Marked advances in the treatment of congenital heart disease (CHD) have resulted in a growing population of adults with CHD, currently estimated at >1 million adults (at least equal to the pediatric population). Many of these are young women, facing decisions about childbearing, and will require expert care. A recent analysis of a large nationally representative hospital discharge database in the United States demonstrated that annual deliveries for women with CHD increased 34.9% from 1998 to 2007 compared with an increase of 21.3% in the general population. In several recent pregnancy registries, CHD was the most common form of heart disease complicating pregnancy in the Western world (accounting for 74% of cases in the Canadian Cardiac Disease in Pregnancy [CARPREG] registry and 66% of cases in the European Registry on Pregnancy and Cardiac Disease [ROPAC] registry), whereas in less developed countries, rheumatic heart disease plays a larger role Table 1).

This expanding population of patients encompasses a large variety of congenital heart defects with a wide variety of anatomic abnormalities of varying complexity and continually changing surgical and interventional procedures to address these defects. To define the long-term needs of these patients, the 32nd Bethesda Conference in 2001 classified CHD defects as simple, moderate, or complex defects based on the expected outcomes, particularly in the case of single ventricle physiology or surgically palliated defects. There may be preexisting ventricular dysfunction, particularly in the case of single ventricle physiology or with an anatomic right ventricle functioning in the systemic circulation. Arrhythmias are common in adults with CHD as a result of preexisting substrates such as cardiac chamber dilatation, pressure or volume overload leading to myocardial hypertrophy and fibrosis, surgical scars, trauma to the conduction tissues, and presence of patch tissue in the myocardium. Supraventricular arrhythmias are more common than ventricular arrhythmias in this patient population. Arrhythmias may be poorly tolerated in these patients and may result in hemodynamic deterioration. Arrhythmias occurring in the CHD patient during pregnancy may occur as a result of underlying hemodynamic derangements or may be precipitated or worsened by the physiological changes of pregnancy, including the electrophysiological effects of hormonal changes, cardiac dilatation, and changes in autonomic tone. Some patients with CHD are at increased risk of embolic events, and the prothrombotic state of pregnancy may further increase this risk.

Impact on Fetal Outcomes
Fetal outcomes are also impaired in some mothers with CHD. This includes higher rates of intrauterine growth retardation with lower birth weight, increased risk of preterm birth, increased risk of miscarriage or intrauterine demise, and increased perinatal mortality. Decreased maternal cardiac output and maternal cyanosis are significantly associated with an increase in fetal complications. These complications may have long-term implications for the offspring, because lower birth weight appears to be a risk factor for increased metabolic and cardiovascular complications in later life.

General Approach to Pregnancy for Women With CHD
The American College of Cardiology and American Heart Association (ACC/AHA), the European Society of Cardiology (ESC), and the Canadian Cardiovascular Society have all

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© 2014 American Heart Association, Inc.
Circulation is available at http://circ.ahajournals.org
DOI: 10.1161/CIRCULATIONAHA.113.002105
created guidelines addressing the optimal management of ACHD patients, including recommendations for reproductive issues. Most of these guidelines are based on expert consensus opinion, but serve as excellent resources.17–22

Preconceptual Counseling

Optimally, women with CHD should be evaluated before conception to advise potential mothers about contraceptive choices and the risk of pregnancy for themselves and their fetus. These issues should be addressed in adolescence, before sexual activity. However, many young patients with CHD have limited or inaccurate knowledge of their disease, including their birth diagnosis, type of repair, risk for long-term complications, and their prognosis. Their risks for pregnancy may not have been addressed in adolescence.23,24 Contraceptive management is extremely important for this population of patients and should be performed by personnel with expertise in contraceptive options and the specific consequences of such for women with CHD.25

Pregnancy Risk Assessment

Determining which woman are at increased risk for pregnancy requires evaluation of the patient’s birth anatomy and type of repair, assessment for residual lesions and development of sequelae from their original repair, and evaluation of their current functional status. The history of their birth defect and surgical or device interventions should be obtained, which often requires obtaining childhood records. Functional capacity can be measured by history or by formal stress testing as recommended by the ESC guidelines.21 The presence of chronotropic incompetence on formal exercise testing is a valuable predictor of pregnancy outcome.26,27 An ECG should be obtained and echocardiography performed to assess anatomy, function, and hemodynamic status. Imaging should focus on ventricular function, valve function, presence of pulmonary hypertension, and assessment of prosthetic materials and patches. In select patients, cardiac MRI, cardiac CT, or cardiac catheterization may help the clinician to evaluate anatomy and hemodynamic status to further assess the risk of pregnancy for a specific patient. Access to experts in ACHD should be used for imaging and hemodynamic assessment as appropriate.

Genetic Risks

Both genetic and environmental factors play a role in the development of CHD. Both male and female patients with CHD have an increased risk of CHD in their offspring, and these genetic risks should be addressed before conception. Assessment of specific genetic risk requires obtaining a detailed family history of CHD and specific diagnoses for affected members. For isolated cases of CHD without a family history of CHD (eg, sporadic defects), the recurrence risk of CHD in the offspring is reported to be in the range of 3% to 8%, although the absolute risk varies with the specific type of defect and which parent is affected.28–32 In general, the risk of recurrence is higher if the mother is the affected parent. If >1 sibling is affected by CHD, the recurrence risk may be as high as 10%.33 Up to 20% of asymptomatic 1st-degree relatives of patients with left heart obstructive lesions (particularly bicuspid aortic valves) may have CHD.34

Approximately 18% of patients with CHD have a specific defined genetic abnormality. This includes chromosomal anomalies with either loss or gain of chromosomal material (eg, Down syndrome, Turner syndrome, Williams syndrome, and velocardiofacial syndrome), Mendelian syndromes attributable to single gene mutations or microdeletions (eg, Noonan syndrome, Holt-Oram syndrome, heterotaxy syndrome), or nonsyndromal single gene disorders. Patients with syndromic disorders may have other medical problems, including cognitive defects, endocrine, neurological, collagen vascular, or orthopedic disorders, which may also complicate their management.

For genetic defects such as Marfan syndrome or other autosomal dominant syndromes, the risk of fetal transmission is 50%, although variable penetrance of particular traits may

### Table 1. Types of Heart Disease Seen in Pregnant Women

<table>
<thead>
<tr>
<th>Author</th>
<th>Countries</th>
<th>N</th>
<th>Types of heart disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Siu, et al (CARPREG)3</td>
<td>Canada</td>
<td>599</td>
<td>74% CHD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>22% acquired heart disease</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>4% arrhythmias</td>
</tr>
<tr>
<td>Roos-Hesselink, et al (ROPAC registry)4</td>
<td>38 European countries</td>
<td>1321</td>
<td>66% CHD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>25% valve disease</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>7% cardiomyopathy</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>2% ischemic heart disease</td>
</tr>
<tr>
<td>Avila, et al6</td>
<td>Brazil</td>
<td>1000</td>
<td>56% RHD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>19% CHD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>9% Chagas disease</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>5% arrhythmias</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>4% cardiomyopathy</td>
</tr>
<tr>
<td>Liu4</td>
<td>China</td>
<td>1741</td>
<td>38% arrhythmias</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>30% CHD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>19% cardiomyopathy</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>9% RHD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>3% preeclampsia</td>
</tr>
</tbody>
</table>

CARPREG indicates Cardiac Disease in Pregnancy registry; CHD, congenital heart disease; RHD, rheumatic heart disease; and ROPAC, Registry On Pregnancy And Cardiac disease.
be seen. Conotruncal abnormalities (including truncus arteriosus, transposition complexes, double outlet right ventricle, and tetralogy of Fallot) may be associated with chromosomal abnormalities (trisomy 21, trisomy 13, and trisomy 18) or may be associated with a 22q11.2 microdeletion. Up to 10% to 15% of cases of tetralogy of Fallot or pulmonary atresia may be associated with 22q11.2 microdeletion. With a 22q11.2 deletion, there is a 50% risk of transmission to the offspring. Genetic counseling should be offered to all patients with CHD with referral for genetic testing in specific situations.

Environmental Risk Factors
Maternal environmental risk factors also play a role in the risk of congenital heart defects in the fetus. Maternal diabetes mellitus is associated with a 5-fold increase in risk of CHD, and strict control of blood sugar before conception and throughout pregnancy appears to decrease the risk of CHD. Obesity and cigarette smoking are both associated with increased risk of CHD. Thus, weight control and smoking cessation before conception will like decrease risk. Folic acid supplementation appears to also decrease the risk of CHD in offspring. All of these modifiable risk factors should be addressed with potential mothers before conception to minimize risk to the fetus.

Pregnancy Management
Optimally, patients with CHD should be evaluated at an ACHD center for preconceptual counseling and risk assessment, and mothers with moderate or complex defects or other high risk profiles should be followed throughout their pregnancy and delivery at an ACHD center or with continued involvement of an ACHD cardiologist. Cardiac status should be optimized before conception, including control of arrhythmias and surgical or percutaneous intervention for significant residual lesions (particularly obstructive lesions). A review of all medications should be performed with a view toward teratogenicity or other adverse fetal effects, and a plan should be created to manage anticoagulation if required. The frequency of clinic visits depends on maternal risk with women in the highest risk groups (based on maternal anatomic and physiological complexity) requiring more frequent follow up during the pregnancy. The initial visit should address the risk for pregnancy (if not determined before pregnancy) and to address a plan for pregnancy management, including a plan for delivery and postpartum monitoring. Fetal echocardiography is often recommended between weeks 18 to 22 to evaluate fetal cardiac anatomy. The mode of delivery should be individualized for each patient. Certain obstetric conditions may mandate Cesarean section, but vaginal delivery (either spontaneous or induced) is preferred in most cases because of a lower risk of complications. Cesarean is often recommended for patients who are anticoagulated, mothers with Marfan syndrome or other aortic aneurysm, and for critically ill patients, but the decision should be individualized. Cesarean may also be considered for patients with severe aortic stenosis or severe pulmonary hypertension. Anesthetic management should be planned and individualized for each patient. Heart rate and BP monitoring should be performed for all patients, and pulse oximetry is reasonable for mothers with chronic cyanosis. Continuous ECG monitoring may be indicated for some patients, and invasive monitoring of intraarterial pressure and central venous pressure may be indicated for certain high risk pregnancies. Routine antibiotics are not recommended for most mothers with heart disease but are recommended for patients with prosthetic heart valves, prior endocarditis, and patients with CHD with prosthetic materials or residual shunts.

Termination of Pregnancy
Termination should be considered in mothers with CHD whose pregnancy is determined to be high risk. Elective termination is felt to be the most safe when performed in the first trimester, before major hemodynamic shifts occur. Termination should be done in a hospital setting with precise anesthetic management and the ready availability of ACHD cardiology support and ICU level care. The availability of on-site cardiac surgery may be required in some cases. Dilatation and evacuation is the preferred method of termination within the first and second trimester.

Post-Pregnancy
Postpartum monitoring is important, and close follow up in the first 4 to 6 weeks postpartum is recommended for patients at moderate or high pregnancy risk, as cardiac decompensation may occur up to 6 weeks after delivery. Long-term follow up after pregnancy is important as well, because there is some evidence that deterioration in valvular function, ventricular function, and functional class may persist after delivery.

Review of Selected Defects
Left to Right Shunts
Uncomplicated left to right shunts (atrial septal defects [ASD], ventricular septal defects [VSD], and patent ductus arteriosus [PDA]) without evidence of pulmonary hypertension are usually well tolerated in pregnancy, with good outcomes. Secundum ASDs are most common, but sinus venous defects and partial atroventricular septal defects may also be seen. Patients with ASDs are at some risk for paradoxical embolism, but the reported incidence is quite low. An air filter on intravenous lines is reasonable. Small VSDs or PDAs without pulmonary hypertension or associated defects pose no increased risk for women during pregnancy. Mothers with large ASDs, VSDs, or PDA associated with pulmonary hypertension attributable to either large shunt flow or increased pulmonary vascular resistance do have increased risks in pregnancy and may develop arrhythmias, ventricular dysfunction, and worsening of their pulmonary hypertension, although specific guidelines defining unsafe limits for pregnancy have not been established. After delivery, reassessment of the shunt magnitude and referral for shunt closure if appropriate is warranted. Patients with previously repaired shunts with either surgical or device closure in the absence of ventricular dysfunction or elevated pulmonary vascular resistance tolerate pregnancy well. Patients with Eisenmenger syndrome attributable to a large shunt have a high risk of maternal mortality and morbidity, as well as decreased fetal survival, and...
this diagnosis is considered to be an absolute contraindication to pregnancy (see below).

Atrioventricular septal defects may be partial or complete defects and patients have often had surgery in childhood. For unrepaired and previously repaired atrioventricular septal defects, evaluation for the presence of residual lesions (atrial or ventricular level shunts, mitral regurgitation, left ventricular [LV] outflow tract obstruction, aortic regurgitation, and heart block) as well as pulmonary hypertension, ventricular dysfunction, and arrhythmias should be done. Although most mothers with previous successful atrioventricular septal defect repair without significant residual lesions tolerate pregnancy well, there is some increased risk. Worsening functional class, arrhythmias, and worsening of valvular regurgitation have all been described. Pregnancy is not advised in the setting of pulmonary hypertension or ventricular dysfunction.

Obstructive Lesions

Congenital Aortic Stenosis

CHD is the main cause of aortic stenosis (AS) in women of childbearing age. Mild to moderate AS is usually well tolerated in pregnancy, but more severe AS may be problematic. Women with severe AS may be asymptomatic. Prepregnancy evaluation of women with congenital AS should include echocardiography to assess the severity of AS and exercise testing to evaluate functional capacity, symptoms, blood pressure response to exercise, and arrhythmias. Symptomatic mothers, women with LV dysfunction, and women with an abnormal response to stress testing should be counseled against pregnancy and intervention on the valve should be performed before pregnancy.

In mothers with severe AS with stable severity of stenosis, absence of symptoms of stress testing, and normal BP response, pregnancy outcomes are adequate. Heart failure symptoms may develop in 10% and arrhythmias may occur in 2% to 35% of mothers during pregnancy, but maternal mortality is rare. Diuretics may be used and activities restricted for mothers who develop symptoms of heart failure. Pregnancy care should be coordinated between high-risk obstetrics and expert cardiology care. Patients require regular follow-up during pregnancy to evaluate for development of symptoms, and routine echocardiographic follow-up of valve function and LV function is also recommended during the pregnancy. Obstetric complications are increased in women with severe AS, including hypertensive-related disorders and preterm labor. Fetal complications may also occur in up to 25% of mothers with severe stenosis, including intrauterine growth retardation, preterm birth, and low birth weight. Delivery should be planned in advance, with vaginal delivery preferred for most patients. Patients with severe symptoms of heart failure despite medical management may be considered for percutaneous balloon valvuloplasty if valve anatomy is appropriate. Early delivery by Cesarean followed by urgent surgical valve replacement may be required for life-threatening symptoms if a percutaneous approach is not feasible.

Patients with bicuspid aortic valves may have an associated aortopathy making them prone to dilation and potentially dissection of the aortic root or ascending aorta. In women with bicuspid aortic valves, the aortic root and ascending aorta should be evaluated before pregnancy. Although data are limited, it is usually recommended that patients with aortic root dilatation associated with congenital bicuspid aortic valves should be advised to avoid pregnancy if their aortic root dimensions are \( \geq 4.5 \) cm.

Other Forms of LV Outflow Tract Obstruction

More complicated forms of LV outflow obstruction include subaortic stenosis and supravalvular stenosis. There may be multiple levels of obstruction in some patients. In patients with mild to moderate subaortic stenosis without previous repair, pregnancy is usually well tolerated although individual risk and pregnancy management should be discussed with ACHD experts. Supravalvular aortic stenosis refers to fixed obstruction above the level of the sinus of Valsalva (beyond the origin of the coronary arteries), sometimes extending into the ascending aorta. The coronary arteries may be abnormal, with ostial narrowing or complete obstruction, ectatic, or aneurysmal coronaries. Supravalvular AS is common in Williams syndrome, an autosomal dominant disorder. Pregnancy is contraindicated in symptomatic patients with subaortic or supravalvular aortic stenosis, particularly with significant LV outflow tract obstruction or coronary artery abnormalities.

Pulmonary Stenosis

Mothers with isolated pulmonary valve stenosis in the absence of right ventricle (RV) dysfunction usually tolerate pregnancy well, even with moderate to severe stenosis. Patients with severe stenosis, particularly with RV dysfunction, may be at some risk for symptoms of right heart failure. Percutaneous pulmonary valvuloplasty has been successfully performed during pregnancy in cases of severe symptomatic obstruction. A patent foramen ovale may be associated with pulmonary stenosis, and in the setting of severe pulmonary stenosis may result in right to left shunting at the atrial level. This may be unmasked by performing treadmill testing with pulse oximetry. Mothers with significant hypoxemia that is refractory to oxygen administration may have increased risk of fetal loss related to their cyanosis.

Coarctation of the Aorta

Coarctation of the aorta can occur as an isolated lesion or in association with other forms of CHD. There are few data on outcomes in mothers with unoperated coarctation, but in a single small series from the Mayo Clinic there was no maternal mortality. There is a small risk of aortic dissection in both unoperated and postoperative patients as well as an increased risk of preeclampsia and hypertension. Using a national database of hospitalizations, a study by Krieger et al demonstrated that women with coarctation of the aorta have a higher risk of hypertensive complications during pregnancy (including preexisting hypertension, pregnancy-induced hypertension, preeclampsia, and eclampsia), a higher risk of adverse cardiovascular outcomes (including heart failure, arrhythmia, stroke, and other embolic events), a higher risk of Cesarean section, increased hospital stays, and higher hospital costs. Aggressive blood pressure control in mothers with unoperated coarctation may result in decreased uterine
blood flow with compromise of fetal growth. The ESC guidelines suggest an increased risk of aortic rupture and rupture of intracerebral aneurysms in mothers with unrepaired coarctation and in mothers with previous repair who remained hypertensive.

Adult patients may have undergone previous intervention with either surgery or catheter intervention. Blood pressure should be checked in all 4 extremities in both unoperated and postoperative patients to assess for a gradient between upper and lower extremities. Arm blood pressures may be misleading if there is anomalous origin of a subclavian artery associated with the coarctation, or if the left subclavian artery was used to patch the area of repair or if the repair compromised blood flow to the left subclavian artery. Assessment for the severity of coarctation (or recurrent coarctation), aortic root dilatation, and the presence of other associated lesions should be undertaken before pregnancy, and elective coarctation repair before pregnancy is recommended for significant obstruction.

**Ebstein Anomaly**

Ebstein anomaly is a rare congenital anomaly with displacement of the tricuspid valve into the RV, resulting in varying degrees of tricuspid valve dysfunction (usually regurgitation) and impairment of RV function. Pulmonary stenosis, interatrial shunts, and electrophysiological abnormalities may also be associated. If an atrial level shunt is present, the patient may be cyanotic. Patients diagnosed with Ebstein anomaly in childhood frequently reach child bearing age, but are often asymptomatic. Adults presenting in adulthood may be asymptomatic or may have predominantly arrhythmias. Patients may have exercise intolerance, fatigue, right sided heart failure, arrhythmias, and cyanosis and are at risk for paradoxical embolism, arrhythmias, progressive RV dysfunction, and tricuspid regurgitation. In the absence of cyanosis, symptoms of heart failure, or significant arrhythmias, pregnancy appears to be well tolerated, both for un repaired and repaired patients, but is associated with an increased risk of premature birth and fetal loss. Mothers with significant cyanosis (oxygen saturation below 85%) or with symptomatic heart failure should be advised against pregnancy.

**Tetralogy of Fallot**

Tetralogy of Fallot is one of the most common forms of cyanotic CHD and most adults have had previous repair. In women with unrepaired tetralogy of Fallot, including those with previous palliative shunts, pregnancy is not advised because of poor maternal and fetal outcomes. After surgical repair, pregnancy is generally well tolerated in those women who have good functional capacity and no hemodynamically significant residual lesions. A significant percentage of patients develop pulmonary regurgitation after repair of tetralogy of Fallot, which may lead to RV enlargement and dysfunction, decreased exercise tolerance, and arrhythmias. Other residual lesions include residual RV outflow tract obstruction, residual VSD, aortic dilatation, and aortic regurgitation. With residual lesions, maternal complications may occur, including arrhythmias and heart failure, and there is an increased risk of fetal loss and intrauterine growth retardation.

Increased risk of obstetric complications was reported in 1 series for mothers with residual pulmonary regurgitation. Elective pulmonary valve replacement is recommended before pregnancy for severe regurgitation with RV enlargement or dysfunction.

**D-Transposition of the Great Arteries**

D-Transposition of the great arteries is the most common form of cyanotic CHD seen in the neonatal period. Associated lesions may occur, including VSD, subpulmonary outflow obstruction, and coarctation of the aorta. Most adults with D-Transposition of the great arteriesshave undergone either an atrial baffle procedure or an arterial switch repair in childhood. The atrial switch creates a complex atrial baffle system to redirect systemic venous return to the anatomic RV and pulmonary venous return to the RV. The anatomic RV serves as the systemic ventricle, whereas the anatomic LV serves as the venous ventricle. Maternal and fetal outcomes have been described in this group of patients in several small series. Arhythmias, worsening systemic ventricular function, heart failure, obstruction of the atrial baffle tissues, pulmonary hypertension, stroke, and death have been reported. Deterioration of the systemic RV resulting in clinical heart failure is the major concern and may be irreversible. Comprehensive cardiac evaluation is essential before pregnancy to understand their risk for pregnancy and to manage them during pregnancy.

The arterial switch repair is now the preferred procedure for D-transposition of the great arteries and is associated with excellent quality of life, arrhythmia-free survival, and long-term survival. Comprehensive cardiac evaluation is essential before pregnancy for these patients. Development of LV dysfunction, RV outflow tract obstruction, abnormal coronary blood flow, chronotropic incompetence, neoaortic and pulmonary regurgitation, and dilatation of the neoaoartc root may occur after the arterial switch and may cause complications in pregnancy. There are limited data on pregnancy outcomes after the arterial switch, but outcomes appear to be good in those women with a well functioning repair.

**Congenitally Corrected Transposition**

Congenitally corrected transposition is a relatively rare form of CHD with 2 anatomic abnormalities resulting in physiological correction of the circulation. The RV serves as the systemic ventricle, and many patients present with ventricular dysfunction and heart failure in midadulthood. Patients with associated defects such as VSD or pulmonary stenosis may have had previous surgery. Because of abnormal positioning of the AV node and elongation of the AV bundle, patients are at increased risk of complete heart block (occurs in ~2% of patients per year) as well as reentrant arrhythmias. Prepregnancy assessment with a particular focus on the systemic ventricular function and systemic AV valve regurgitation is imperative. In 2 series, there was an increased rate of fetal loss and cardiovascular morbidity (including worsening systemic AV valve regurgitation, heart failure, endocarditis, worsening cyanosis, and stroke). Systemic ventricular dysfunction, greater than mild systemic AV valve regurgitation, and cyanosis predict patients at risk for maternal complications.
**Chronic Cyanosis**

Chronic cyanosis occurs with uncorrected forms of CHD, with previous palliative shunting, and in some patients who have developed complications from their original repair. Precise delineation of the patient’s anatomy and the cause of their cyanosis before conception is recommended whenever possible. The risk of pregnancy will depend on the cause of the cyanosis, and catheter or surgical intervention to relieve cyanosis before conception may improve fetal outcome and decrease maternal risks. In adults with uncorrected congenital heart defects who have survived to adulthood without surgical intervention, pregnancy can be contemplated with some increased risk. One series assessed pregnancy outcomes in women with cyanotic CHD and found that fetal survival was 92% with maternal arterial saturations at ≥90% but was only 12% with arterial saturations <85%.

Maternal outcome is dependent on the underlying cardiac defect. Cyanotic mothers are at risk for paradoxical emboli resulting from right to left shunting, and meticulous management of intravenous lines and use of air filters is usually advised.

**Pulmonary Hypertension**

Pulmonary hypertension occurs in 4% to 10% of all patients with CHD, often as a result of long-term left to right shunting and excessive pulmonary circulation. Pulmonary hypertension may occur with simple or complex congenital defects, in patients with un repaired shunts, and in some patients late after surgical repair. In general, pregnancy is not recommended in women with increased pulmonary vascular resistance, although specific guidelines defining unsafe limits have not been established. In some patients with large left to right shunts at birth, the pulmonary pressure equals the systemic pressures, resulting in shunt reversal and cyanosis (known as the Eisenmenger syndrome), the most extreme form of pulmonary hypertension associated with CHD. Compared with idiopathic pulmonary arterial hypertension, pulmonary hypertension associated with CHD may have a better prognosis with longer survival, although recent registry data suggest that this may not be the case. Pregnancy in mothers with Eisenmenger syndrome is contraindicated because of high maternal and fetal mortality. Maternal mortality rates as high as 50% have been noted in some reports, although lower rates have been reported in some series. Common modes of death during pregnancy are pulmonary hypertensive crisis, in situ pulmonary thrombosis, and advanced right heart failure. Death occurs most commonly in the third trimester or first few weeks after successful delivery, so that continued monitoring in the postpartum period is required. Fetal loss is also significantly increased with spontaneous abortion occurring in up to 40% to 50%, and termination of pregnancy is often recommended early in the pregnancy and should be undertaken in a facility with expertise in the management of both CHD and pulmonary hypertension. If a mother opts to continue the pregnancy, a similar team of specialists is required to manage the patient throughout the pregnancy, particularly at the time of delivery and postpartum.

A variety of therapeutic agents are now available for treatment of pulmonary hypertension, but not all are appropriate for use in pregnancy. Endothelin receptor antagonists are teratogenic, and strict measures should be undertaken to avoid conception. If pregnancy does occur, these agents should be stopped and switch to alternative agents. Prostacyclin analogs and phosphodiesterase inhibitors may be used during pregnancy and may help to improve outcomes. Meticulous planning of delivery and postpartum care is needed to optimize outcomes.

**The Fontan Operation**

Patients born with complex defects that cannot be repaired to recreate a 2-ventricular circulation commonly undergo a series of operations to create a Fontan connection. The Fontan procedure is a palliative procedure that relieves cyanosis but does not restore normal cardiac anatomy. After the Fontan operation, systemic venous blood returns directly to the pulmonary arteries and the functional single ventricle supplies both the systemic and pulmonary circulation in series, rather than two separate circulations. Patients frequently do well for many years after this procedure, but there are many complications that occur over time, including arrhythmias, ventricular dysfunction, development of abnormal venous collaterals, protein losing enteropathy, and chronic liver congestion that can eventually lead to liver fibrosis, and in some cases cirrhosis.

Survival after the Fontan operation was reported at 80% at 1 year, 78% at 5 years, and 71% at 10 years in 1 report and 94% at 5 years, 90% at 10 years, 98% at 15 years, and 83% at 20 years in a more recent report. Late deaths are most often attributable to arrhythmia, thromboembolism, or heart failure. Young women of childbearing age after a Fontan procedure often are interested in pregnancy, particularly if they have not experienced complications from their Fontan circulation. Fertility may be an issue and pregnancy complications are frequent in mothers with a Fontan circulation, particularly if they have had previous cardiac issues such as arrhythmias. Even in mothers with a good Fontan, miscarriage rates and prematurity rates are high, low fetal birth weight is common, and maternal complications such as arrhythmias, heart failure, and embolic complications may occur. Pregnancy in a woman after a Fontan operation must
be undertaken with great deliberation, and the risks to both mother and fetus should be understood. These patients should be followed during the pregnancy (and throughout their life) in a center with expertise in ACHD patients.

**Marfan Syndrome**

Marfan syndrome is an autosomal dominant genetic disorder of connective tissue caused by mutations in the FBN1 gene. The cardinal features of Marfan syndrome include the cardiovascular system, ocular system, and the skeletal system but aortic complications account for most of the mortality for these patients. An increased risk of aortic dissection or rupture has been well described in women with an aortic root diameter >40 to 45 cm and for those with previous aortic root surgery.\(^{10,11}\) This risk of aortic complications increases throughout the pregnancy, and complications may also occur after delivery. The risk is low for aortic diameters <4.0 cm but pregnancy is absolutely contraindicated in women with an aortic root diameter >4.4 cm and surgical repair of the aortic root is recommended before pregnancy for aortic root diameters >4.0 to 4.5 cm.\(^{21,47,111}\) Other congenital aortopathies such as Loeys-Dietz have been identified which also place patients at increased risk of aortic complications, requiring careful management. The risk for dissection may vary according to the specific type of aortopathy, and special expertise in these disorders is required to counsel women about their pregnancy risk and outcomes.

**A General Approach to Risk Stratification**

Several registries provide information that can be used to counsel women with CHD about their pregnancy risks. The CARPREG registry was the first to identify specific risk factors for maternal complications of pregnancy, and several other registries have identified additional risk factors that contribute to increased risk.\(^{3,6,14,29,77,112}\) Two studies have also identified chronotropic incompetence on exercise testing as a risk factor.\(^{26,27}\) Table 2 lists the risk factors that have been associated with maternal complications in pregnancy. These risk factors appear to be additive, with increased pregnancy risk associated with multiple risk factors. Risk factors for poor fetal outcomes include poor maternal functional class, systemic ventricular dysfunction, left sided obstructive lesions, maternal cyanosis, anticoagulation, multiple gestation pregnancy, smoking, maternal age <20 or >35years, and chronotropic incompetence. The ZAHARA investigators, a consortium representing eight Dutch medical centers, have revised the CARPREG registry risk score, incorporating additional risk factors and using an exponential weight to adjust the risk score to result in a better predictive value.\(^{112}\) However, this risk score has not been validated prospectively.\(^{112}\) Finally, the ESC Guidelines on cardiovascular disease and pregnancy recommend using a modified World Health Association (WHO) classification for mothers with cardiac disease.\(^{(Table 3)}\) For patients in WHO class I, there is no increased risk of maternal mortality and no or only a mild increase risk in morbidity with pregnancy. For women in WHO class IV, pregnancy is absolutely contraindicated. The ESC further divides women at some risk into class II, II to III, and III depending on maternal risk. Mothers in WHO class III require extensive counseling before pregnancy and need intensive cardiac and obstetric follow-up throughout pregnancy.

**Summary**

The population of adults with CHD continues to expand, and thus the number of women with CHD who contemplate

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**Table 3. World Health Organization Proposed Classification of Pregnancy Risk**

<table>
<thead>
<tr>
<th>Modified WHO Class</th>
<th>Definition</th>
<th>Types of CHD</th>
</tr>
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<tbody>
<tr>
<td>WHO I</td>
<td>No increased risk of maternal mortality</td>
<td>Mild pulmonary stenosis</td>
</tr>
<tr>
<td></td>
<td>No or mild increased risk of maternal morbidity</td>
<td>Small, uncomplicated PDA</td>
</tr>
<tr>
<td>WHO II</td>
<td>Small increase in risk of maternal mortality, or Moderate increase in risk of maternal morbidity</td>
<td>Unoperated ASD or VSD</td>
</tr>
<tr>
<td>WHO II–III</td>
<td>Moderate increase in risk of maternal morbidity</td>
<td>Repaired tetralogy of Fallot</td>
</tr>
<tr>
<td>WHO III</td>
<td>Significantly increased risk of maternal mortality or severe morbidity</td>
<td>Systemic right ventricle</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Fontan circulation</td>
</tr>
<tr>
<td>WHO IV</td>
<td>Extremely high risk of maternal mortality or severe morbidity. Pregnancy is contraindicated.</td>
<td>Pulmonary hypertension of any cause</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Severe systemic ventricular dysfunction (EF &lt;30%, NYHA class III–IV)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Severe symptomatic AS or MS</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Native severe coarctation</td>
</tr>
</tbody>
</table>

Adapted from Regitz-Zagrosek et al\(^{21}\) with permission of the publisher. Copyright ©2011, Oxford University Press (UK), European Society of Cardiology. AS indicates aortic stenosis; ASD, atrial septal defect; CHD, congenital heart disease; EF, ejection fraction; LV, left ventricle; MS, mitral stenosis; NYHA, New York Heart Association; PDA, patent ductus arteriosus; PAPVR, partial anomalous pulmonary venous return; VSD, ventricular septal defect; and WHO, World Health Organization.
pregnancy or become pregnant is also growing. Mothers with low-risk defects can be managed by general cardiologist, whereas those with more complex defects should be managed by or with the assistance of ACHD cardiologists. It is important to acknowledge that all patients with CHD may have unique anatomy or physiology, despite their classification as having a simple, moderate, or complex defect. As such, clinicians evaluating these patients should have adequate knowledge and expertise when assessing patient’s risk for pregnancy, when performing imaging or hemodynamic studies, and when managing these patients during pregnancy. The American Board of Medical Specialties has recently recognized ACHD as a subspecialty of cardiovascular disease to treat the specialized needs of these patients in adulthood. ACHD experts can provide expertise in the management of specific defects or lesions, imaging techniques, pre-pregnancy risk assessment, and can manage these patients or comanage them with other medical providers during their pregnancy. Because many of these ACHD patients are lost to follow-up in adulthood, pregnancy represents a time when these patients seek medical care (and for some, represents a time of vulnerability and increased risk). This represents an opportunity to establish or reestablish care with ACHD specialists and to reestablish continuing long-term care for their CHD. Pregnancy also provides an opportunity to create partnerships between primary care physicians, adult cardiologists, and ACHD specialists to provide optimal care for these women throughout their lives.

Disclosures

None.

References


KEY WORDS: congenital heart defect ■ congenital heart disease ■ pregnancy
Cardiovascular Management in Pregnancy: Congenital Heart Disease
M. Elizabeth Brickner

Circulation. 2014;130:273-282
doi: 10.1161/CIRCULATIONAHA.113.002105

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