The Adolescent and Adult Congenital Heart Disease Program

The Heart Center at Columbus Childrens Hospital
The Ohio State University

Reproductive Health, Pregnancy and Congenital Heart Disease

Congenital Heart Disease in Women:
Statistics have shown a dramatic increase in survival of individuals with congenital heart disease. Today, we expect that most children treated for congenital heart disease will survive to adulthood and lead a full adult life. Thus, more women with congenital heart disease are interested in pregnancy. **Although most women with congenital heart disease can successfully carry a pregnancy, some are at substantial risk.** We strongly encourage all adolescent girls and women with congenital heart disease to discuss reproductive health care and pregnancy with their OB-Gyn doctor and cardiologists, **prior to becoming pregnant.**

Menstruation:
Menarche is the onset of menstrual periods. Compared to adolescent girls without heart disease, menarche occurs slightly later in girls with congenital heart disease, and this is especially true for women with bluish discoloration of the skin, lips, fingernails, and tongue (cyanosis). Women with cyanosis may also be more likely to have irregular menstrual periods. All women 18 years of age and older, and particularly those that are sexually active should have annual gynecologic examinations (pelvic, pap smear, breast examination). Gynecological examinations are important for reproductive health, education related to sexuality, sexually transmitted diseases (STDs), pregnancy and birth control options.

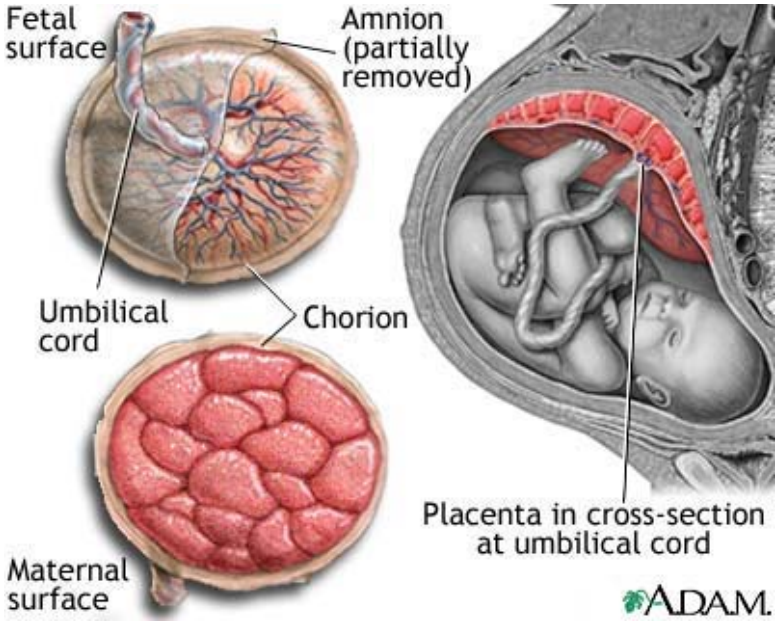
Birth Control:
The reliability, advantages and disadvantages of the more common forms of birth control are shown in ***Table 1***. Some forms of birth control are not appropriate in women with certain forms of congenital heart disease, and we recommend women discuss the various options with her health care providers to select the safest and most appropriate. In general, women with complex heart disease, cyanosis, or pulmonary hypertension, should **not** use birth control pills that contain estrogen because there is an increased of forming blood clots. Depo-Provera and progesterone only birth control pills may be better alternatives in some patients. Intrauterine devices (IUDs) may place some women at higher risk of endocarditis. These women should be given antibiotics prior to insertion of an IUD.

Table 1: Birth Control Options

Method	Lowest expected failure rate	Typical Failure rate in 1st Year	Advantages	Disadvantages
Abstinence	0%	0%	Consistent abstinence provides 100% reliable contraception and avoids contracting STDs	Requires discipline, commitment
Fertility Awareness	1%	20%	No hormones	Requires daily monitoring of symptoms Must be abstinent during "unsafe" days Does not protect from STDs
Male Condoms	3%	14%	Condoms protect against STD (Animal condoms do not prevent against HIV) Available without prescription	Decreased spontaneity; Messy Nonoxyl 9 spermicidal may make it easier to contract HIV
Diaphragm, cervical cap, sponge	6-9%	12-20%	No hormones Don't affect menstrual cycle	Must be inserted at time of intercourse
Birth control pills, Patch, Vaginal Ring	<1%	8%	Reduced menstrual flow and cramping, Regulation of periods; Decrease in anemia Less acne Lowers risk of ovarian and uterine cancer	No STD protection Breakthrough bleeding Nausea, breast tenderness, mood swings Increase risk of blood clots Effectiveness reduced with some medications
Depo-Provera Injection	<1%	<1%	12 week effectiveness Privacy Safe during breastfeeding Menstrual cycles may cease	Requires injection every 90 days No STD protection,Weight gain; Hair loss Unpredictable menstrual cycles May cause bone loss over long term use
Intrauterine Device (IUD)	<1%	<1%	Spontaneity May be left in for up to 5 or 10 years IUD with hormones reduces menstrual cramps and bleeding	Cramps and heavier/longer periods with copper IUD No periods or light periods with levonorgestrel IUD Requires insertion by health care provider
Permanent Sterilization	<1%	<1%	Highly reliable Permanent	Very difficult or impossible to reverse No STD protection Requires surgery
Emergency Contraception	25%	25%	Pills given as soon as possible after unprotected sex (Plan B)	Not as effective as regular use of other methods

Pregnancy:

Pregnancy is a special time in every woman's life. At around 12 days after conception, an extensive vascular system (placenta) develops to provide nutrients for the baby and eliminates its waste products. The placenta is implanted in the wall of the uterus, where it receives Mother's blood, and carries out wastes from the baby. The placenta is a barrier that filters out harmful substances to the baby, however not all substances are filtered. Alcohol, viruses and some medications can cross the placenta and can be harmful to the developing baby.



The extra demand for nourishment (oxygen) and removal or wastes increases the work load on the mother's cardiovascular system. In response to the increased demands, the mother's heart must beat faster and stronger. As the pregnancy advances toward term, the mother's cardiovascular demands increase and pregnancy mimics mild exercise. Although most women tolerate pregnancy well, most experience symptoms related to the changes that occur with pregnancy. The effects of pregnancy, labor and delivery and postpartum are summarized in ***Table 2.*** Normal and abnormal symptoms during pregnancy are summarized in ***Table 3***.

Table 2: Cardiac Physiology Changes that Occur During and After Pregnancy

	Volume	**Cardiac Output**	**Heart Rate**	**Blood Pressure**
1st Trimester	↑			↓
2nd Trimester	↑↑↑	↑	↑	
3rd Trimester	↑	↑	↑↑	
Labor/Delivery		↑↑		
Postpartum	↓	↓	↓	

Table 3: Normal and Abnormal Symptoms during Pregnancy

Normal	***Abnormal***
Fatigue	Symptoms at rest
Chest pain	Chest pain with activity
Mild shortness of breath	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	Palpitations; racing heart
Fainting without activity	Fainting with activity

Prior to becoming pregnant:
Women with heart disease are strongly urged to use a reliable form of birth control until they are emotionally and physically ready to become pregnant. Although many women with congenital heart disease can have a successful pregnancy, it is important that you discuss pregnancy with your cardiologist ***before*** becoming pregnant. It will be necessary for each potential mother with congenital heart disease to undergo a complete cardiac evaluation prior to becoming pregnancy. The evaluation may be as simple as a history and physical examination. In more complex congenital heart disease, the evaluation may also include EKG, echocardiography, transesophageal echocardiography (TEE), stress testing, cardiac MRI or CT scan, Holter monitor, and/or electrophysiology study (EPS). The purpose of the preparatory evaluation is to ensure the healthiest outcome for Mother and Baby.

***Medications and pregnancy*:**
Prospective mothers should discuss medication use with experts in OB-Gyn, primary care and/or cardiology team prior to becoming pregnant since some medications are potentially harmful to the baby. Moms with heart disease may be advised to continue heart medications when this is healthiest for Mom, and therefore would be healthiest for the baby. It is extremely important to review all medications (prescription and over-the-counter) because many medications used to treat heart problems can adversely affect the baby. Any unnecessary medications are discouraged.

High Risk Maternal-Fetal Health Care:
Women with heart disease are advised ***expert obstetric care*** from a team experienced with a variety of heart conditions and the effect of pregnancy on those conditions. Such a team is introduced to the Mother early in pregnancy, follows the mother and baby during pregnancy and together, plans the induction of labor and delivery. Induction and planned delivery ensure that the appropriate team members (high risk obstetrician, cardiologist, etc) are available at the time of delivery. Prior to the planned induction, you will have a chance to discuss anesthesia and method of delivery (vaginal or Cesarean section). We work with Dr. David Colombo and his colleagues in the High Risk Maternal-Fetal Unit at Ohio State University. Dr. Colombo's patients deliver at Ohio State University. Vaginal delivery is the usual and preferred method of delivery. Cesarean section is usually necessary because of problems with delivering the baby (i.e. breech presentation).

Potential Complications in Women with Heart Disease

	Arrhythmias	***Heart Failure***	***Blood clot Complications***	***Other***
Intracardiac shunt	√	√	√*	
Valve replacement	√	√	√	
TOF	√	√	√*	
DORV	√	√	√*	
TGA	√	√	√*	
Single Ventricle	√	√	√*	
CAD/Cardiomyopathy	√	√	√	
Pulmonary hypertension	√	√	√*	
Aortic Coarctation				**Aneurysm ↑ blood pressure**
Marfan Syndrome				**Aneurysm**

Intracardiac shunt=ASD/VSD (atrial/ventricular septal defect); Valvular stenosis=aortic, mitral, pulmonary or tricuspid stenosis; TOF=tetralogy of Fallot; DORV=double outlet right ventricle; TGA=transposition of the great arteries; Single ventricle=tricuspid, pulmonary atresia; CAD=coronary artery disease; *-increased risk of blood clotting complications is due to unrepaired or residual intracardiac shunt or fenestration

Arrhythmias:
The baby's needs are met by increased blood volume and output from Mother's heart. Increased blood volume and stretch in Mother's heart can cause Mother's heart to skip or beat fast. Cardiac arrhythmias can be dangerous, particularly in women with heart disease. Thus, the occurrence of palpitations and racing should be reported to the cardiology team. Rhythm disturbances may require monitoring and medications.

Heart Failure:
When the heart cannot keep up with the demands of pregnancy, women may experience heart failure symptoms-fatigue, shortness of breath, swelling in the legs and abdomen. For some women, these symptoms appear or worsen in the second or third trimester of pregnancy. Pregnant women with heart failure need to be watched closely and may require medications, bed rest, or hospitalization with special IV therapy.

Mechanical Heart Valve Replacement:

Strong blood thinning medications (warfarin/Coumadin) keep mechanical heart valves functioning normally. Changes during pregnancy such as weight gain and increased breakdown of medications can alter the correct dose of blood thinning medications, requiring more frequent blood thinner testing, and close communication with your physicians or nurses. Fetuses whose mothers require blood thinning medications (warfarin; Coumadin) are at increased risk of developing birth defects. The baby is also at increased risk of bleeding. Enoxaparin (Lovenox) injections or subcutaneous heparin injections can be substituted in some cases. Although there is an increased risk of birth defects, warfarin/Coumadin is more reliable in preventing mechanical valve failure due to blood clot formation. Some women choose heart surgery to undergo removal of the mechanical valve with a valve made from human or animal tissues (bioprosthetic) in order to avoid the need for blood thinning medicate.

Other:

In addition the increased workload on the heart, pregnancy causes the mother's blood system to produce more blood clotting factors. This helps protect Mother from excessive bleeding as her placenta (a rich vascular organ) is delivered from the uterus. Although the increased tendency to clotting is beneficial, there is an increased chance that small blood clots formed in the uterus will be carried in the blood stream back to the heart, where they are filtered by small vessels in the lungs and are eventually dissolved. In women with small holes in the heart (atrial septal defect-ASD; ventricular septal defect-VSD; or small communications between the veins and arteries within the lungs) there is an increased risk that blood clots will bypassing the lungs and flow across the hole in the heart, and cause serious complications such as stroke. Larger holes in the heart cause oxygen-poor blue blood to mix with oxygen-rich red blood, causing a bluish discoloration of the skin, lips and mouth (cyanosis). Thus, all women with "cyanotic" heart disease are advised to take blood thinning medications (aspirin, Coumadin, heparin, Lovenox) to help prevent complications caused by blood clot formation during pregnancy, labor and after delivery.

Risk Stratification of Pregnancy in Women with Congenital Heart Disease

Low	***Moderate***	***Very High***
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)
Mild atrial and ventricular arrhythmias	Hypertrophic cardiomyopathy	Uncorrected cyanotic congenital heart disease
		Marfan syndrome or coarctation of the aorta with aortic aneurysm
		Ischemic Cardiomyopathy

Fetal Risk:
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 4***. 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.

Table 4: Risk Factors Associated with Congenital Heart Disease

Alcohol or drug abuse during pregnancy
Exposure to certain environmental agents (pesticides, lead)
Maternal viral infection, such as German measles
Maternal fever early in pregnancy or around conception
Maternal diabetes (not gestational diabetes)
Maternal obesity
Poor nutrition
Chromosomal or genetic abnormalities (Down syndrome) in the child
Certain medications taken during pregnancy (ACE inhibitors, Coumadin)

OPTIONAL INFORMATION

Heart-related conditions in otherwise healthy women who become pregnant (gestational).

Peripartum cardiomyopathy is a rare form of heart muscle weakness that occurs most often in women beyond 30 years of age, and most commonly during the last trimester of pregnancy or within six months after delivery. It can be a serious or even life-threatening risk for the mother and can also put her unborn child at risk.

Pregnancy-induced hypertension (high blood pressure) can affect the supply of oxygen to both mother and fetus and can increase the risk of stroke and seizures for Mother, and developmental delay in the fetus. Risk factors for pregnancy-induced hypertension include: smoking, overweight, diabetes, family history of high blood pressure, and multiple birth.

Gestational diabetes (non-insulin-dependent or type 2 diabetes) can develop during pregnancy, with return to normal blood sugar level after delivery. Approximately one-half of women with gestational diabetes will type 2 diabetes within 15 years of gestational diabetes, and are also more likely to develop gestational diabetes with future pregnancy.