Labor Analgesia for a Patient with Primary Restrictive Cardiomyopathy and Paroxysmal Ventricular Tachycardia

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Case Report: A 17 year-old pregnant female at 26 weeks gestation with idiopathic restrictive cardiomyopathy was placed under surveillance until delivery at the request of the high-risk obstetrics team. Prior to admission, she had been recently removed from the heart transplant list due to increasing pulmonary hypertension and paroxysmal ventricular tachycardia and managed by the life vest external defibrillator.

At 33 weeks, rupture of membranes precipitated labor that was unresponsive to tocolysis. The anesthesia team was called to the operating suite for a STAT Cesarean section when persistent late decelerations occurred. However, fetal heart tones returned to baseline and the obstetrician we decided to continue with the vaginal delivery trial. Defibrillator pads were applied and an arterial line was placed, then epidural catheter was used for analgesia. Vasodilators, inotropes, and antarrhythmic agents were prepared but the patient did not experience any cardiovascular events in the immediate perioperative period. She remained in the OR and was able to successfully deliver a viable male after several hours and did not experience any immediate peripartum complications.

However, at four days post-partum she suffered a thrombotic CVA and underwent treatment with tPA. Evaluation with TEE revealed no intra-cardiac thrombus. In spite of aggressive therapy, the patient remained hemiplegic at the time of discharge. An ICD was placed 18 days after delivery and she was advised against future pregnancies.

Discussion: Idiopathic restrictive cardiomyopathy accounts for 2.5-5% of cardiomyopathies in pediatric patients, the majority of which will die within five years of diagnosis without heart transplant. Restrictive cardiomyopathy carries a grave prognosis with no consistent favorable pharmacologic response leaving transplant as the sole effective treatment option. Cardiac transplant itself carries a high mortality rate and even with a successful transplant patients may be limited in their functionality. Some studies report death in as high as 20% of pediatric patients within one year of heart transplant.

There is little data available concerning patients with restrictive cardiomyopathy and pregnancy, but the increased cardiac workload further increases the acuity of the management needed for vaginal or operative delivery. Detachment of the placenta after delivery could potentially overwhelm an already stressed heart when returning blood to the systemic circulation, precipitating frank pulmonary edema. Bilateral atrial dilation combined with markedly decreased compliance of the left ventricle could lead to heart failure or ventricular arrhythmia after auto transfusion from placental circulation. Due to these concerns, a pediatric cardiologist was involved with the case preoperatively and was physically present in the operating room for the duration of the patient's labor and post-delivery recovery in the ICU.