Congenital heart disease in pregnancy

K. Babazadeh M.D.
Interventional Pediatric Cardiologist
Babol University of Medical Science

Women of Childbearing Age with Cardiac Disease: Approach to Contraception and Pregnancy

Contraception

- • Patient education regarding contraception options

Preconception

- • Preconceptual counseling should begin in adolescence
- • All women with heart disease should receive preconception counseling
- • Maternal cardiac risk of pregnancy
- • Fetal and neonatal risks during pregnancy
- • The safety of medication use during pregnancy
- • The risk of transmission of congenital heart disease to offspring

Pregnancy

- • For intermediate and high-risk patients, assessment by a multidisciplinary team including cardiologists and obstetricians should occur early in the pregnancy
- • The patient should be advised of antepartum follow-up requirements
- • Women with CHD or who are spouses of men with CHD should be offered fetal echocardiography
- • Peripartum, intrapartum and postpartum management issues

PHYSIOLOGIC CHANGES DURING PREGNANCY

- Maintenance of adequate oxygen delivery is achieved by:

1. maternal circulating blood volume, (in sixth week of gestation about 50% more than the prepregnant state)
2. red cell mass, (as much as 40% above prepregnancy levels)
3. peripheral vascular compliance and resistance, (decreases beginning in the fifth week of gestation)
4. heart rate, (The mean heart rate increases to approximately 10 to 20 beats above prepregnancy levels by term)
5. cardiac output (Increases during pregnancy as a result of increases in both heart rate and stroke volume)

- Increase in cardiac output (fifth week of gestation): early increase in stroke volume, later increase in the heart rate.
- Pregnant women with underlying cardiac disease have lower CO than pregnant women with normal cardiac function.
- Cardiac output can fall acutely if the inferior vena cava is compressed by the gravid uterus in the supine position.
- During labor and delivery pain, anxiety and uterine contractions result in tachycardia, hypertension, and further increases in cardiac output.
- Immediately following delivery, cardiac output may transiently increase to as much as 80% above prelabor values due to relief of inferior vena cava compression and autotransfusion from the placenta.

CARDIAC FINDINGS IN NORMAL PREGNANCY

- Fatigue, dyspnea, light-headedness, and palpitations are normal symptoms associated with pregnancy
- The hemodynamic changes of pregnancy that can mimic cardiac disease: displacement of the apical impulse, prominence of the jugular venous pulsation, wide splitting of the first and second heart sounds, soft systolic flow murmurs, and continuous murmurs.
- Sinus tachycardia and premature atrial or ventricular ectopic beats are normal.
- Electrocardiogram: leftward shift in the frontal axis, T-wave inversion in lead II, and ST-segment depression.
- Echocardiography: increase in dimensions of all four cardiac chambers and left ventricular wall thickness and mass, increases in transvalvular flow velocities, increase in mitral, tricuspid, and pulmonic annular diameters and increase valvar regurgitation.

ASSESSMENT OF PREGNANCY RISK IN WOMEN WITH CONGENITAL HEART DISEASE: GENERAL CONCEPTS AND GLOBAL EVALUATION

- Risk factors of pregnant women with heart disease: poor functional status (NYHA > II) or cyanosis, systemic ventricular systolic dysfunction, left heart obstruction, and history of cardiac events prior to pregnancy (arrhythmia, stroke, or pulmonary edema), the presence of a prosthetic valve or conduit , occurrence of an obstetric complication such as preeclampsia, and use of anticoagulants or teratogenic drugs.
- Based on these predictors: low (0 predictor), intermediate (1 predictor), or high (>1 predictor) risk categories, a 5%, 25%, or >75% chance of developing an adverse cardiac event during pregnancy respectively.
- Adverse fetal and neonatal complications, include fetal or neonatal death, premature birth (RDS, IVH), or SGA neonates.

Risk Factors for Adverse Maternal Cardiac and Fetal/Neonatal Events During Pregnancy in Women with Heart Disease

- Maternal cardiac adverse event

- Risk Factor
- • Poor functional class (NYHA Class Ill or IV) or cyanosis
- • Systemic ventricular ejection fraction <40%
- • Left heart obstruction (mitral valve area <2 cm^2, aortic valve area <1.5 cm^2, or peak LVOT gradient >30 mm Hg)
- • Cardiac event (sustained or symptomatic arrhythmias requiring treatment, stroke, pulmonary edema) prior to pregnancy
- • Subpulmonic ventricular dysfunction
- • Significant pulmonary regurgitation
- • Mechanical heart valves
- • Moderate or severe subaortic or subpulmonary atrioventricular valve regurgitation
- • Known lesion-specific risk

CONTINUE

- Fetal and/or neonatal adverse event

- Risk Factor
- • Poor maternal functional class (NYHA Class Ill or IV) or cyanosis
- • Maternal left heart obstruction (defined above)
- • Maternal age <20 or >35 years
- • Obstetric risk factors
- • Multiple gestation
- • Smoking during pregnancy
- • Anticoagulant therapy
- • Known lesion-specific risk

LESION-SPECIFIC RISKS AND OUTCOMES

Cardiac shunts

- Unrepaired ASD, VSD, PDA have low risk because of the pregnancy-associated fall in peripheral vascular resistance.
- Potential complications: atrial arrhythmias and heart failure, paradoxical embolization if systemic vasodilation and/or elevation of pulmonary resistance result in transient right-to-left shunting (in ASD or patent foramen ovale)
- Atrioventricular septal defects (AVSDs) are more complex, and pregnancy may be less well tolerated.

Right ventricular outflow tract obstruction

- Mild pulmonic stenosis is well tolerated during pregnancy.
- In severe pulmonic stenosis, the increase in preload associated with pregnancy may result in atrial arrhythmias or right heart failure.

Tetralogy of Fallot

- In general, pregnancy is well tolerated, but risk of complications is increased in presence of residual and surgical sequelae (residual shunts, right ventricular outflow tract obstruction, pulmonary regurgitation, right ventricular dilation or dysfunction, and atrial or ventricular arrhythmias).

Left ventricular outflow tract obstruction

- Left ventricular outflow tract obstruction lead to heart failure, ischemia, or hypotension, arrhythmias, and angina.
- Women with symptomatic aortic stenosis should undergo correction prior to pregnancy.
- Selected aortic valvuloplasty prior to a planned pregnancy or during pregnancy.
- Aortic dissection has been reported in women with bicuspid aortic valve and aortopathy.

Coarctation of the aorta

- Persistent or recurrent systemic hypertension may manifest after repair.
- Significant coarctation of the aorta impedes delivery of blood to the arterial tree distal to the coarctation site; that may impact on the placental circulation. Upper body hypertension and concomitant aortic valve disease pose additional risks.
- Women with Turner syndrome with the aortopathy .
- Severe unrepaired coarctation: pregnancy-induced hypertension and preeclampsia, intrauterine growth restriction and premature labor.
- Control of upper body hypertension: hypotension distal to the coarctation site with potential adverse impact on oxygen delivery to the fetus.

Marfan syndrome

- Increased cardiac output and hypervolemia: increased risk of aortic dilation and dissection.
- Ideal aortic root size prior to pregnancy is <45 mm.
- Prophylactic root replacement prior to pregnancy does not guarantee a safe pregnancy (because of possible dissection).
- Preconception counseling
  1. If seen first in early pregnancy they should be offered termination.
  2. Discussion of the increasing risk with increasing maternal age.
  3. 50% chance of transmitting the syndrome to offspring.
- Increased risk for fetal, neonatal, and obstetric complications, and **preterm delivery** due to premature rupture of membranes and cervical incompetence .

Ebstein anomaly

- The ability of the heart to tolerate a pregnancy varies according to the severity of the disease.
  1. Mild Ebstein anomaly(acyanotic): uncomplicated pregnancy,
  2. severe Ebstein anomaly: unable to tolerate the increased preload and cardiac output of pregnancy (functional deterioration, right heart failure, and arrhythmia, increase cyanosis due to right-to-left interatrial shunt).
- Fetal complication: fetal loss, prematurity, and congenital heart disease in the offspring.

Transposition of the great arteries

1. **Atrial switch** operation (Mustard or Senning procedure): late complications include sinus node dysfunction, atrial arrhythmias, systemic ventricular dysfunction, and systemic atrioventricular valve regurgitation, heart failure and deterioration in cardiac function.
2. **Arterial switch** operation (Jatene operation): complications include valve thrombosis in a woman with a mechanical valve and ventricular arrhythmia.
3. **Congenitally corrected transposition of the great arteries**: systemic ventricular dysfunction and systemic atrioventricular valve regurgitation.

- Associated lesions: VSD, pulmonary stenosis, and complete heart block.
- Potential problems: heart failure as a result of a dysfunctional subaortic (systemic) right ventricle and/or increased subaortic (tricuspid) atrioventricular valve regurgitation, atrial arrhythmias, atrioventricular block, and late sequelae from prior surgical interventions (e.g., dysfunction of a right ventricular to pulmonary artery conduit).

Fontan circulation

- Palliation the condition by directing right atrial or caval blood into the pulmonary artery (thus improves cyanosis and volume overload of the subaortic (systemic) ventricle)
- Limitation in increasing in cardiac output, scarring and remodeling of the atria (atrial arrhythmias and atrial thrombi), supraventricular tachycardia, atrial fibrillation, and deterioration of NYHA functional class.
- Fetal and neonatal adverse outcomes remain common
- The incidence of first trimester miscarriage is high.

Cyanotic heart disease

- Adverse maternal cardiac events (in 32%): heart failure, arrhythmias, pulmonary artery thrombosis, and cerebral infarction.
- In low maternal oxygen saturation (≤85%) the live birth rate is 12%.
- Eisenmenger syndrome: decrease in afterload can lead to increase in right- to-left intracardiac shunting (increasing hypoxemia and cyanosis)
- Volume depletion and hypotension during labor and delivery (fatal within the first few weeks postpartum).
- Preconceptual counseling should advise against pregnancy and termination of pregnancy .
- If pregnancy continues, the use of pulmonary vasodilators may reduce PAP.
- Adverse fetal events in Eisenmenger syndrome (common): Perinatal mortality and prematurity(in 37%)

Prosthetic heart valves

- Risks of complications : depends on the type of valve and its position and the type of anticoagulant used.
- Normally functioning bioprosthetic valves often are tolerated well.
- Pulmonary autograft aortic valve replacement (Ross procedure) are tolerated well.
- Mechanical valve prostheses have increased risk for thromboembolic events (4% to 9% of pregnancies).
- Warfarin embryopathy (5 mg of warfarin per day)

TRANSMISSION OF CARDIAC DISEASE TO OFFSPRING

- Genetic counseling should be offered.
- In patients with congenital heart disease, the recurrence risk of CHD to offspring is approximately 3% to 5%.
- Parental left heart obstructive lesions are associated with higher rates of transmission (13% to 18%).
- Autosomal dominant conditions such as Noonan syndrome, Williams syndrome, Holt Oram syndrome, Marfan syndrome, or 22q11.2 deletion syndrome confer a 50% risk of recurrence in an offspring.
- Preconception use of multivitamins containing folic acid
- A fetal echocardiogram is indicated.

MANAGEMENT ISSUES DURING PREGNANCY

- Guidelines from the American Heart Association/American College of Cardiology, the Canadian Cardiovascular Society, and the European Society of Cardiology.

1-Preconception Issues

- Preconceptual counseling should be offered to all women with cardiac disease (assessment of the maternal risk of pregnancy and the effects of the maternal cardiac condition on fetal outcomes).
- The risks and benefits of drug therapy .
- Termination of the use of the teratogens such as alcohol, hydantoin, lithium, retinoic acid, valproic acid, and warfarin.
- Adjustment of drug dosing and frequency of administration (because of changes in volume of distribution, glomerular filtration rate, and hepatic metabolism).

2-Antepartum Issues

- Women with low risk heart disease: management in local obstetric units.
- Women with intermediate or high risk for complications: should receive care in a high-risk obstetrics unit.
- The optimal frequency of cardiac follow-up: an early assessment in first trimester, another follow-up near the end of the second trimester (Peak cardiac output occurs at this time), a third visit around the end of the eighth month.
- Echocardiography is the cardiac imaging modality of choice.
- Cardiac MRI should be performed after the first trimester (Gadolinium is contraindicated during pregnancy).
- Ionizing radiation (computed tomography, cardiac catheterization, nuclear imaging) should be avoided during pregnancy.
- Ambulatory ECG monitoring is useful in symptomatic palpitations.

3-Management of Heart failure

- Women with limited cardiac reserve: at risk of heart failure because of increased hemodynamic burden of pregnancy, gestational hypertension, hyperthyroidism, and anemia.
- Women with symptomatic heart failure: activity limitation, oxygen, diuretics, and afterload-reducing agents such as hydralazine.
- Nitroprusside is toxic to the fetus
- Women with preexisting systemic ventricular dysfunction: beta-blockers usage with caution
- Digoxin is safe in pregnancy.
- Angiotensin-converting enzyme inhibitors and angiotensin receptor blockers are contraindicated.

4-Management of Arrhythmias

- The hemodynamic changes of pregnancy may provoke arrhythmias.
- History of arrhythmias: increase risk for adverse maternal cardiac events like arrhythmia recurrences and then increase in adverse fetal/ neonatal events.
- Pharmacologic therapies: just in patients with severe symptoms.

1. Paroxysmal SVT (AVNRT and AVRT): beta-blockers

- Acute management of SVT: intravenous adenosine or beta-blockers
- Amiodarone: contraindicated (impaired neonatal thyroid function)
- Cardioversion is safe.

2. Ventricular tachycardia: beta-blockers

- Acute management of VT: intravenous procainamide, sotalol, amiodarone, or beta-blocker

3. Bradyarrhythmias: Pacemakers and ICD

5-Management of Anticoagulation

- Pregnancy increase the risk of thrombosis and thromboembolism.
- Options include warfarin, unfractionated heparin, low-molecular-weight heparin (LMWH), and adjunctive aspirin.
- Warfarin has potential for embryopathy.
- Warfarin: feral intracranial bleeding
- Warfarin replacement by heparin at least 2 weeks prior to labor
- Heparin: maternal thrombocytopenia and osteoporosis
- For women with mechanical valves: Guidelines for the use of anticoagulants have been offered by the American Heart Association/American College of Cardiology, the American College of Chest Physicians, and the European Society of Cardiology.

6-High-Risk Conditions

- Cyanotic cardiac lesions or symptomatic obstructive lesions: should be repaired prior to conception.
- Women with Eisenmenger syndrome: early hospitalization, supplemental oxygen and possibly empiric anticoagulation, pulmonary vasodilators
- Marfan syndrome: risk increases in proportion to aortic root size
- Prophylaxis in Marfan syndrome: beta-blocker

7-Cardiac Surgery During Pregnancy

- Cardiovascular surgery during pregnancy: significant maternal and feral mortality
- Feral adverse events: provocation of uterine contractions in association with extracorporeal circulation, maternal hypotension and consequent placental hypoperfusion (fetal hypoperfusion, hypoxia, and bradycardia).
- Fetal mortality vary with maternal age >35 years, maternal functional class, reoperation, emergency surgery, the type of myocardial protection, and anoxic time.

MANAGEMENT ISSUES DURING LABOR AND DELIVERY

- Caesarean delivery: in women with aortic dissection or Marfan syndrome with dilated aortic root or if there has been a failure to switch from warfarin to heparin at least 2 weeks prior to labor.
- Vaginal delivery: all other pregnancies in the absence of obstetric contraindication
- Preterm induction of labor is rarely indicated.
- Subcutaneous administration of unfractionated heparin should be discontinued at least 12 hours prior to induction
- Epidural anesthesia is the anesthetic technique of choice.
- Epidural fentanyl: does not lower peripheral vascular resistance (ideal in cyanotic patients or in patients with significant aortic stenosis).
- Intra-arterial monitoring and central venous pressure monitoring: ideal in severe AS, PH, severe systemic ventricular systolic dysfunction, or preload-dependent physiology (Fontan operation).
- Prevention of Infective Endocarditis is not necessary for uncomplicated vaginal deliveries based on AHA guideline

Risk Classification for the Use of Combined Hormonal Contraceptive in Women with Congenital Heart Disease

WH01
Always useable Condition with no restriction for the use of the contraceptive method

- • Mitral valve prolapse with trivial mitral regurgitation
- • Bicuspid aortic valve with normal function
- • Mild pulmonary stenosis
- • Repaired coarctation with no hypertension or aneurysm
- • Simple congenital lesions successfully repaired in childhood and with no sequelae (atrial or ventricular septal defect, PDA or total anomalous pulmonary venous drainage)

WH02
Broadly useable
Condition where the advantages of the method generally outweigh the theoretical or proven risks

- • Most arrhythmias other than atrial fibrillation or flutter
- • Uncomplicated mild native mitral and aortic valve disease
- • Tissue prosthetic valve lacking any of the features noted in WHO 3 or 4 columns
- • Surgically corrected congenital heart disease lacking any of the features noted in WHO 3 or 4 columns
- • Small left-to-right shunts not reversible with physiologic maneuvers (i.e., small VSD, small PDA)
- • Hypertrophic cardiomyopathy lacking any WHO 3 or 4 features
- • Past cardiomyopathy, fully recovered, including peripartum cardiomyopathy
- • Uncomplicated Marfan

WH03
Caution in use
Condition where the theoretical or proven risks usually outweigh the advantages of using the method

- • Atrial fibrillation or flutter even on warfarin
- • Bileaflet mechanical valve even on warfarin
- • ASD with left-to-right shunt that may reverse with physiologic stress (i.e., Valsalva maneuver)
- • Repaired coarctation with aneurysm and/or hypertension
- • Previous thromboembolism

WH04
Do not use
Condition that represents an unacceptable health risk if the contraceptive method is used

- • Atrial fibrillation or flutter, if not anticoagulated
- • Bjork Shiley or Starr Edwards valve even if taking warfarin
- • Fontan circulation even if on warfarin
- • Cyanotic heart disease
- • Pulmonary arteriovenous malformation
- • Prior left ventricular dysfunction from any cause (i.e., dilated cardiomyopathy) (LVEF <30%)
- • Coronary artery disease
- • Coronary arteritis (i.e., Kawasaki disease with coronary involvement)

با تشکر از توجه شما