Maternal and fetal outcomes of pregnancy with Fontan circulation: a multicentric study of 59 pregnancies

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Objectives: Despite serious long-term sequelae, more and more women with Fontan palliation reach childbearing age. However there is paucity of data on the mid to long-term pregnancy outcomes and management of this condition. We sought to determine maternal and fetal outcomes of pregnancy in women with Fontan palliation.

Methods: This multicentric, retrospective study included women with Fontan circulation followed in 13 French centers from January 2000 to June 2014. All pregnancies were reviewed, including miscarriages, abortions, premature and term births. We reviewed maternal and fetal outcomes.

Results: Thirty-seven patients had 59 pregnancies. Mean age was 27±5 years at first pregnancy. There were 16 miscarriages (27%) and 36 live births with 1 twin pregnancy. Cardiac events occurred in 10 (17%) pregnancies, with no maternal death. The most common cardiac complication was atrial arrhythmia, which occurred in 3 women. Prior atrial arrhythmia was a significant predictor of atrial arrhythmia during pregnancy (p=0.03). Hematological complications including thromboembolic/hemorrhagic events occurred in 5 women antepartum, and 4 women postpartum. There was a high incidence of prematurity (n=25/36, 69%), with a mean gestational age of 34 weeks and a mean birth weight of 1980 g (10th percentile). Anticoagulation therapy, regardless of medication or dose, was significantly associated with neonatal events (p<0.01). In contrast there were more live births in women receiving low molecular weight heparin. After a median follow-up of 24 months, there was no significant worsening of clinical status/cardiac function/worsening thromboembolic disease noted.

Conclusions: Women can successfully complete pregnancy with Fontan circulation. Although there is an increase in cardiac and hematological events during pregnancy, there is no long-term impairment due to the pregnancy of the single ventricle function or the Fontan circulation, and there is no maternal death. At least prophylactic anticoagulation is recommended during pregnancy and postpartum period to prevent thromboembolic complications.