did not require a transfusion. The entire case from incision to closure was 2 hours and 20 minutes, with an anesthesia time of 3 hours and 50 minutes. The last echocardiogram reading in the operating room revealed no change in the EF of 40%, with moderate LV function. Her vital signs throughout the case were as follows: blood pressures, 110 to 150 over 70 to 80 mm Hg; heart rate, 75 to 100/min; and BIS, 40 to 45. Breath sounds remained clear and free from crackles or rales, oxygen saturation was 96% to 99% with FiO₂ of 60%, and the administration of diuretics was well tolerated.

The patient was left intubated and sedated in the ICU overnight to ensure adequate ventilation, hemodynamic stability, and adequate pain control before extubation. On

the first postoperative day, the patient remained hemodynamically stable, with effective postoperative epidural analgesia, and the patient was extubated without incident. Early mobilization was encouraged, and aggressive chest physiotherapy and pain control were maintained.

The patient's oral medications were reintroduced along with thrombosis prophylaxis. Members of the anesthesia, cardiology, medicine, and obstetrics departments continued to follow up the patient throughout her hospital stay. The patient was transferred out of the ICU to the postpartum unit on postoperative day 3, where she was first able to be with her baby. On postoperative day 5 the patient was discharged to home in stable condition. Her cardiologist and obstetrician continued to follow up the patient.

The patient had impaired ventricular function as a result of hypertension and previous cardiomyopathy, and her symptoms were exacerbated, as evidenced by shortness of breath, EF less than 40%, and poor activity level during her pregnancy. Her symptoms, however, resolved shortly after delivery. An echocardiogram was planned for 6 months postpartum.

Discussion

• *Diagnosis/Presentation.* Making the proper diagnosis is the initial challenge in caring for patients with PPCM. Although there is extensive overlap of symptoms with the normal postpartum patient, the diagnosis of PPCM is made when these symptoms are more pronounced. Although undiagnosed and spontaneously resolving cases of PPCM occur, patients with preexisting diseases such as obesity, hypertension, or orthopnea typically present with marked symptoms of heart failure. Symptoms include fatigue, shortness of breath, CHF, nocturia, and palpitations. Radiographic assessments may reveal an enlarged heart, pulmonary congestion, pericardial and pleural effusions, or signs of pulmonary embolism. Because these symptoms are common to many other diseases, it is important to rule out all other possible causes before diagnosing PPCM. The radiographic finding of LV systolic dysfunction by echocardiogram is an important criterion for making the diagnosis. The National Heart, Lung, and Blood Institute and the Office of Rare Diseases of the National Institutes of Health have recommended strict echocardiographic criteria of LV dysfunction: EF less than 45%, or M-mode fractional shortening less than 30%, or both, and end-diastolic dimension more than 2.7 cm/m².^{8,9} In addition to being an important diagnostic tool in PPCM, echocardiography may provide prognostic information with regard to recovery of cardiac function.¹⁰

• *Medical Management.* Medical therapy is the standard treatment of heart failure due to systolic dysfunction; however, presentation of heart failure in women who are going to be delivering presents unique challenges. Salt intake is restricted, and loop diuretics are

used to decrease volume overload and pulmonary congestion. In patients with systolic dysfunction, afterload is usually reduced with vasodilation and is achieved with nitrates. Hydralazine for blood pressure control appears to be reasonably safe for the fetus, although a few cases of fetal thrombocytopenia have been reported. In the postpartum period, an angiotensin-converting enzyme (ACE) inhibitor and digoxin can then be added to the regimen. For women with hypertension and heart failure symptoms, β -blocker therapy has also safely been prescribed. Management goals include preload optimization, afterload reduction, and increased contractility.

As in other forms of heart failure, PPCM can lead to both thrombotic and thromboembolic complications; therefore, anticoagulation should be considered. To this end, subcutaneous heparin administration with or without the patient wearing compressive stockings during the hospital stay is recommended. For those patients with clinical evidence of deep venous thromboses postpartum, warfarin therapy may be initiated. The need for long-term anticoagulation therapy should then be reassessed depending on the state of LV recovery.

- **Anesthetic Management.** Caesarean delivery avoids the increase in stroke volume and cardiac output that uterine contractions produce. Trouton et al¹¹ proposed that cesarean delivery with the patient under general anesthesia should be used for parturients who showed impaired LV function following acute myocardial infarction, since rapid infusion of fluids to epidural-related hypotension could worsen the performance of the impaired left ventricle. Others argue that regional anesthetics may be used because of the decreased cardiac sympathetic response and the potential to elicit symptoms.^{12,13}

Our patient, however, had the risk of aspiration, a probable difficult intubation, the potential for acute cardiac decompensation, and the inability to tolerate a lengthy repeat cesarean delivery. Our patient could not tolerate lying flat for the procedure because of her pulmonary dysfunction, and thus being awake for delivery was not possible. As a result of these considerations, we chose an awake fiberoptic intubation to secure the patient's airway and placement of a lumbar epidural catheter for perioperative pain control, with small increments of lidocaine and a continuous infusion of bupivacaine once the baby was delivered. With cardiac function impaired, invasive monitoring became very useful for the management of the patient's hemodynamic status. An arterial line for minute-to-minute blood pressure measurements and laboratory blood draws, and a central venous catheter, which could accommodate a pulmonary artery catheter if needed, allowed management of the patient's cardiac response to the procedure. When indicated, the anesthetist may need to infuse inotropic or vasoactive drugs. Agents such as nitroglycerin or nitroprusside, dopamine, dobutamine, or milrinone should be readily

available for titration. Because we anticipated that general anesthesia would cause neonatal depression, the neonatal resuscitation team was present in the operating room and ready to care for the baby upon delivery and the neonatal intensive care unit was prepared for the baby's arrival.

We elected to place a TEE probe at the beginning of the surgical procedure to assess the ventricular function, volume status, pulmonary hypertension, and any possible valvular dysfunction. The probe remained in position until the end of the procedure. Although no cardiac medication was necessary during our case, TEE is very helpful in assessing the response to drug and fluid therapy. As in patients with PPCM, TEE provides accurate, real-time, structural, and physiologic data in patients undergoing operations—information that is not obtained by conventional intraoperative monitors. Information obtained from TEE may directly alter surgical management and anesthetic management in approximately 30% and 50% of patients, respectively.¹⁴

Conclusion

A coordinated plan for this patient's cesarean section was initiated. Appropriate staff was present during the delivery of the baby including the neonatal specialists to care for the infant. Multiple anesthetists collaborated in the operating suite to safely anesthetize and monitor the patient. The obstetricians and perinatologist worked together on the best approach for the patient's peripartum management and operation. Cardiologists and pulmonologists gave their input on how to aggressively treat the PPCM symptoms. This case illustrates the prompt recognition of the risk of PPCM in pregnancy, the initiation of appropriate medical management, and the collaboration with multiple disciplines for delivery, all of which are essential to provide the best possible outcome for both mother and infant.

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