

Pregnancy and Congenital Heart Disease

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Introduction:

Most women with congenital heart disease will do well during pregnancy. To ensure that pregnancy will be well tolerated, planning for the care of a pregnant patient with congenital heart disease starts prior to conception and continues after delivery. Ideally, appropriate counseling should begin during pediatric care. The following provides guidelines for the general care for all women with congenital heart disease. This link will provide more comprehensive, disease specific information. <http://www.heartdiseaseandpregnancy.com/>

Key Components:

- Preconception Counseling (Wald, 2009)
 - What are the risks of pregnancy?
 - Evaluate risks related to both mother and baby
 1. Increased risk to mother, pregnancy discouraged
 - a. Severe pulmonary hypertension
 - b. Severe left heart obstructive lesions
 - c. Marfan syndrome with increased aortic root diameter
 - d. Cardiomyopathies with ventricular dysfunction
 2. Increased risk to fetus
 - a. Poor maternal functional class
 - b. Cyanosis
 - c. Left heart obstruction
 - Evaluation of maternal risks include:
 1. Thorough history
 2. Physical examination
 - a. 4 extremity blood pressures
 - b. Oxygen saturation
 3. 12-lead electrocardiogram
 4. Cardiopulmonary exercise testing
 5. Cardiac imaging
 - a. Echocardiogram
 - B. MRI/CT imaging as needed

- Is the patient on medications that would need to be discontinued prior to pregnancy?
 - Cardiovascular medications
 1. Almost all cardiovascular medications cross the placenta
 2. Use medications with lowest risk profile
 - Medications with increased risk profile
 1. Angiotensin-converting enzyme (ACE) inhibitors
 2. Anticoagulants – especially warfarin
 3. Antiarrhythmics
- What type of cardiac surveillance would be required during pregnancy?
 - Maternal surveillance
 1. Follow plan created prior to conception
 2. Determined by risk factors and tolerance to physiological changes that normally occur during pregnancy
 - Fetal surveillance
 1. Fetal echocardiography by 18-20 weeks gestation
 2. Follow up as indicated
- Are there genetic testing that should be done prior to pregnancy?
 - All patients with congenital heart disease should be counseled on the risk of their fetus having congenital heart disease.
 - 22q11 deletion testing is recommended for all patients with (Genetic Basis for congenital heart defects, Circ 2007)
 1. Tetralogy of Fallot
 2. VSD with aortic arch anomaly
 3. Truncus arteriosus
 4. Interrupted aortic arch
 5. Discontinuous branch PA's
- Pregnancy
 - For a low risk pregnancy most patients can be evaluated around 20 weeks by their cardiologist, and deliver in the community.
 - All patients should have a fetal echo at 20 weeks.
 - For patient with moderate to severe risk, cardiac and high risk obstetrics care should be provided at a center with expertise in congenital heart disease.
- Interdisciplinary Care Meetings
 - Care meetings that include representation from the ACHD cardiology team, high risk obstetrics and anesthesia are of great benefit. Other teams such as pulmonary hypertension, electrophysiology can be included on an as needed basis.
 - Initiated by preconception plan
 - Frequency and team members will vary based upon:
 1. Needs of both mother and baby

- 2. Services provided at delivering hospital
 - Needs to clearly communicated to all facets of care
 - Needs to include both mother and father
- Held at least bi-monthly
- Topics covered should includes
 - Cardiac diagnosis (diagrams are very helpful)
 - Potential risks of pregnancy
 - Identification of all members of the care team with contact information
 - Due Date
 - Planned mode of delivery
 - Planned location of delivery and post-partum care
 - Need for a specialized nursing plan
 - 1. For very high risk deliveries the presence of a critical care RN in the delivery room
 - 2. For post-partum care delivered in a critical care unit there should be a plan for scheduled delivery of post-partum nursing care by an obstetric RN.
 - Discussion regarding medication regime and ability of patient to breast feed.

References:

Pierpont ME, Basson CT, Benson DW Jr, et al: Genetic basis for congenital heart defects: current knowledge: a scientific statement from the American Heart Association Congenital Cardiac Defects Committee, Council on Cardiovascular Disease in the Young: endorsed by the American Academy of Pediatrics, *Circulation* 115(23):3015-38, 2007.

Wald RM, Colman JM: Pregnancy and contraception. In Warnes CA, editor: *Adult Congenital Heart Disease*, Oxford, 2009, Wiley-Blackwell.