



Preoperative Management of Twin Gestation Pregnancy in a Parturient with Eisenmenger Syndrome

Kyle M. Struzyk MD¹, Camron Wipfli MD¹, Molly Houser MD FACOG²

Department of Anesthesiology¹, Department of Obstetrics & Gynecology²; The University of Tennessee Health Science Center; Memphis, TN



Introduction

Women with Eisenmenger syndrome who elect to attempt pregnancy and delivery of children are at significant risk for peripartum morbidity and mortality to themselves and their progeny. Historically, patients with Eisenmenger syndrome are strongly advised to abstain from pregnancy or terminate pregnancy within the first trimester. Of primary concern, is the right-to-left shunt fraction that is progressively worsened throughout the pregnancy. Furthermore, the autotransfusion period immediately post-partum in conjunction with underlying pulmonary hypertension rapidly exacerbates this shunt fraction, leading to precipitous hypoxia. This recommendation is even more paramount when considering multiple gestation. However, with modern medical management, these congenital heart patients are surviving to later ages and requiring obstetric care.

We present a woman with severe pulmonary hypertension due to Eisenmenger Syndrome associated with a congenital unprepared ASD, who successfully underwent cesarian delivery of her di/di twin pregnancy.

Materials & Methods

Cardiac Surgery was consulted on behalf of cardiac anesthesia and Maternal Fetal Medicine for prepartum ECMO introducers. Perfusion services remained on standby with bypass on standby.

Patient consent was obtained, and no identifiable patient information was used.

Case Presentation

Our patient is a 29F, G2P1011, with PMH remarkable for severe pulmonary hypertension (unrepaired ASD c/b Eisenmenger Syndrome) who presented with 30w5d di/di twin IUP. Patient presented from MFM clinic visit and found to be hypoxic, with SpO₂ at 87% and she was promptly admitted to the cardiac ICU for preoperative planning. An ECHO was obtained revealing a severely dilated RA, RV with severe hypertrophy, moderate TR and a preserved LVEF at 60%. A PA line was placed revealing admission PA pressures of 102/40 mmHg, consistent with known severe pulmonary hypertension.

Management

The patient was counseled regarding perioperative management. Bilateral 16g IVs and radial arterial line was obtained the night prior to scheduled C-section. The patient received supplemental oxygen to goal >94% for fetal oxygenation and was continued on her home Treprostinil. NO was initiated with minimal change to her pulmonary pressures.

On the day of surgery, the patient received an epidural with test dose confirmation. Intermittent doses of 2% lidocaine, were used to achieve an adequate surgical level. The patient was taken to the operating room where bilateral access was obtained via small 5fr introducers as a precaution for potential ECMO.

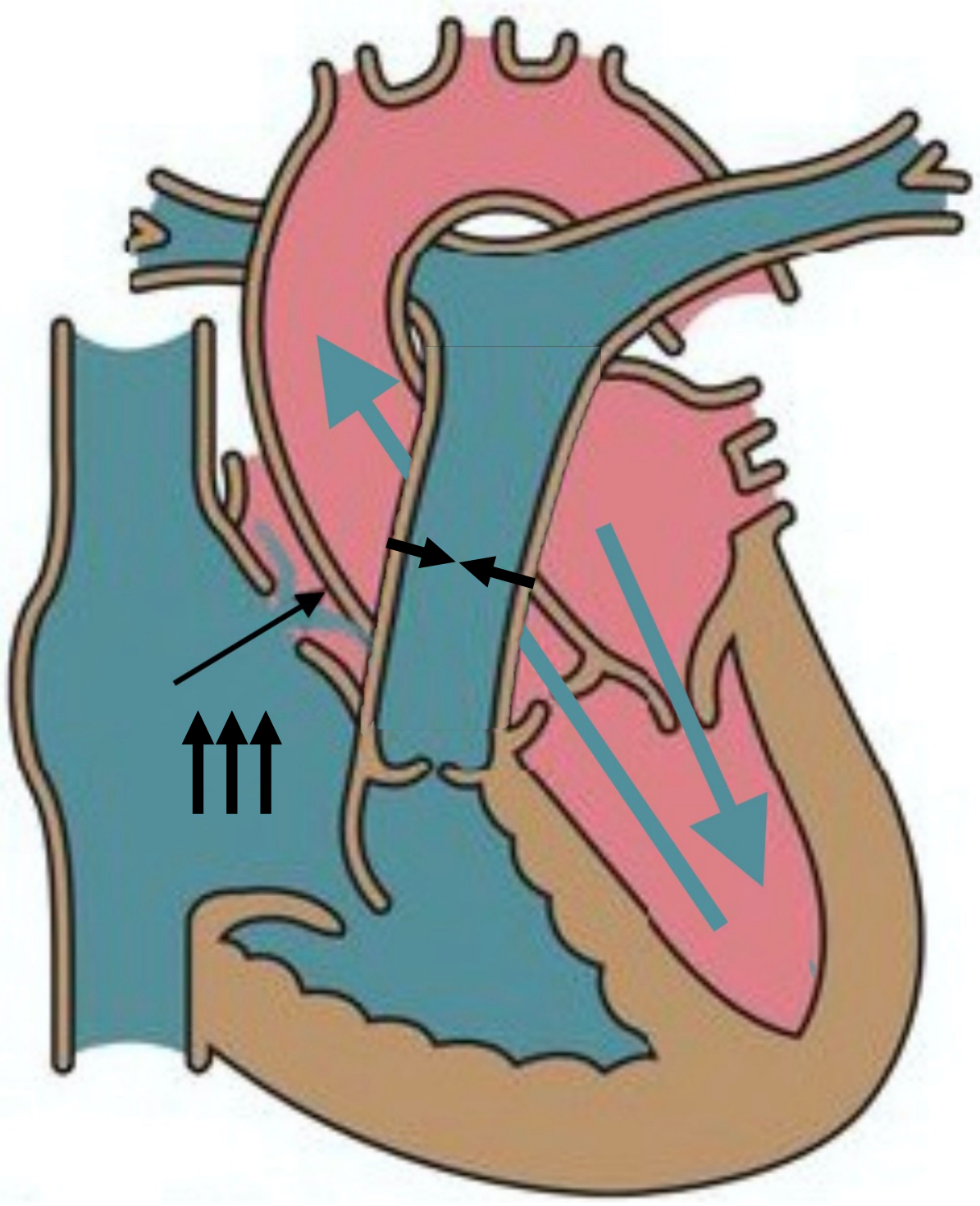
C-Section was performed, with unremarkable delivery of both baby A and baby B within 8 minutes of skin incision. After delivery, 30 units of oxytocin was diluted into 500ml LR and an infusion was promptly started. The patient was started on a vasopressin drip to support the hypotension experienced from oxytocin. Intermittent boluses of vasopressin were also administered.

Management Cont.

After delivery, the patient's systolic pulmonary artery pressure began to exceed her systolic blood pressure increasing her right-to-left shunt with subsequent worsening hypoxemia. The patient's SpO₂ dropped into the mid 80s and supplemental oxygenation was increased to settle in the low 90s. Supra-systolic PA pressures continued for approximately 20 minutes post-delivery. Face mask oxygen was continually titrated during this time in conjunction with coaching the patient to hyperventilate as tolerated.

Eventually her supra-systolic PA pressures subsided and her hypoxemia improved. Cesarean closure was performed without complication and the ECMO introducers were removed. The patient recovered without further complication.

Shunt Physiology



Discussion

Patients with Eisenmenger's syndrome present unique and challenging management strategies for anesthesia providers. This is especially the case in the parturient patient, where physiologic changes in both pregnancy and delivery can be fatal to the mother and the baby. Management of these patients requires a coordinated and well-planned approach in order to deliver the best care. In this case, it was the coordinated planning and efforts from maternal fetal medicine, pediatric critical care, cardiac surgery, and cardiac and obstetric anesthesiology that were able to come together to best optimize and safely deliver care to this mother and her children.

References

Brida M, Gatzoulis MA. Pulmonary arterial hypertension in adult congenital heart disease. *Heart* 2018;**104**:1568-1574.

Dachlan EG, Amirah, Cininta N, et al. High Maternal Neonatal Mortality and Morbidity in Pregnancy with Eisenmenger Syndrome. *J Pregnancy*. 2021;2021:3248850. Published 2021 Sep 27. doi:10.1155/2021/3248850

Liu Y, Li Y, Zhang J, et al. Pregnancy outcomes of women with Eisenmenger syndrome: A single-center study [published online ahead of print, 2022 Dec 13]. *Int J Cardiol*. 2022;S0167-5273(22)01888-5. doi:10.1016/j.ijcard.2022.12.014

Lopez BM, Malhamé I, Davies LK, Gonzalez Velez JM, Marelli A, Rabai F. Eisenmenger Syndrome in Pregnancy: A Management Conundrum. *J Cardiothorac Vasc Anesth*. 2020;34(10):2813-2822. doi:10.1053/j.jvca.2020.02.053

Naguib MA, Dob DP, Gatzoulis MA. A functional understanding of moderate to complex congenital heart disease and the impact of pregnancy. Part II: tetralogy of Fallot, Eisenmenger's syndrome and the Fontan operation. *Int J Obstet Anesth*. 2010;19(3):306-312.

Neema PK. Eisenmenger syndrome: An unsolved malady. *Ann Card Anaesth* 2012;15:257-8.

Rathod S, Samal SK. Successful pregnancy outcome in a case of eisenmenger syndrome: a rare case report. *J Clin Diagn Res*. 2014;8(10):OD08-OD9. doi:10.7860/JCDR/2014/9120.5050

Yadav V, Sharma JB, Kachhawa G, Kriplani A, Mishra S, Kriplani I. Successful pregnancy outcome in Eisenmenger's syndrome during pregnancy. *Eur J Obstet Gynecol Reprod Biol*. 2018;223:146-147. doi:10.1016/j.ejogrb.2018.02.016

Yuan SM. Eisenmenger Syndrome in Pregnancy. *Braz J Cardiovasc Surg*. 2016;31(4):325-329. doi:10.5935/1678-9741.20160062

