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Multidisciplinary Approach to Delivery of a Preeclamptic Parturient with Severe Left Ventricular Noncompaction

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Introduction: Left ventricular non-compaction (LVNC) is characterized by spongiform left ventricular trabeculae, and deep intertrabecular recesses. A thickened myocardial wall with a thick endocardial layer and a thin, compacted epicardial layer are thought to originate by arrest of normal embryogenesis. Though widely under diagnosed, the prevalence of LVNC ranges at approx. 0.05% to 0.24% with men reportedly more affected than women. In 2006, LVNC was classified as a primary cardiomyopathy of genetic origin by the American Heart Association with multiple genes identified. Clinical manifestations include arrhythmias leading to sudden cardiac death, left ventricular systolic/diastolic dysfunction, and systemic thromboemboli. Echocardiography remains the most widely utilized and efficient means of diagnosing LVNC although no consensus on echocardiographic criteria exist currently.

Case Report: We present a case of a 27 year-old asymptomatic female with a known diagnosis of LVNC, admitted for observation at 32 weeks gestation for preeclampsia. Though maintaining NYHA class I functional status, TTE findings demonstrated an EF of 15-30%, RVSP of 97 mmHg, severe thickening of the myocardium with multiple deep recesses, severe global LV hypokinesis, septal and anterior wall akinesis, severe tricuspid regurgitation, and new RV systolic dysfunction. Given the limited information in the literature regarding this condition during pregnancy, a multidisciplinary consultation conference was convened with maternal-fetal medicine, cardiology, and anesthesia to discuss plans for timing and mode of delivery, as well as monitoring during delivery. At 33 weeks gestation, due to worsening preeclampsia, we proceeded with cesarean delivery under general anesthesia in the main operating room suite. Members of both the obstetric and cardiac anesthesia teams were present to manage potential worsening of ventricular failure or pulmonary hypertension during delivery. Neuraxial anesthesia was not utilized to prevent a decrease in systemic vascular resistance. Awake arterial line placement was followed by rapid sequence induction with etomidate, fentanyl, and succinylcholine, after which intubation and ultrasound guided right internal jugular vein catheterization commenced. Intraoperative TEE demonstrated severe LVNC and global akinesis. The patient successfully tolerated anesthesia and surgery without complication after which she was monitored in the intensive care unit. The infant displayed normal cardiac anatomy via prior fetal echocardiogram and was successfully delivered without complication (Apgar 7, 7).

Discussion: We describe a multidisciplinary approach to management of an asymptomatic parturient with LVNC Resulting in pulmonary hypertension and severe left ventricular dysfunction who required cesarean delivery due to worsening preeclampsia at 33 weeks gestation. Intraoperative use of TEE was useful for guiding the patient’s anesthetic management.