

## **Pregnancy and Congenital Heart Disease**

What the Nurse Caring for a Patient with Congenital Heart Disease Needs to Know

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### **Introduction:**

Most women with congenital heart disease (CHD) will reach child-bearing age. Many women with complex CHD have a moderate to high risk for both the mother and her fetus during pregnancy. For women wishing to have children, planning for pregnancy for a patient with CHD starts prior to conception and continues after delivery. All practitioners involved in the care of a patient need to be aware of the most up-to-date guidelines and information about CHD and pregnancy. Ideally, appropriate counseling should begin during pediatric care. The following document provides guidelines for the care of pregnant women with CHD. This link will provide more comprehensive, disease specific information. <http://www.heartdiseaseandpregnancy.com/>

### **Key Components:**

- Preconception Counseling (Wald, 2009)
  - Recommended for any patient with CHD
  - Risks of pregnancy
    - Evaluate risks related to both mother and baby
    - Increased risk to mother, pregnancy discouraged
      - Severe pulmonary hypertension
      - Severe left heart obstructive lesions
      - Marfan syndrome with increased aortic root diameter
      - Cardiomyopathies with ventricular dysfunction
    - Increased risk to fetus
      - Poor maternal functional class
      - Maternal cyanosis
      - Maternal CHD with left heart obstruction
- Prenatal History (Canobbio, 2017)
  - Evaluation of maternal risks is most reliable when using a risk algorithm.
  - Risk scores/stratification systems are helpful to identify at risk patients to predict/plan for adverse maternal cardiac events.

- Recommendations are supported by the World Health Organization (WHO) and the American Heart Association (AHA).
- The Modified WHO Classification of Pregnancy Risk:
  - Class I conditions:
    - Associated with no detectable increased risk of maternal mortality and no/mild increase in morbidity
    - Conditions include uncomplicated, small patent ductus arteriosus, mild pulmonic stenosis, or mitral valve prolapse; successfully repaired simple lesions (atrial or ventricular septal defect, patent ductus arteriosus, or anomalous pulmonary venous drainage); and isolated atrial or ventricular ectopic beats
  - Class II conditions:
    - Associated with small increased risk of maternal mortality or moderate increase in morbidity
    - Conditions include unrepaired atrial or ventricular septal defect, repaired tetralogy of Fallot, most arrhythmias
  - Class II to III conditions:
    - Depends on individual
    - Conditions include mild left ventricular impairment, hypertrophic cardiomyopathy, native or bioprosthetic valvular heart disease not considered WHO I or IV, repaired coarctation, Marfan syndrome with aortic dimension <40mm without aortic dissection, and bicuspid aortic valve with ascending aorta diameter <45mm
  - Class III conditions:
    - Associated with significantly increased risk of maternal mortality or severe morbidity
    - Conditions include a mechanical valve, systemic right ventricle, Fontan circulation, cyanotic heart disease (unrepaired), other complex congenital heart disease, bicuspid aortic valve with ascending aortic diameter of 45 to 50 mm, and Marfan syndrome with aortic diameter of 40 to 45 mm
  - Class IV conditions:
    - Associated with extremely high risk of maternal mortality or severe morbidity
      - Pregnancy is contraindicated
- Risk Stratification (See Table 1 below)

***Table 1. Risk Stratification of Pregnancy in Women with Congenital Heart Disease***

<b><i>Low</i></b>	<b><i>Moderate</i></b>	<b><i>Very High</i></b>
Atrial septal defect	Repaired heart disease	Pulmonary hypertension
Mild/moderate valvular regurgitation	Mechanical valve replacement	Shunt Lesions with Eisenmenger syndrome
Mild/moderate pulmonary stenosis	Uncomplicated repaired coarctation of the aorta	Severe heart muscle disease (cardiomyopathy)
VSD PDA		

Mild atrial and ventricular arrhythmias	Hypertrophic cardiomyopathy	Uncorrected cyanotic congenital heart disease
Successful repaired lesions: Secundum ASD VSD PDA TAPV		Marfan syndrome or coarctation of the aorta with aortic aneurysm
		Ischemic Cardiomyopathy

- Planning for conception and pregnancy
  - Individualized
    - For a low risk pregnancy
      - Evaluate around 20 weeks by their cardiologist
      - Deliver in the community
      - Fetal echo at 20 weeks.
    - For moderate to severe risk pregnancy
      - Provide care at a center with expertise in CHD
      - Involves individualized plan based upon:
        - Maternal risk factors
        - Complexity of CHD
        - Functional capacity
        - Other existing or potential clinical issues
        - Social situation
        - Insurance coverage
        - Location of high-risk obstetric and adult CHD providers
  - Involves interdisciplinary team
    - Patient, significant other, other family members as desired
    - High-risk perinatal/neonatologists
    - Adult Congenital Heart Disease Cardiologists
    - Anesthesiologists
    - Nurse practitioners
    - Additional specialists as indicated (electrophysiology, hematology, pulmonology, etc.)
  - Interdisciplinary Care Meetings (See Attachment A for suggested content of documentation for team meetings)
    - Initiated by preconception plan/birth plan
    - Frequency and team members will vary based upon:
      - Needs of both mother and baby
      - Services provided at delivering hospital
    - Needs to clearly communicate all facets of care
    - Needs to include both mother and father
    - Held at least bi-monthly

- Topics include:
  - Vary depending on events during pregnancy
  - 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
  - Cardiac Monitoring during and/ after deliver
  - IV lines: CVP? Art line?
  - Specialized nursing plan
  - For very high risk deliveries consider the presence of a critical care RN in the delivery room
  - For delivery in a critical care unit consider the presence of an obstetric nurse during the delivery and post-partum periods
- Discussion regarding medication regime
- Desire/ability of patient to breast feed
- Presence of family during delivery and post-partum period
- Update of family members during delivery
- Cardiac surveillance required during pregnancy.
  - **Maternal surveillance**
    - Follow plan created prior to conception (See components of plan outlined above and documentation in Appendix A)
    - Determined by risk factors (See Table 1 on risk stratification above)
    - Determined by tolerance to normal physiological changes during pregnancy (See Table 2 below)
      - Fluctuations in circulating blood volume
      - Increase in BNP
      - Decrease in plasma albumin
      - Increase in heart rate with lower threshold for arrhythmias
      - Remodeling of arterial vasculature to accommodate increased blood volume

	<b>Volume</b>	<b>Cardiac Output</b>	<b>Heart Rate</b>	<b>Blood Pressure</b>
<b>1<sup>st</sup> Trimester</b>	↑			↓
<b>2<sup>nd</sup> Trimester</b>	↑↑↑	↑	↑	
<b>3<sup>rd</sup> Trimester</b>	↑	↑	↑↑	

<b>Labor/Delivery</b>		↑↑		
<b>Postpartum</b>	↓	↓	↓	

**Table 2. Normal Physiological Changes during Pregnancy**

- Assessment includes:
  - Physical examination (See Table 3 below for normal and abnormal cardiovascular findings)
  - Diagnostic and genetic testing
  - Symptoms (See Table 4 below)
  - Hemodynamic changes (See Table 2 above)
  - Complications (See Table 5)

**Table 3. Normal and Abnormal Physical Exam during Pregnancy**

<i>Exam</i>	<i>Abnormal</i>
<p><b>Observed color:</b></p> <p>No color changes</p>	<p><b>Observed color findings:</b></p> <p>Cyanosis</p> <p>Clubbing</p>
Dependent edema	
Rales	
Distended neck veins	
<p><b>EKG Changes:</b></p> <p>Nonspecific ST and T-wave changes</p> <p>Shift in electrical axis, leftward to rightward with physical displacement of the heart.</p> <p>PMI: laterally displaced by displacement of heart</p>	<p><b>EKG Findings:</b></p> <p>Arrhythmias</p>



Chest pain	Chest pain with activity
Mild shortness of breath, increased frequency as gestation advances (15% 1 <sup>st</sup> trimester; 75% by 3 <sup>rd</sup> trimester)	Severe shortness of breath with activity
Unable to lie without pillows	Unable to lie without pillows
Rapid breathing	Awakening due to shortness of breath
Palpitations – due to greater cardiac workload and increased blood volume	Palpitations, racing heart, strong, fast, “galloping” heart beats, tachycardia - greater chance due to heart enlargement, may indicate serious problem
Fainting without activity	Fainting with activity

**Table 5. Potential Complications in Pregnancy in Women with Heart Disease**

	<i>Arrhythmias</i>	<i>Heart Failure</i>	<i>Blood clot complications</i>
<i>Intracardiac shunt</i>	√	√	√
<i>Valve replacement</i>	√	√	√
<i>TOF</i>	√	√	*
<i>DORV</i>	√	√	*
<i>TGA</i>	√	√	*
<i>Single Ventricle</i>	√	√	*
<i>CAD/Cardiomyopathy</i>	√	√	√
<i>Pulmonary hypertension</i>	√	√	*
<i>Aortic Coarctation</i>			<b>Aneurysm, ↑ blood pressure</b>
<i>Marfan Syndrome</i>			<b>Aneurysm</b>

○ **Fetal surveillance**

- *All patients with congenital heart disease should be counseled on the risk of their fetus having congenital heart disease.*
- Babies born to women with cyanotic heart disease or reduced cardiac output may experience poorer fetal growth due to lower maternal oxygen or inadequate blood flow during fetal life.
- The risk of heart disease in the baby is higher if either parent has a congenital heart defect. Studies show that the risk of the baby inheriting congenital heart disease from the father is between 1.5 and 3 percent. The risk can be as high as 18 percent if the mother has congenital heart disease.
- It may be helpful for potential parents to speak with experts in Genetics prior to becoming pregnant since a genetic cause in the parent can help

answer questions about the risk of transmitting the genetic condition to the baby. The cause of congenital heart disease is unknown in most cases.

- Risk factors associated with an increased rate of congenital heart disease are shown in Table 6 below. Because of the increased risk of transmitting congenital heart disease, fetal ultrasound is recommended.
- The fetal ultrasound is performed by specially trained sonographers and physicians between 20 and 24 weeks of gestation to check the baby’s heart for congenital defects.
- 22q11 deletion testing is recommended for all pregnant patients with:
  - Tetralogy of Fallot
  - VSD with aortic arch anomaly
  - Truncus arteriosus
  - Interrupted aortic arch
  - Discontinuous branch PA’s
- Follow up as indicated by identified risk factors or fetal diagnosis of CHD

**Table 6. Fetal Risk Factors Associated with Congenital Heart Disease**

Maternal alcohol or drug abuse during pregnancy
Exposure to certain environmental agents (pesticides, lead)
Maternal CHD – risk of fetal CHD increased by 18%
Paternal CHD – risk of fetal CHD increased by 1.5-3%
Maternal heart disease with cyanosis or reduced cardiac output – increased risk of intrauterine growth retardation (IUGR)
Maternal viral infection, such as German measles
Maternal fever early in pregnancy or around conception
Maternal diabetes (not gestational diabetes)
Maternal obesity
Poor maternal nutrition
Chromosomal or genetic abnormalities (Down syndrome) in the fetus
Certain medications taken during pregnancy (ACE inhibitors, Coumadin)

- Medication management during pregnancy
  - Identify any medications that would need to be discontinued prior to pregnancy

- Remember that almost all cardiovascular medications cross the placenta
- Anticoagulation:
  - All forms increase the risk of:
    - Spontaneous abortion
    - Retroplacental bleeding
    - Stillbirth
    - Fetal death
  - Increased risk of clot formation in pregnancy
    - Hypercoagulable state
    - Platelet adhesion with decreased fibrinolysis
    - Increased risk of valve thrombosis or embolic events
    - Specifically in pts with mechanical valve in MV position
  - Medications
    - Coumadin
      - Safest for mother
      - Teratogenic – see midline deformities
        - 9% risk, less if daily dose < 5 mg
        - Avoid 1<sup>st</sup> trimester
        - Acceptable during 2<sup>nd</sup> trimester to middle of 3<sup>rd</sup> trimester
      - Concern during 3<sup>rd</sup> trimester related to immature fetal liver
    - Heparin
      - Often used in 1<sup>st</sup> trimester until 13-14 weeks
        - May then resume warfarin
        - Increased risk of clot
          - Prosthetic valve in mitral position
          - Disc type of prosthetic valve
          - TPA/streptokinase does not cross placenta.
      - Dose – prolong aPTT by 2X control 6 hours after administration initiated
    - Low molecular weight Heparin (LMWH)
      - Increased risk of thrombosis in prosthetic valves
      - More predictable bioavailability.
      - Anti-Xa monitoring, 4 hours post dose, weekly with goal of 1.0-1.2 unit/ml
      - Aspirin may be used adjunctively.
    - ASA
      - Acceptable during pregnancy
      - Dose - 81 mg dose safe without premature fetal duct closure
- Fetotoxic medications
  - Angiotensin converting enzyme (ACE)/angiotensin receptor blocker (ARB)/aldosterone antagonists
    - Strictly contraindicated
    - Increased risk of fetal renal malformation, IUGR

- Alternative afterload-reducing agents
  - Aldosterone
  - Oral isosorbide dinitrate
- Systemic afterload
  - Hydralazine
  - Nitrates
- Antiarrhythmic medications
  - Clinically significant arrhythmias common
    - Increased with history of arrhythmias
    - Lower threshold for ventricular arrhythmias, reentrant SVT
    - Often related to:
      - Extra volume load
      - Enhanced adrenergic receptor excitability
      - Presence of surgical scar
  - Place patients with documented arrhythmias during pregnancy on continuous cardiac monitoring (direct or tele monitoring) throughout labor, delivery, and the postpartum period
  - Medications
    - Beta blockers
      - Extensively used
      - Generally safe, except for atenolol
      - Atenolol
        - Associated with pre term labor
        - Crosses placenta
          - Associated with fetal growth retardation
          - Newborn bradycardia
          - Hypoglycemia.
    - Calcium channel blockers
      - Most experience with verapamil
      - Less with diltiazem and nifedipine
      - Generally safe
      - Use with caution with magnesium
      - Less desirable near delivery/breastfeeding.
    - Digoxin
      - Extensive experience
      - Crosses placenta but not associated with teratogenicity
    - Diuretics
      - Generally safe
      - Aggressive use may decrease placental blood flow
      - Limited data on safety of aldactone

### **LESION Specific Concerns During Pregnancy**

- Complications in women with CHD (See Table 5 above for summary of potential complications in women with CHD)
  - Associated with specific lesions
    - Aortic valve (AV) stenosis

- Mild/Asymptomatic-> well tolerated
- Moderate->usually well tolerated, provided normal LV function
- Severe (AVA<1cm<sup>2</sup>, mean grad >50 mmHg)
  - Decreased PVR
    - Will exaggerate gradient
    - May provoke symptoms
    - Management
      - Bedrest
      - Beta blockers to increase diastolic filling time
      - May consider balloon valvuloplasty – shield gravid uterus
    - Acute afterload reduction following delivery is Particularly dangerous
    - May require invasive monitoring for 24 hours during and post delivery
- Pulmonary Stenosis
  - Usually well tolerated unless RV hypertension present
  - See accelerated degeneration of a bioprosthetic valve in any position
    - May occur during or shortly after delivery.
- Mitral Stenosis
  - Common problem in areas where rheumatic heart disease is endemic.
  - Increased HR and volume lead to:
    - Increased atrial stretch
    - Pulmonary congestion
  - Mild to moderate – management
    - Beta blockers
      - Slow heart rate
      - Increase diastolic filling time
    - Aspirin
      - Decreases the risk of embolic events
    - Diuretics
      - Use with caution
      - Improve volume status
  - Severe – may consider balloon valvuloplasty
- Simple Congenital Lesions
  - Atrial septal defect (ASD)
    - Well tolerated, even large defects
    - Exceptions
      - Pulmonary hypertension (PH)
      - Atrial fibrillation
    - Risk of paradoxical embolism increased
  - Ventricular septal defect (VSD)
    - Usually well tolerated

- Unless PH present
- Patent ductus arteriosus (PDA)
  - Small->well tolerated
  - Large->increased volume load
  - High risk if PH is present
- Aortic abnormalities
  - Coarctation
    - Generally well tolerated
    - May assist stage II with a C-section
    - Deliver by C-section for any concern for aortic instability
  - Aortic Risks in connective tissue disease
    - Greatest risk of complications including aortic dissection – last trimester and early post-partum period
    - Risk stratification:
      - Growth rate
        - Normal = 1.3 mm/year
        - Increased = 1.9 mm/year with bicuspid AV
        - Rate of progression
      - Aortic diameter
        - Size  $\geq 45$ mm – should replace prior to pregnancy or deliver with C-section
    - Age
    - BSA
    - Family history of dissection
    - Connective tissue disease (Syndromes: Marfan, Turners, Ehlers-Danlos, Loeys-Dietz)
  - Management of conditions with connective tissue disease
    - Marfan Syndrome
      - Increased risk for dissection for aorta  $>40$ - $45$ mm
      - Continue beta blockade
      - Surveillance echo imaging every 6-8 weeks
      - Invasive arterial pressure monitoring and assisted 2<sup>nd</sup> stage with C-section
    - Turner Syndrome
      - Associated with left-sided cardiac abnormalities – BAV (20%), CoA (12%)
      - Increased risk of aortic dilation/dissection (50%), even without risk factors
      - Have increased risk of spontaneous dissection
      - Associated with vascular abnormalities
      - Hypertension (50%)
      - Pregnancy occurs with infertility interventions

- Frequent surveillance echo once aorta > 2cm/m<sup>2</sup> due to increased risk of dissection
      - C-section indicated for aorta  $\geq 27$  mm/m<sup>2</sup>
    - Ehlers-Danlos Syndrome
      - Recommended to deliver all patients by C-section.
  - Tetralogy of Fallot
    - Following surgical correction
      - Risk dependent on RV function, pulmonary insufficiency (PI), and tricuspid regurgitation (TR)
      - Requires assessment of hemodynamics and arrhythmia status prior to pregnancy
  - Ebstein
    - Risk dependent on degree of TR
    - Higher risk in the setting of atrial communication and/or atrial arrhythmias
    - Assess for bypass tracts/SVT risk prior to pregnancy
  - Cyanotic Heart Disease
    - High Risk for mother and fetus
    - Decrease in PVR may worsen cyanosis by increase Qs and decrease Qp
    - Increased chance of paradoxical embolism
    - Assess RV function prior to pregnancy
  - Pulmonary hypertension (HTN)
    - Higher risk if pulmonary pressures >60% systemic
    - Eisenmenger syndrome, mortality approaches 50%
    - Termination should be considered given risks
    - ICU delivery with invasive pressure monitoring
    - Left lateral decubitus positioning may be helpful.
    - Implement DVT Bundle to prevent PE
    - Highest risk period is 24 hours following delivery due to:
      - Sudden shifts in volume status and anemia
      - Acute afterload reduction
      - Profound vagal changes
- Heart-related conditions in otherwise healthy women who become pregnant (gestational)
  - Peripartum cardiomyopathy
    - Rare form of heart muscle weakness
    - Occurs most often in women over 30 years of age
    - Most common during the last trimester of pregnancy or within six months after delivery

- Can be a serious or even life-threatening risk for the mother
- Can put the fetus at risk
- EF<45% in the absence of other causes
- Risk of recurrence reported as high as 50%
- Risk decreases for women with recovered EF
- Pregnancy-induced hypertension (high blood pressure)
  - Can affect the supply of oxygen to both mother and fetus
  - Can increase the risk of stroke and seizures for mother
  - Developmental delay in the fetus.
  - Risk factors for pregnancy-induced hypertension include:
    - Smoking
    - Obesity
    - Diabetes
    - Family history of high blood pressure
    - Multiple birth
  - Typically diagnosed after the 20<sup>th</sup> gestational week
  - Manage with labetalol and methyldopa
  - Consider evaluation for CoA
    - Significantly hypertension
    - Young patient
    - Widened pulse pressure with blunted systolic pressure after 32 weeks gestation
      - Diastolic BP normally decreases up to 10 mmHg at 28-32 weeks gestation, then steadily increases
      - With CoA, diastolic BP decreases more than systolic BP. Results in widened pulse pressure.
- Gestational diabetes (non-insulin-dependent or type 2 diabetes)
  - Can develop during pregnancy
  - Normal blood sugar level after delivery
  - Half will develop type 2 diabetes within 15 years of gestational diabetes
  - More likely to develop gestational diabetes with future pregnancy
- Amniotic fluid embolism
  - Rare
  - Occurs at the time of placental membrane rupture and bearing down (i.e. during contractions) or with cesarean section
    - Amniotic fluid returning to the heart
      - Enters blood vessels in the lungs
      - Causes a rapid heartbeat or shock
    - Amniotic fluid crosses through an intracardiac shunt
      - Enters systemic circulation
      - Causes a stroke or interferes with blood supply to the baby

**References:**

Balci,A., Sollie-Szarynska, K. M., van der Bijl, A. G., et al. (2014). Prospective validation and assessment of cardiovascular and offspring risk models for pregnant women with congenital heart disease. *HEART*, 100, 1373-1381.

Brickner, M. E. (2014) Cardiovascular management in pregnancy: Congenital heart disease. *Circulation*, 130, 273-282.

Canobbio, M. M., Warnes, C. A., Aboulhosn, J., et al. on behalf of the American Heart Association on Cardiovascular and Stroke Nursing; Council on Clinical Cardiology; Council on Cardiovascular Disease in the Young; Council on Functional Genomics and Translational Biology; and Council on Quality of Care and Outcomes Research. (2017). [Management of pregnancy in patients with complex congenital heart disease. A scientific statement for healthcare professionals from the American Heart Association](#) [Published online January 12, 2017]. *Circulation*, 135;XXX-XXX. doi:10.1161/CIR.00000000000000458

Pierpont, M. E., Basson, C. T., Benson, D. W. Jr., et al. (2007). 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.

Wald, R. M., Colman, J. M. (2009). Pregnancy and contraception. In Warnes CA, editor: *Adult Congenital Heart Disease*, Oxford, Wiley-Blackwell.

## APPENDIX A

### Template for Documentation of Perinatal High-risk Team Conference

#### **Patient Care Coordinate Note:**

Providers:  
MFM:  
Cardiologist:

Distance from OHSU:

Cardiac diagnosis:

Last echo:

PMH:  
PSH:  
OB Hx:

Fetal status:

Medications:

CARPREG score:  
NYHA Class:  
Anticoagulation: Y/N  
Bleeding risk: high/low  
Heart failure risk: high/low  
Arrhythmia risk: high/low  
- Preferred first-line antiarrhythmic if high risk:  
Aortic dissection risk: no/high/low  
Stroke risk: high/low

Antepartum plan:

- Fetal echocardiogram
- Anesthesia consult
- Discussion at MFM/ACHD conference: [dates]

Delivery Plan:

- contact ACHD team upon admission for delivery
- Timing of delivery:
- Delivery location: L&D, OR, CVICU
- Post-delivery ICU care needed: Y/N
- ICU bed arrangements made: Y/N
- discussed with ECMO team
- Mode of delivery:
- Cesarean
- Vaginal delivery with second stage assistance
- Vaginal delivery without second stage assistance
- Mode of anesthesia:
- Monitoring: telemetry, I/Os
- IV access/lines:
- Antibiotic prophylaxis: Y/N
- Contraception: - postpartum ppx