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Peripartum anesthetic management of a patient with Dilated Cardiomyopathy secondary to pathogenic RNA-binding motif protein 20 variant

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Background

- Pathogenic RNA-binding motif protein 20 (RBM20) variant is a gene that codes for post-translational slicing in sarcomeres and causes cardiac fibrosis
- This gene has been linked to non-ischemic forms of dilated cardiomyopathy (DCM)
- Due to advancements in treatment, more women with hereditary forms of DCM are surviving to the reproductive years
- As a result, DCM is an increasing cause of pregnancy-related mortality in the United States ^[1]
- Parturients with DCM offer a unique challenge to obstetric anesthesiologists regarding labor analgesia and intraoperative management in cesarean deliveries

Case Presentation

- 32-year-old G1P0 at 32-6 weeks gestation presented for induction of labor for worsening orthopnea
- mWHO class IV, NYHA class II secondary to non-ischemic DCM caused by RBM20 variant
- Diagnosed at age 19 with serial transthoracic echocardiograms displaying EF of 45% without right heart strain
- 1st trimester cardiac MRI revealed severely dilated left ventricle with moderately reduced systolic function
- 3rd trimester TTE showed LVEF 40% with moderate mitral valve regurgitation

[1] Ituk, U. S., Habib, A. S., Polin, C. M., & Allen, T. K. (2015). Anesthetic management and outcomes of parturients with dilated cardiomyopathy in an academic centre. *Canadian journal of anaesthesia = Journal canadien d'anesthésie*, 62(3), 278–288.



Preoperative Management

- A slow induction of labor was planned and initiated with a Foley balloon and vaginal misoprostol
- This was complicated by tachysystole, fetal variable decelerations and a sustained three-minute deceleration
- Foley bulb was removed, and vaginal lavage improved her strip however, cesarean section was offered to the patient at this time
- Radial arterial and epidural lines were placed
- Lower extremity arterial duplex study was performed to assist in femoral access if ECMO was required
- Epidural slowly titrated to the T4 level using plain 2% lidocaine without epinephrine

Intraoperative Management

- Patient was administered 10 mg oxytocin IM after fetal delivery rather than the IV form, which requires an additional 500 cc of crystalloid administration
- Point of care ultrasound (POCUS) was essential in guiding fluid management and choice of vasopressor
- End of case POCUS revealed a severely dilated and hypokinetic left ventricle despite the patient receiving only 300 cc of IV crystalloid fluid throughout the case



Figure 1. TTE, apical four chamber view, showing a severely dilated left ventricle

- Patient was given 10 mg IV furosemide and her cardiac function improved

Postoperative Management

- Patient was transferred to the cardiovascular ICU for 24 hours of post-operative monitoring
- Her recovery was uneventful
- Patient was discharged home on post-operative day five in stable condition and continued on goal-directed therapy consisting of:
 - Enalapril
 - Metoprolol succinate
 - Spironolactone

OUTCOMES

- This case illustrates an example of favorable outcomes after primary cesarean section under neuraxial anesthesia in a patient with severe systolic dysfunction secondary to DCM
- The optimal anesthetic management of patients with DCM undergoing cesarean section requires ^[2]:
 - Thorough preoperative assessment
 - Vigilant perioperative and postoperative monitoring
 - Suitable anesthetic catered to the underlying differences from normal physiology
 - Optimization of fluid management
 - Stable hemodynamic status
- Given our patient was found to have a severely hypokinetic left ventricle on POCUS examination, the optimal vasoactive agent of choice for hypotension would be epinephrine, which provides inotropic support via β -adrenergic agonism leading to improved myocardial contractility
- On the contrary, phenylephrine would cause an acute increase in afterload ultimately worsening cardiac output

DISCUSSION

- In patients with DCM, epidural anesthesia is preferred over spinal anesthesia due to the ability to better control hemodynamic changes as the onset of blockade is slower and redosing is easy
- The hemodynamic state in DCM includes increased preload, increased afterload, decreased contractility, increased to normal heart rate and equivocal pulmonary vascular resistance ^[3]
- Plain 2% lidocaine without epinephrine was used to avoid further increases in heart rate, preload and afterload
- By carefully titrating anesthetic and analgesic doses, obstetric anesthesiologists can judiciously and appropriately supplement volume, as necessary.
- Though parturients with dcm are prone to hypotension because of neuraxial anesthesia, venous pooling, prior beta-adrenergic blockade and diuretic therapy, prophylactic use of vasopressors and intravascular volume loading are best avoided ^[3]
- Point of care ultrasound can be useful in guiding management to avoid excessive administration of volume and proper selection of vasoactive agent



[2] Chen, C. Q., Wang, X., Zhang, J., & Zhu, S. M. (2017). Anesthetic management of patients with dilated cardiomyopathy for noncardiac surgery. European review for medical and pharmacological sciences, 21(3), 627–634.

[3] Choudhury M. (2018). Neuraxial anaesthesia in parturient with cardiac disease. Indian journal of anaesthesia, 62(9), 682–690. https://doi.org/10.4103/ija.IJA_474_18