

*Clinicians and Facilities:***RESOURCES WHEN CARING FOR WOMEN WITH ADULT CONGENITAL HEART DISEASE OR OTHER FORMS OF CARDIOVASCULAR DISEASE***Abha Khandelwal, MD, MS – Stanford University School of Medicine**Julie Arafah, MSN, RN – Lucile Packard Children’s Hospital, Stanford University***INTRODUCTION**

This chapter includes a summary of guidelines published by the American College of Cardiology and the American Heart Association in conjunction with other professional groups that manage adult cardiovascular disease.<sup>1,2</sup> These guidelines are based on scientific evidence reviewed by experts in their field of practice. The purpose of the guidelines is to give clinicians the most current evidence upon which to base management of adults with specific cardiac disease. This synopsis is intended to provide information to clinicians who care for women with cardiac disease about current resources and management strategies. Key components of comprehensive, evidence-based care include resources consisting of diagnostic testing, imaging and experienced multidisciplinary staff. Recommendations for appropriate resources when providing care for adults with cardiac disease are also included.<sup>3</sup>

**OB PROVIDERS: ADULT CONGENITAL HEART DISEASE (ACHD) GUIDELINES.<sup>1,2</sup>**

- Estrogen-containing oral contraceptives are not recommended for patients with Adult Congenital Heart Disease (ACHD) at risk of thromboembolism such as those with cyanosis, intra-cardiac shunt, severe pulmonary arterial hypertension (PAH) or Fontan repair.
- Patients with ACHD should consult with an ACHD expert before pregnancy to develop a plan for management of labor and the postpartum period.
- Preconception counseling is recommended for women receiving chronic anticoagulation with warfarin.
- Patients with intra-cardiac right to left shunt should have fastidious care of IV lines to avoid air embolus.
- Fetal echocardiography is recommended between 18 and 20 weeks in women with personal history of congenital heart disease.

Table 1, on the next page, is an overview and does not replace evaluation and management by an ACHD physician, which should be pursued in all ACHD patients.<sup>1-3</sup>

**Table 1: Adult Congenital Cardiac Lesions: Management and Expected Outcomes in Pregnancy**

<b>Lesion</b>	<b>Overview *Note: This does not supplant evaluation and management by an ACHD physician, which should be performed in all ACHD patients.<sup>1-3</sup></b>
<b>Shunt lesions</b>	
Atrial Septal Defect (ASD)	<ul style="list-style-type: none"> <li>Well tolerated in the absence of pulmonary arterial hypertension (PAH).</li> <li>Repair should be considered in patients with large ASDs prior to pregnancy in the absence of PAH.</li> <li>Pregnancy is not recommended in patients with ASD and severe PAH or Eisenmenger syndrome due to excessive maternal and fetal mortality.</li> </ul>
Ventricular Septal Defect (VSD)	<ul style="list-style-type: none"> <li>Small VSDs without PAH and no associated lesions do not have increased CV risk and pregnancy is usually well tolerated.</li> <li>Prior to pregnancy, repair should be considered in patients with large VSDs in the absence of PAH.</li> <li>Pregnancy is not recommended in patients with VSD and severe PAH or Eisenmenger syndrome due to excessive maternal and fetal mortality.</li> </ul>
Atrioventricular Septal Defects (AVSD)	<ul style="list-style-type: none"> <li>Usually well-tolerated post repair in the absence of PAH.</li> <li>Pregnancy is not recommended in patients with AVSD (repaired or unrepaired) and severe PAH or Eisenmenger syndrome due to excessive maternal and fetal mortality.</li> </ul>
<b>Left-sided obstruction</b>	
Aortic Stenosis (AS)	<ul style="list-style-type: none"> <li>Mild or moderate stenosis is usually well tolerated in pregnancy.</li> <li>Vaginal delivery is preferred except in critical AS or if associated with aortic disease (dissection or aneurysm).</li> </ul>
Supravalvular or Subvalvular AS	<ul style="list-style-type: none"> <li>Those with significant obstruction, coronary involvement or aortic disease should be counseled against pregnancy.</li> </ul>
Coarctation of Aorta	<ul style="list-style-type: none"> <li>Patients with severe obstruction or aortic aneurysm should have hemodynamic assessment and treatment prior to getting pregnant.</li> </ul>
<b>Right-sided obstruction</b>	
Pulmonic Stenosis and Right Ventricular Outflow Tract Obstruction	<ul style="list-style-type: none"> <li>Mild to moderate obstruction is well tolerated.</li> <li>Severe obstruction should be treated prior to pregnancy.</li> </ul>
Tetralogy of Fallot (TOF)	<ul style="list-style-type: none"> <li>TOF should be repaired prior to pregnancy. In patients with repaired TOF and a competent pulmonary valve, pregnancy is well tolerated in those with good functional capacity and without residual lesions.</li> <li>Severe symptomatic pulmonary regurgitation should be treated prior to pregnancy in the presence of severe right ventricle (RV) dilatation. Patients should be screened for arrhythmias prior to pregnancy.</li> </ul>
<b>Other lesions</b>	
Single Ventricle Lesions Post Fontan Repair	<ul style="list-style-type: none"> <li>Successful pregnancy is reported after Fontan repair but arrhythmias, ventricular dysfunction, thrombotic complications and edema have been reported.</li> <li>Increased risk for spontaneous abortion or premature birth.</li> </ul>
Ebstein's Anomaly	<ul style="list-style-type: none"> <li>Generally well tolerated in the absence of severe tricuspid regurgitation, arrhythmias and cyanosis; however, there is an increased risk of low birth weight, and fetal loss if significant cyanosis is present. The risk of CHD in offspring is approximately 6%.</li> </ul>

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## Obstetric Providers:

### RESOURCES WHEN CARING FOR ADULTS WITH CONGENITAL HEART DISEASE

#### FOR ADOLESCENTS:

- Adolescents with congenital heart disease (CHD) should have a coordinated, collaborative and comprehensive healthcare transition to adult cardiac specialists with services similar to the level of care they received as children.

#### FOR ADULTS:

- Each provider of adult congenital heart disease (ACHD) care and each facility where ACHD patients receive care should be in contact with a regional ACHD center of excellence.
- Regional ACHD centers are responsible for the organization of ACHD healthcare including:
  - Staff with expertise in cardiology: physicians, nurses, advanced care providers, anesthesiologists
  - Diagnostic testing and imaging
  - Interdisciplinary care teams for special patient populations including obstetrics and neonatology
  - Mechanisms for consultations, referrals, review of policies and protocols, quality assessment
- Patients with ACHD should possess documents that describe their condition including how to access local healthcare and the regional center.
- Each patient with ACHD should have a primary health care provider who has her current medical records and a consultation arrangement with local and regional ACHD experts.
- Patients with moderate or complex ACHD should be followed by a provider with expertise in that level of ACHD, or their primary provider should be in frequent consultation with an expert in CHD. Plans for referral to a higher level of expert care should be in place in the event the patient's condition becomes unstable.
- Adults with moderate or complex CHD should have the following procedures or evaluations in the regional center of excellence:
  - Diagnostic and interventional procedures
  - Surgery that necessitates conscious sedation or general anesthesia
  - Sudden onset or emergent cardiac or non-cardiac conditions.

## Obstetric Providers:

### ADULT VALVULAR DISEASE GUIDELINES

#### PRECONCEPTION EVALUATION AND INTRAPARTUM MONITORING:

- Prior to pregnancy, all patients with known or suspected valve disease should be evaluated by a cardiologist with expertise in managing patients with valvular heart disease during pregnancy who can provide preconception counseling.
- Transthoracic echo (TTE) is recommended in the evaluation of all women with known or suspected valvular heart disease as part of preconception counseling and assessment.
- Exercise tolerance testing should be considered prior to pregnancy in patients with severe valve disease.
- Symptomatic severe valve disease should be treated prior to pregnancy and treatment might be indicated in selected asymptomatic patients.
- Asymptomatic valve disease should be monitored by a cardiologist and may require additional testing during pregnancy.
- Pregnant patients with severe valve stenosis or regurgitation should be monitored in a tertiary care center with a dedicated heart team consisting of cardiologists, anesthesiologists and obstetricians with expertise in the management of high-risk cardiac patients during pregnancy. A cardiothoracic surgeon should be part of the team in select cases.
- Use of angiotensin-converting enzyme (ACE) inhibitors and angiotensin receptor blockers is contraindicated in pregnant and breastfeeding patients (yet, due to increased risk of angioedema, caution and monitoring of ACE inhibitor use in pregnant African-American women is appropriate); however, the use of beta-blockers and diuretics may be reasonable for symptomatic relief.

#### RHEUMATIC FEVER / ENDOCARDITIS PROPHYLAXIS:

- Secondary prevention of rheumatic fever with antibiotics is indicated in patients with rheumatic heart disease.
- Patients with prosthetic heart valves, previous endocarditis, cardiac transplant recipients with valve regurgitation, congenital heart disease repaired with foreign material within six months of repair, and unrepaired cyanotic heart disease are at highest risk for infective endocarditis; therefore, antibiotic prophylaxis is indicated.

## ANTICOAGULATION / PROSTHETIC VALVES:

- Certain valve lesions will require anticoagulation and should be monitored by a cardiologist at a center with expertise in anticoagulation.
- All pregnant patients with prosthetic valves should undergo clinical evaluation and a baseline TTE evaluation by a cardiologist experienced in valvular heart disease. Pregnancy should be monitored at a tertiary care center with a dedicated heart team.
- Anticoagulation should be given to all pregnant patients with mitral stenosis and atrial fibrillation unless contraindicated.
- All pregnant patients with mechanical valves should ideally be maintained on warfarin in therapeutic range in the second and third trimester. Warfarin crosses placenta and therefore should be discontinued close to the delivery time. Specifically, the current recommendation is to stop warfarin no later than 36 weeks' gestation and start therapeutic doses of unfractionated heparin (UFH) prior to planned vaginal delivery. In the first trimester, it may be reasonable to utilize warfarin if the dose is less than 5 mg. Warfarin may be teratogenic at high doses but birth defects are rare at low doses. Other options include UFH or low molecular weight heparin (LMWH). Due to the unpredictable response and side effect profile of prolonged use of UFH, LMWH is preferred. However, LMWH should not be given unless LMWH levels are frequently monitored 4 hours post-dose to consistently maintain target anti Factor X-a level of 0.8 U/mL to 1.2 U/mL (LMWH level).
- All pregnant patients with mechanical and bioprosthetic valves should additionally be maintained on a daily low dose aspirin 75-100 mg in the second and third trimesters.

## REFERENCES

1. Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. *J Thorac Cardiovasc Surg.* 2014;148(1):e1-e132.
2. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. *J Am Coll Cardiol.* 2008;52(23):e143-263.
3. Yancy CW, Jessup M, Bozkurt B, et al. 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. *J Am Coll Cardiol.* 2013;62(16):e147-239.