

Congenital Heart Disease in Pregnancy

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KEYWORDS

- Pregnancy • Congenital heart disease • CARPREG score • Contraindications
- Infective endocarditis • Eisenmenger syndrome • Pulmonary hypertension • Tetralogy of Fallot

KEY POINTS

- For cardiac patients, prepregnancy counseling before conception is strongly recommended.
- It is important for the clinician to understand the normal hemodynamic changes during pregnancy.
- Pregnant patients can be risk-stratified into low, medium, and high cardiac risk based on the CARPREG Risk Score.
- Cardiac absolute contraindications against pregnancy include: pulmonary hypertension, Marfan syndrome with dilated aortic root (4 cm), severe left heart obstruction, and systemic ventricular function less than 30%.
- Medications to avoid or use with caution in pregnancy include: angiotensin converting enzyme inhibitors, angiotensin-II receptor blockers, amiodarone, warfarin, spironolactone.
- Pregnant patients with congenital heart disease can be managed successfully with collaboration between the maternal–fetal medicine specialist and an adult congenital heart disease cardiologist.

INTRODUCTION

Each patient with known congenital heart disease (CHD) should ideally be seen before pregnancy by a cardiologist who is experienced in adult CHD and a maternal–fetal medicine specialist for a preconception consult.^{1,2} The discussion should include the effect of pregnancy on the mother and the impact of the mother's heart disease on the fetus. However, in many circumstances, this discussion does not occur. Drenthen showed that while 69% of Fontan patients discussed pregnancy with their cardiologist, and 49% had pregnancy discouraged, 77% were still considering pregnancy.³

MATERNAL RISK

Risk stratification of the mother based upon her cardiac diagnosis can be helpful in the

preconception evaluation. In general, regurgitant lesions are well-tolerated, while stenotic lesions convey higher risk. The risk varies from very low (eg, similar to the general population), as in mild mitral regurgitation, to very high, as in severe coarctation. Siu and colleagues,⁴ as part of the Cardiac disease in Pregnancy (CARPREG) investigators found that, in their cohort of 599 women with heart disease, there was a 13% incidence of a cardiac event, notably pulmonary edema, arrhythmia, stroke, or cardiac death. The authors developed a score to predict adverse maternal outcomes (**Box 1**).

Functional capacity assessment by history and physical examination, and hemodynamic status with an echocardiogram and electrocardiogram are recommended as part of the prepregnancy clinic visit. Recent data also suggest that cardio-pulmonary exercise testing should be performed,

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Box 1**CARPREG Risk Score: Predictors of maternal cardiovascular events**

New York Heart Association (NYHA) functional class >II

Cyanosis (room air saturation <90%)

Prior cardiovascular event

Systemic ventricular ejection fraction <40%

Left heart obstruction (eg, mitral valve area <2 cm² or aortic valve area <1.5 cm², or left ventricular [LV] outflow gradient of >30 mm Hg)

CARPREG risk score: for each CARPREG predictor that is present, a point is assigned. Risk estimation of cardiovascular maternal complications.

0 points-5%

1 point-27%

>2 points-75%

Data from Siu SC, Sermer M, Colman JM, et al, Cardiac Disease in Pregnancy (CARPREG) Investigators. Prospective multicenter study of pregnancy outcomes in women with heart disease. *Circulation* 2001;104(5): 515–21.

since an abnormal chronotropic response is predictive of maternal and neonatal events.⁵ If indicated, cardiac catheterization or cardiac magnetic resonance imaging should be performed in moderate to high-risk pregnancy patients. In patients who are found to be very high risk, other options should be recommended, such as adoption, surrogacy, and appropriate contraception or sterilization.

Contraindications to Pregnancy

In conditions where the maternal risk of death exceeds 10%, pregnancy should be routinely avoided. Due to the prohibitively high risk to the mother and/or fetus, pregnancy should be avoided in the conditions listed in **Box 2**.

FETAL RISK

Maternal heart disease is associated with fetal and neonatal complications, such as intrauterine growth restriction, fetal loss, and prematurity. These complications are more common in cases of maternal cyanosis, anticoagulation, poor NYHA functional class, left heart obstructive lesions, maternal smoking, or multifetal gestation.⁸ In women who have CHD, the risk of CHD in the offspring is 7% to 8%, for conditions with no chromosomal abnormality or family history.³ When the father

Box 2**Contraindications to pregnancy**

- Pulmonary arterial hypertension of any cause
- Marfan syndrome with dilated aortic root >40 mm
- Aortic dilatation >50 mm in aortic disease associated with bicuspid aortic valve
- Severe left heart obstructive lesions (severe mitral stenosis, severe symptomatic aortic stenosis (AS), or native severe coarctation)
- Severe systemic ventricular dysfunction (LV ejection fraction [LVEF] <30%, NYHA III-IV)
- Previous peripartum cardiomyopathy with any residual impairment of LV function

Data from Refs.^{4,6,7}

has CHD, the risk of CHD in the fetus is roughly 3% to 4%.⁶ Nonetheless, CHD in the fetus approaches 50% in single gene disorders such as Marfan syndrome. Genetic counseling is recommended if there is a dysmorphism or a chromosomal abnormality suggested.⁹

PRECONCEPTION MEDICAL MANAGEMENT

Medical or surgical intervention may be necessary before conception in order to optimize cardiac function and to minimize the risks of pregnancy. This should include a review of all cardiac medications, with cessation of any contraindicated drugs and substitution of suitable alternatives (**Box 3**). Comorbidities should also be well-controlled, including hypertension, diabetes, and obesity. Surgery or percutaneous intervention (eg, balloon dilation or transcatheter septal defect closure) should be strongly considered for significant valve disease, and any arrhythmias should be well-controlled.

Box 3**Cardiac drugs to avoid/use with caution**

Angiotensin converting enzyme (ACE) inhibitors
 Angiotensin-II receptor blockers (ARBs)
 Amiodarone
 Warfarin^a
 Spironolactone

^a Use during first trimester is contraindicated. Warfarin may be used from weeks 13–35, and then heparin may be used until delivery.¹⁰

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