The Successful Approach to Perioperative Management of a Parturient with Unrepaired Truncus Arteriosus and Cyanotic Congenital Heart Disease for Cesarean section

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Introduction: Major advances in the treatment of women with congenital heart disease (CHD) have enabled them to survive to childbearing age without surgical correction and this number is rising by 5% per year. However, the hemodynamic demands of pregnancy are associated with a 50% maternal mortality (with pulmonary hypertension) and increased fetal mortality (1).

Case Presentation: Despite heart lung transplant refusal, numerous miscarriages and preconception counseling, a 30 year old G4P0030 at 25 weeks with cyanotic CHD presented to her cardiologist with worsening dyspnea. She had a known history of unrepaired Type II Truncus Arteriosus, type B Interrupted Arch, ventricular septal defect and Eisenmenger’s syndrome.

She was admitted, loaded with intravenous Treprostinil, and her medications optimized to include ASA, Enoxaparin, and Sildenafil. Maternal fetal medicine monitoring showed fetal growth retardation. A decision was made to continue the pregnancy as long as her functional status was not deteriorating and the fetus was growing. Meanwhile, a team of anesthesiologists, obstetricians, cardiologist, cardiac surgeon, neonatologist and intensivists was formed for perioperative planning for the cesarean section. Specifics were delineated: the utility of left sided arterial line (mean arterial pressure) versus right (cerebral perfusion pressure), TEE, and CVP. Options for uterine tone post delivery were explored.

Preop evaluation included platelets, TTE, cardiac MRI and 6 minute walk test. At 29 weeks, fetal umbilical artery Doppler revealed reversal of end diastolic flow confirming worsening growth restriction requiring delivery.

Preop vitals included an oxygen saturation of 85% on room air, blood pressure 100/60, heart rate 118, height 66 inches, and weight 84 kg. Arterial and central line were placed preop with platelet infusion. Goal hemodynamics were to maintain preload, a sinus tachycardia and saturations of 85%. Induction was performed with Fentanyl, Rocuronium and Etomidate. Anesthesia was maintained with Remifentanil infusion, Midazolam and Desflurane. The neonate was delivered 7 minutes later and only needed nasal cannula. The patient was extubated and taken to recovery.

Hours later, she required a hysterectomy due to bleeding. Edema from resuscitation prevented abdominal closure and extubation. After 2 weeks of meticulous intensive care, her abdomen was closed and she was extubated.

Discussion: There are now 1 million patients with CHD that survive to adulthood. The most important predictor is functional status and problems relate to arrhythmias and heart failure. The concern remains that labor can lead to a deterioration in cardiac status, and a cesarean delivery under general is preferred (2). A multidisciplinary approach involving preconception risk counseling as well as careful perioperative planning is necessary to improve both maternal and fetal outcomes (3). Optimal care for parturients with unrepaired CHD, despite their small number and increased expense, remains at specialized centers that offer centralization, consistency and expertise in complex physiology management.

References:
1) NEJM 2000;342(4):256-63;
2) J Am Coll Cardiol 2007;49:2303â11;