Pregnancy and Adult Congenital Heart Disease

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KEYWORDS

• Adult congenital heart disease • Pregnancy • Heart failure • Obstetrics • Obstetric anesthesia • Congenital heart disease • Arrhythmia • Preconception counseling

KEY POINTS

- Best practice in the care of the pregnant patient with congenital heart disease (CHD) includes multidisciplinary cardiology/obstetric/anesthesiology preconception evaluation and planning.
- Heart failure and arrhythmias remain the primary complications in pregnant individuals with heart disease.
- Recommendations for pregnancy and delivery should be individualized according to congenital anatomy, current physiology, and the effects of superimposed pregnancy-related physiology.
- Many CHD patients are lost to follow-up in adulthood, and pregnancy is an opportunity to re-establish longitudinal adult congenital cardiac care.

INTRODUCTION

The population of adults with congenital heart disease (ACHD) continues to grow at an impressive rate because these individuals are now surviving beyond childhood as a result of medical and surgical advances. These medical and surgical advances result in an expanding number of women of childbearing age with congenital heart disease (CHD) with a significant increase in number of deliveries among women with CHD in recent years.[1] Importantly, with the high loss to follow-up rates in the ACHD population, many of these individuals only return to the health care system when contemplating pregnancy or already pregnant. This time is an opportunity to aid these women in resuming lifelong cardiac care, recognizing that, ideally, preconception counseling is paramount. In this review, general issues applicable to the ACHD population are addressed, including preconception counseling, risk assessment, lesion-based review of pregnancy physiology, and clinical management of potential complications for the general cardiologist (**Table 1**). Pregnancy in the ACHD population requires a multidisciplinary team, and therefore, the primary cardiologist, local (or remote but consulting) ACHD center, high-risk obstetrics or maternal fetal medicine, and obstetric anesthesia should all be involved in pregnancy planning, including patient selection and intrapartum and delivery planning. The entire care team, including the nurses who will spend the most time at the bedside during labor and delivery, should be included in the planning and education process. Individuals with ACHD can increasingly have successful pregnancies when planned with their team and with close communication throughout the process.

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Table 1
Adult congenital heart disease disease-specific risks and recommendations for pregnancy management

	Evaluate/Exclude	Potential Risks	Recommendations
VSD	PAH Ventricular dysfunction	Arrhythmia Endocarditis	Antibiotics if unrepaired
ASD (unrepaired)	PAH Ventricular dysfunction	Arrhythmia Paradoxic embolism	Low-dose aspirin thromboembolic prophylaxis (if bed rest)
Coarctation (repaired)	Re-coarctation Aortic/brain aneurysm	Pre-eclampsia/HTN Aortic dissection Left heart failure	β-Blocker if HTN Avoid placental hypotension Consider C-section if aneurysm
TOF (repaired)	Severe RVOT obstruction Severe PR RV dysfunction DiGeorge syndrome	Arrhythmia/VT Right heart failure Endocarditis	Volume control Peridelivery Consider preterm delivery if RHF Antibiotic prophylaxis
Mitral stenosis	Severe MS Pulmonary venous HTN	Atrial fibrillation Thromboembolic event Pulmonary edema	β–Blockers Diuresis 3rd-trimester bed rest Antibiotic prophylaxis
AS	Severe AS (peak >80 mm Hg) Symptoms ST depressions LV dysfunction	Arrhythmia Angina Heart failure/shock Endocarditis	± 3rd-trimester bed rest Diuretics as needed If HF: valvotomy or preterm C-section Antibiotic prophylaxis
Systemic RV	Ventricular dysfunction Severe systemic TR Arrhythmia Symptoms Venous pathway obstruction	Heart failure Arrhythmia Thromboembolic event	Restore NSR if needed Aspirin 81 mg daily ± Rhythm monitoring
Cyanotic lesions without PAH	Ventricular dysfunction Saturation <85%	Hemorrhage Thromboembolic events Increased cyanosis Heart failure Endocarditis	Bed rest and O_2 Thromboembolic prophylaxis
Fontan circulation	Ventricular dysfunction Arrhythmia Prior heart failure	Thromboembolic events Increased cyanosis Heart failure Endocarditis	Anticoagulation or aspirin Avoid dehydration during delivery Restore NSR quickly Avoid calcium channel blockers Antibiotic prophylaxis
Marfan syndrome	Aortic root >4.0	Type A dissection	β–Blocker C-section if root >4.0–4.5 cm
Eisenmenger syndrome	Ventricular dysfunction Arrhythmia	30%–50% risk of death	Discuss termination If late for termination: • O_2 • Bed rest • Thromboembolic prophylaxis • Prolonged postpartum stay • Permanent contraception

Abbreviations: HTN, hypertension; MS, mitral stenosis; NSR, normal sinus rhythm; PAH, pulmonary arterial hypertension; PR, pulmonic regurgitation; RHF, rheumatic heart failure; VT, ventricular tachycardia.