

Perivable HELLP Syndrome in a Patient with a Fontan Heart

Aaron M. Dom, MD

Carolinas Medical Center, Department of Obstetrics & Gynecology

Rebecca Pollack, MD

Carolinas Medical Center, Department of Obstetrics & Gynecology, Maternal Fetal Medicine

Background/Synopsis

There is a growing number of adult women with congenital heart disease. Pregnancy can pose unique, life-threatening risks to these patients.

Objective

To highlight the maternal risks associated with some adult congenital heart diseases during pregnancy, as well as the association of elevated AFP with hypertensive disorders of pregnancy.

Methods

Our patient is a 24 year old G2P0100 who presented at 23w5d by LMP consistent with 9 week ultrasound with worsening dyspnea and orthopnea. Her prenatal course was remarkable for a complex maternal cardiac anomaly: dextrocardia, double inlet left ventricle (single ventricle), pulmonary stenosis, transposition of the great arteries, ventricular septal defect, and NYHA class III congestive heart failure, status post bilateral Glenn anastomosis and Fontan procedure. She also had chronic hypertension, abdominal situs inversus, chronic hepatitis C, and a history of HSV. Prior obstetric history included a hysterotomy secondary to intrauterine fetal demise in the setting of HELLP syndrome and complete placenta previa at 23w0d. She was noted to have placentomegaly and an elevated AFP of 6.6 MoM. Her platelets were noted to be 61k, and her AST/ALT were over 2x the upper limit of normal.

Results

Due to suspicion for HELLP syndrome, she was given antenatal corticosteroids. She underwent classical cesarean section under cardiac anesthesia at 24w0d. Her neonate weighed 450g and had APGARs of 2, 2, and 5 at 1, 5, and 10 minutes, respectively.

Conclusion

It is critical to have multidisciplinary care teams, including cardiology and anesthesia, for adult patients with congenital heart disease in pregnancy. It is also important to have suspicion for the development of hypertensive disorders of pregnancy in the setting of an elevated AFP.