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A Successful Case of PAH Management Through Pregnancy and Delivery

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Background: Patients with pulmonary arterial hypertension (PAH) of child-bearing potential are counseled to avoid pregnancy due to increased fetal and mortality risk associated with physiologic and hemodynamic changes. We present a PAH patient whose pregnancy was managed by a multidisciplinary team with a favorable outcome.

Case Presentation: The patient is a 41-year-old G1P1 woman with history of anemia, epistaxis, pulmonary arteriovenous malformations, and inflammatory bowel disease who was found to have echocardiographic evidence of pulmonary hypertension during a hospitalization for pneumonia. At approximately 12 weeks gestation she was urgently referred to our center.

Management: Right heart catheterization confirmed the diagnosis of PAH WHO Group 1 (Table 1). She was promptly seen by several specialty clinics at UCSF. The Pregnancy and Cardiac Treatment (PACT) Clinic diagnosed hereditary hemorrhagic telangectasia (HHT). Both the PACT Clinic and Maternal-Fetal Medicine (MFM) / High Risk obstetrics programs counseled the patient regarding high risk of pregnancy in the setting of her pulmonary hypertension and HHT, with risk of mortality estimated in the range of 10-25%. She elected to continue with the pregnancy. She was referred to our Pulmonary Hypertension Clinic, and thereafter hospitalized for initiation of intravenous epoprostenol to optimize hemodynamics and minimize peripartum risk. She tolerated initiation of IV epoprostenol with only mild side effects, and was discharged at a dose of 8.5 ng/kg/min, with plans to continue up-titration.

A multi-disciplinary team convened to plan for delivery, including the Cardiology (PACT), Pulmonary Hypertension, MFM, Cardiac Anesthesia, and ICU services. She was scheduled for a repeat low transverse cesarean section in a cardiac operating room, with cardiac anesthesia and extra-corporeal membrane oxygenation (ECMO) on standby. The plan included transferring the infant immediately to UCSF Children's Hospital. She went into labor 4 days prior to her scheduled delivery and the team was urgently mobilized. Throughout her peripartum course, the epoprostenol dose was 24 ng/kg/min. Post-partum, the patient was transferred to the cardiac intensive care unit for invasive hemodynamic monitoring. Her immediate post-partum course was uneventful and she was discharged home 4 days later on the same epoprostenol dose.

Follow-up: She had an IUD placed at her MFM PACT postpartum visit. Follow-up echocardiogram at 6 weeks post-delivery showed preserved right ventricular function with tricuspid regurgitation and pulmonary hypertension. RHC at 8 weeks post-delivery showed severe pulmonary hypertension with preserved cardiac index and normal right atrial pressure (Table 1). The patient strongly preferred switching to oral therapy, and epoprostenol was transitioned to selexipag (max dose reached 1200 mcg BID). At post-discharge follow-up in PH Clinic she reported improved energy with stable WHO Functional Class II symptoms.

Discussion: Despite advances in available therapies, maternal risk remains an important consideration in pregnancy in PAH and patients should be counseled accordingly. For those electing to continue with pregnancy, early initiation of IV therapy such as epoprostenol may be utilized to optimize hemodynamics. Referral to an accredited Pulmonary Hypertension Care Center (PHCC) is advisable for PAH patients who are pregnant. Comprehensive planning for delivery and coordination for appropriate multidisciplinary team presence at delivery is recommended.

PH PN SYMPOSIUM

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Table 1: Right Heart Catheterization Data

Date	***RA***	***PAS***	***PAD***	***PAM***	***PCWP***	***CO/CI***	***PVR***
3/12/18 *(OSH)*	7	74	22	39	23	4.63/3.02	3.5
3/12/18 + NO		67	30	42			
8/13/18 *(19-20 weeks gestation)*	3	70	31	44	4	5.8/3.5 (TD)	6.6
2/13/19 *(2 months post-partum on epoprostenol)*	7	75	27	45	6	5.1/3.1 (TD)	8