Pregnancy outcomes in patients with pulmonary arterial hypertension associated with congenital heart disease.
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Take Home Points:
- Maternal cardiovascular morbidity is high in women with Congenital Heart Disease Associated Pulmonary Hypertension. [heart failure and severe cyanosis]
- Mortality risk in this heterogeneous contemporary cohort of pregnant women with PAH and CHD appears to be low

Commentary from Dr. Tabitha Moe (Phoenix), section editor of ACHD Journal Watch:
The authors aim to address an important question in this retrospective outcomes study, i.e. of whether improvements in general medical and cardiological care has translated into improved outcomes for women with PAH and congenital heart disease undergoing pregnancy. This relatively small cohort included 28 pregnancies in 20 patients, data being collected by questionnaire from 22 French CHD referral centers between 1997-2015. Study subjects carried the following diagnoses, including: VSD [n=5], Pulmonary Atresia with VSD +/- Aortopulmonary collaterals [n=3], ASD [n=3] and the remaining 9 had any of the following diagnoses: DORV, single ventricle, VSD, Truncus arteriosus, and CCTGA + VSD. Seventeen were considered to have Eisenmenger’s syndrome, 3 having segmental PAH. Only 3 patients were treated with PAH therapies during pregnancy [sildenafil, Tadalafil and oxygen therapy]. 85% of the cohort were in NYHA functional class I or II. The most common cardiovascular adverse event were: worsening of hypoxemia, and heart failure requiring inotrope support occurred in 4 pregnancies, one subject required ECMO support. There was a single maternal mortality in the oldest member of the cohort (42), who died 10 days post-partum. She also had a history of atrial arrhythmia, and moderately impaired RV systolic function. The diagnosis was not provided. Six patients received antiplatelet or anticoagulant therapy, and there were no cases of hemoptysis. The most common neonatal complications were prematurity in 77.78% percent of births, and almost 40% of fetuses had growth restriction.
The most common obstetric complication was bleeding: 4 with post-partum hemorrhage and 2 with abruptio-placentae. In at least half of these cases of bleeding, the subjects were anticoagulated. There were only 18 completed pregnancies, and 12 (67%) were delivered by caesarean section.

Caution should be exercised in the generalization of these results, as the study subjects were a very heterogeneous group in terms of diagnoses and probably also heterogeneous in the degree of pulmonary hypertension. Indeed only 3 had required advanced therapies, suggesting a cohort in a better state of health. Obstetric bleeding was very common and seemed to be associated with formal anticoagulation.