Peripartum Management of a Parturient with Repaired Tetralogy of Fallot and Factor V Leiden and MTHFR Mutation

Abstract Type: Case Report/Case Series
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Introduction: Factor V Leiden mutations are a leading cause of venous thrombosis in both pregnant and nonpregnant women and anticoagulation with heparin is the most common treatment. We present a case where close management of anticoagulation regimen allowed for early labor analgesia in a parturient with repaired Tetralogy of Fallot (ToF) and hypercoagulable state undergoing vaginal delivery.

Case Report: A 35-year-old G3P0 parturient presented at 38 weeks of gestation for elective induction of labor. She had a past medical history of ToF that was successfully repaired at the age of 4. Her last echocardiogram, performed at 38 weeks of gestation showed a severely enlarged right ventricle, right ventricular volume overload and severe pulmonary regurgitation. Her history was also significant for a hypercoagulable state secondary to heterozygous mutations of coagulation factor V Leiden and Methylene-tetrahydrofolate reductase (MTHFR). Her medication regimen included Aspirin 81 mg daily and low molecular weight heparin (LMWH) until 36 weeks of gestation at which time she was switched to unfractionated heparin (UH) 10,000 units twice daily. The last dose of UH was administered 24 hours before admission. A prenatal ultrasound showed ToF in her fetus. On admission the patient was asymptomatic, and no invasive monitoring was deemed necessary. Coagulation studies did not reveal any significant abnormalities. An epidural catheter was placed at L4-L5 level using a midline approach. Adequate pain control was achieved and maintained using a solution containing 0.125% Bupivacaine and Fentanyl 2 mcg/ml. Patient was hemodynamically stable during labor and she underwent an uneventful forceps delivery. The baby was immediately intubated and transferred to neonatal ICU for monitoring.

Discussion: Parturients with repaired ToF are at increased risk of developing atrial or ventricular tachyarrhythmias. Early administration of effective labor analgesia is necessary in order to minimize the cardiac stress associated with pain. Providing neuraxial analgesia is dependent on a normal coagulation profile. The hypercoagulable state associated with pregnancy combined with Factor V and MTHFR mutation (associated with elevated homocysteine levels) predisposes to thromboembolic events. The presented mutations are associated with an increased risk of pregnancy loss. Our patient had a history of miscarriages that prompted the genetic testing that led to her diagnosis. Anticoagulation with heparin is the most widely used treatment to avoid repeat pregnancy loss in patients with an underlying thrombophilic predisposition. Pregnancy complication related to thrombotic events and unpredicted timing of labor can cause unique challenges in regard to using regional anesthesia in anticoagulated parturients. Coordination of care and close management of the anticoagulation regimen allowed for early intervention and successful labor analgesia in our patient.