Heart Diseases During Pregnancy

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Improvements in health care services in recent decades have permitted more frequent identification of pregnant women with congenital and acquired heart disease. Normal pregnancy and the peripartum period are associated with considerable cardiocirculatory changes, which are usually well tolerated by the mother. However, the increased cardiac demands imposed by those changes may at times unmask cardiac disease in presumably normal women or increase morbidity and mortality in women with established heart disease. A recognition and understanding of these changes is essential for providing optimal care to obstetrical patients with cardiac disorders. This article offers a brief review of those cardiocirculatory changes during pregnancy and of the main features of various congenital and valvular heart disorders frequently referred for advice to our cardiology practice, along with recommendations for their management and follow up.

Key words: Heart disease, Pregnancy, Congenital heart disease, Valvular heart disease, Arrhythmias

Normal pregnancy and the peripartum period are associated with significant hemodynamic changes, usually well tolerated by the mother, but the increased cardiac demands imposed by them may at times exacerbate minor preexisting symptoms, unmask cardiac disease in presumably normal women and increase morbidity and mortality in women with known heart disease. The recognition and understanding of those changes and the identification of different congenital and valvular cardiac disorders affected by them during pregnancy, is essential to provide optimal management to these patients.

Cardiac Physiologic Changes in Pregnancy and Periods of Higher Risk

Several hemodynamic and circulatory changes occur during pregnancy (Table 1), although the mechanisms responsible for those changes are incompletely understood. One of the most remarkable changes is an average increase of around 40 percent in cardiac output. That increase starts around the fifth week of gestation and peaks around weeks 15 to 20 and is largely related to an augmentation of the stroke volume (1). Increase in cardiac output during the third trimester, however mostly occurs due to a rise in heart rate. Changes in body position can also cause variations in cardiac output with higher levels occurring while the woman is in a lateral position and lower levels when supine. Compression of the inferior vena cava by the gravid uterus, while the woman is in the supine position is the main cause of those positional changes. The resulting diminution in venous return and hypotension, may negatively affect patients with volume dependent cardiac lesions like aortic valvular stenosis or left ventricular systolic dysfunction. An increase in heart rate is also a frequent cardiovascular change observed in pregnancy and which peaks in the third trimester. The

<table>
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<th>Table 1. Reported Hemodynamic Changes During Pregnancy</th>
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<td>First Trimester</td>
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<td>Stroke volume</td>
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<td>Diastolic blood Pressure</td>
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Legend: ↑ slight increase, ↑↑ moderate increase, ↑↑↑ marked increase, ↓ slight reduction, ↓↓ moderate reduction, ↓↓↓ marked reduction, unchanged, or decrease

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average rise in heart rate is of 10 to 20 beats per minute, however the normal total heart rate rarely exceeds 100 beats per minute (2). Blood volume also considerably increases during pregnancy, starting as early as the 6th week and rising rapidly until midpregnancy when the rise continues but at slower rate. The degree of maximal circulating volume expansion varies significantly between individuals but averages to around 50 percent of the prepregnant value. As the increase in circulating blood volume is greater than the increase in red cell mass, this is reflected in a lower hemoglobin concentration or the so-called “anemia of pregnancy” (3). Another physiologic cardiovascular change of importance during pregnancy is a decrease of approximately 25% in systemic vascular resistance. This change may manifest with a decrease in systemic arterial pressure around midpregnancy and by widening of the pulse pressure observed in pregnant women. The decline in peripheral resistance may positively affect patients with regurgitant valvar lesions, such as aortic or mitral valvar insufficiency, who may profit from afterload reduction (4).

In parallel with changes in cardiac physiology, there are three periods of particular high risk for cardiac decompensation during pregnancy. The first period occurs at the end of the second trimester (24 to 28 weeks gestation), when blood volume and cardiac output reach peak values. A second period of high risk occurs in association to the work of labor, when from one to two units of blood may be auto-transfused from the uteroplacental into the systemic circulation with each uterine contraction, with subsequent return of that same amount of blood to the uteroplacental circulation with each contraction. (1). Pregnant women with cardiac compromise may be unable to tolerate such rapid shifts in circulatory volume. The postpartum period also increases the risk for decompensation in women with heart disease due to the overload associated to varying fluid shifts, and related to the involution of the uterus (with loss of the low resistance circulatory component of the placenta) and the increase in preload after release of the compression of the inferior vena cava by the gravid uterus. Patients particularly sensitive to shifts in blood volume as those with severe pulmonary hypertension and Eisenmenger syndrome may experience considerable morbidity and mortality during this period.

Cardiac Evaluation During Pregnancy

Frequent Symptoms

The diagnosis of heart disease, particularly congenital diseases, may be difficult at anytime but more so during pregnancy. One of the reasons for that difficulty is that symptoms frequently observed in patients with heart disease, like fatigue, shortness of breath, orthopnea, palpitations, chest pain or discomfort, dizziness and leg edema, are also commonly reported during normal pregnancy (5). Palpitations are one of the most frequent symptoms referred by pregnant women. These have been related to a shift of the heart closer to the anterior chest wall, a faster resting heart rate and increased cardiac contractility associated with pregnancy. Ectopic atrial or ventricular contractions may occur in many normal pregnant women and have been linked to increased atrial and ventricular stretch due to the augmented circulatory volume. Increased body awareness by many women in concert with increased exposure to health care providers, can contribute to increased reporting of palpitations during the gestational period. However, palpitations characterized as fast, either regular or irregular and that last more than several minutes and that are accompanied with lightheadedness, presyncope or syncope are most likely related with a significant tachyarrhythmia and demand further investigation (6).

Normal and Abnormal Physical Findings

Due to the substantial cardiocirculatory changes associated with pregnancy, normal physical findings during gestation may be misinterpreted as abnormal. Frequent findings in normal pregnant women include, an increase in heart rate (which peaks during the third trimester and has a mean rise of 10 to 20 beats per minute), increased jugular venous pulsations, lateral displacement of the left ventricular impulse, a physiologic third heart sound and a basal, early peaking ejection systolic murmur. Peripheral edema and venous varicosities are also commonly observed. However, physical findings such as a fourth heart sound, a loud (3/6 intensity) systolic murmur or diastolic murmurs (aside from the occasional physiologic diastolic murmur heard at the left sternal border and usually generated by increased diastolic flow through the internal mammary arteries of the engorged breasts of pregnant women ["mammary souffle"], are considered abnormal and do not occur during normal pregnancy and are thus indicative of a need for further careful cardiac evaluation (7).

Assessment of the Risk of Pregnancy in Patients with Heart Disease

The functional classification of the New York Heart Association (NYHA) is a useful tool for the assessment of the risk of pregnancy in women with heart disease. Women with Class I and Class II functional classification usually tolerate pregnancy very well, with a mortality of less than 1%. The risk increases to around 7% in Class III
and IV patients. These patients must be made aware of the potential for morbidity and mortality prior to becoming pregnant. In general, fixed obstructive cardiac disorders or those associated with pulmonary hypertension are poorly tolerated during pregnancy because of inability for increasing cardiac output. In contrast, valvular regurgitant lesions are relatively well tolerated due to the decrease in systemic vascular resistance and afterload. In presence of cardiovascular lesions placing a mother and the fetus at extremely high risk such as pulmonary hypertension, dilated cardiomyopathy with congestive heart failure, Marfan’s syndrome with dilated aortic root and cyanotic congenital heart disease the advise of avoidance or interruption of pregnancy is considered appropriate (8).

**Congenital Heart Diseases**

Congenital heart diseases represent the most common form of structural heart disease affecting women of childbearing age. Improvement in medical care has permitted that women with these disorders reach such age and consider becoming pregnant. The management of these patients must start prior to conception. Patients and their relatives must understand the risks for both the mother and the fetus and the risk of cardiac malformations for the latter. Fetal echocardiography is to be routinely employed in pregnant women with these disorders to further assess that possibility. Generally, the survival of the mother will depend on the type of disorder, whether the lesion has been corrected or requires correction, the presence of cyanosis and the functional capacity. Pregnancy is considered contraindicated in women with severe cyanotic congenital heart disease. These women have high risk for fetal loss and if reach term usually deliver low-birth-weight infants. It is advisable that when recognized and feasible, congenital heart disease be corrected prior to pregnancy. Most women with congenital heart disorders should have vaginal deliveries. Cesarean sections should only be performed for obstetrical reasons or marked hemodynamic compromise. Hemodynamic monitoring may be required in patients with markedly diminished functional capacity, severe pulmonary hypertension and severe cyanotic disorders (9).

**Atrial septal defect (ASD)**

The ostium secundum ASD is the most common congenital heart disorder reported during pregnancy. It may at times be associated with concurrent mitral valve prolapse. The symptoms and signs of an ASD can be minimal and the lesion may be unrecognized prior to pregnancy. This disorder is generally well tolerated, with no significant effect on morbidity and mortality. However uncorrected ASD is associated with a small increase in the risk for paradoxical embolism. Ostium primum defects are equally well tolerated during pregnancy, unless associated with other significant cardiovascular abnormalities (11).

**Ventricular septal defect (VSD)**

As most ventricular septal defects spontaneously close during childhood, they are not frequently encountered during pregnancy. When present the lesion is usually well tolerated in the absence of associated pulmonary hypertension (12).

**Patent ductus arteriosus (PDA)**

Uncomplicated patent ductus arteriosus is also usually well tolerated during pregnancy in women with small to moderate shunts and normal pulmonary artery systolic pressure.

**Coarctation of the aorta**

Whenever possible surgical correction is advised prior to conception to reduce the possibility of aortic dissection, aortic rupture or death, as the associated maternal mortality rate reported with this lesion is from 3 to 8% (13). A concomitant bicuspid aortic valve is very common. Correction of the coarctation may at times be required during pregnancy if there is severe uncontrollable hypertension or heart failure (14).

**Cyanotic Cardiac Lesions (Right to Left Shunts)**

**Eisenmenger syndrome**

Eisenmenger syndrome may occur with long standing uncorrected congenital cardiac lesions involving large left to right shunts, such as ASD, VSD or PDA, whenever pulmonary vascular resistance exceeds systemic vascular resistance (15). The elevation of the pulmonary vascular resistance causes right to left reversal of the shunt and cyanosis.

Pregnancy is not advised in these patients and therapeutic abortion may have to be considered. Should pregnancy occur, close follow up and early hospitalization around the time of delivery is highly recommended. Vaginal
delivery is preferable, as cesarean section has been associated with higher mortality (16).

**Tetralogy of Fallot (TOF)**
Tetralogy of Fallot is the most common cyanotic cardiac disorder seen during pregnancy. In women with uncorrected lesions the maternal mortality is high and fetal loss can exceed 50%. Signs of poor prognosis include, a maternal hematocrit above 60%, oxygen saturation below 80%, right ventricular hypertension and syncopal episodes (17). Vaginal delivery is preferred in most patients with TOF.

Surgically corrected lesions are associated with similar maternal mortality like that of women without heart disease. Women with pulmonary hypertension or significant LV dysfunction should be counseled about their cardiovascular risk and advised to avoid pregnancy.

**Complex Congenital Heart Diseases**
The maternal and fetal morbidity and mortality with these disorders is very high when the lesion results in maternal cyanosis.

**Transposition of the Great Vessels (TGA)**
The available data reported in women with d-transposition of the great vessels has shown poor maternal and fetal outcome, although partial or complete surgical correction prior to pregnancy has been associated with improved results (18,19).

In women with congenitally corrected transposition of the great arteries (I-transposition; I-TGA) pregnancy can be well tolerated if not complicated by cyanosis, ventricular dysfunction or heart block. This disorder is characterized by atrioventricular and ventriculo-great arterial discordance, and may occur as an isolated anomaly or associated to other congenital disorders like VSD, pulmonary stenosis, anomalies of the atrioventricular valves (Ebstein’s-like malformation), complete heart block, dextrocardia and ASD. However, women in Class III or IV NYHA functional classification, severe systemic ventricular dysfunction (ejection fraction <40%) or significant systemic atrioventricular valve regurgitation should be advised to avoid pregnancy (20,21).

**Other Disorders**

**Pulmonary stenosis**
Pulmonary stenosis is most commonly due to congenital obstruction at the valvular level. Women with isolated pulmonary stenosis frequently tolerate pregnancy well, even if severe but intravascular volume depletion should be avoided (22). If severe symptoms occur, such as syncope, uncontrolled dyspnea and chest pain, percutaneous balloon valvuloplasty may be performed after the first 12 weeks of gestation.

**Marfan syndrome**
Women with Marfan syndrome should be advised to avoid pregnancy in view of the unpredictable risk of death from aortic rupture or dissection (23). This is particularly high if the aortic root diameter exceeds 40 mm by echocardiography (24). If the patient elects to continue her pregnancy, physical activity should be limited and hypertension prevented. Beta-adrenergic blockade have been reported to decrease the progression of aortic dilatation. Serial echocardiography is advised for monitoring aortic root dilatation and termination of pregnancy. Aortic repair is counseled if progressive dilatation occurs. This is one of the few instances in which delivery by cesarean section is recommended, to avoid the hemodynamic stress of labor.

**Hypertrophic cardiomyopathy**
In general, most asymptomatic women with hypertrophic cardiomyopathy (HCM) tolerate pregnancy well and undergo successful vaginal delivery (25). However, patients with heart failure and severe symptomatic restrictive physiology are unlikely to tolerate the hemodynamic changes of pregnancy and they should be advised against it (26). The ability to tolerate pregnancy in patients with HCM tends to correlate closely with preconception NYHA functional status. Beta-adrenergic blocking agents may be of benefit for relieving dyspnea and angina by improving coronary perfusion and allowing a more effective ventricular filling time. Rate control and prompt reversion to sinus rhythm are required in women who develop atrial fibrillation, as this is poorly tolerated due to the effects of the tachycardia and loss of atrial transport. Careful fluid management is very important to maintain ventricular filling but pulmonary congestion must be avoided. In an uncomplicated pregnancy, the main risk to the fetus is the 50% recurrence risk. As HCM is rarely diagnosed in early life, screening of the offspring is recommended until adulthood.

**Valvular Disorders**

**Mitral Stenosis**
Mitral stenosis is the most common valvular disorder seen during pregnancy (27). It is almost always associated to rheumatic heart disease. The increase in cardiac output, heart rate and fluid retention during pregnancy may elevate the pressure gradient across the stenosis and elicit or worsen congestive symptoms. Maternal and fetal
complications are higher with higher NYHA functional classifications. Aggravation of symptoms usually starts around the 20th week of gestation.

In women who are considering pregnancy and who have symptomatic moderate to severe stenosis and a valve area less than 1.0 cm², balloon dilation or valve surgery should be performed before conception. Pregnant women with mild stenosis (a mitral valve area more than 1.5 cm²) may have conservative medical management.

If mitral stenosis is first recognized during pregnancy and the patient becomes symptomatic, initial standard medical therapy is appropriate (28). If this does not control symptoms, balloon valvuloplasty can be performed (with appropriate radiation shielding to the fetus) after the second trimester. Surgical commissurotomy or valve replacement has been performed, but with a fetal loss exceeding 30% (29).

Atrial fibrillation with rapid ventricular response is of particular concern in patients with mitral stenosis, as it may reduce diastolic filling and lead to pulmonary edema. Emergency treatment may include IV verapamil or cardioversion; beta-blockers may also be useful.

The vaginal route with epidural anesthesia is the usual approach for delivery. Use of assisted-delivery devices is encouraged to eliminate the need for pushing during the second stage of delivery (30). Cesarean section may be performed if there are obstetrical indications.

**Mitrail regurgitation**

Is more frequently caused by rheumatic heart disease or myxomatous degeneration of the valve. Mitral valve prolapse is another common cause, which if isolated is not usually associated to further complications. Mitral regurgitation is generally well tolerated during pregnancy due to the decrease in systemic vascular resistance (31). Afterload reduction therapy may be of benefit in symptomatic patients, but ACE inhibitors must be avoided due to teratogenic effects. Nitrates and hydralazine may be utilized as alternative agents. In patients with severe symptoms not responding to medical therapy and a high incidence of fetal loss, valve repair could be considered.

**Aortic stenosis**

Aortic stenosis is uncommonly encountered in pregnancy. Its most common cause in childbearing age women is a congenital bicuspid valve (32). If severe stenosis is recognized prior to conception, balloon valvuloplasty or a surgical commissurotomy should be considered.

Mild to moderate aortic stenosis with preserved left ventricular systolic function (an ejection fraction $>50\%$) is usually well tolerated during pregnancy. Severe aortic stenosis with an aortic valve area of less than 1.0 cm² and a mean systolic gradient above 40 mm Hg significantly increases the risk. In patients with severe symptoms balloon valvuloplasty or aortic valve surgery can be performed, but may be associated with increased fetal loss (33).

Vaginal delivery is the preferred approach with employment of assisted-delivery devices during the second stage to eliminate the need for pushing. Cesarean section should be considered in the presence of obstetrical indications.

**Aortic regurgitation**

Its most common causes are rheumatic heart disease, infectious endocarditis, connective tissue diseases, bicuspid aortic valves and dilation of the aortic root (34). Marfan syndrome should be considered in presence of a dilated aortic root or dissection. It is usually well tolerated during pregnancy due to the decrease in vascular resistance. Should heart failure occur, recommended treatment will require employment of diuretics and afterload reduction with vasodilators, such as hydralazine and nifedipine, but ACE inhibitors are to be avoided.

**Tricuspid valvular disease**

Lesions of this valve are uncommonly seen during pregnancy. An increase in the incidence of tricuspid regurgitation secondary to right-sided endocarditis due to intravenous drug abuse is being observed (35). Tricuspid stenosis is very rare, but when present, intravascular volume depletion should be avoided.

**Infectious Endocarditis Prophylaxis**

The existing guidelines for management of heart disease in pregnancy are included in the 1998 American College of Cardiology/American Heart Association (ACC/AHA) guidelines on valvular disease. Those guidelines do not recommend routine antibiotic prophylaxis for women undergoing uncomplicated vaginal delivery or cesarean section. However, if vaginal infection is present, bacterial endocarditis prophylaxis should be started promptly. In addition, antibiotic prophylaxis seems appropriate for women in high-risk categories like those with prosthetic valves or prior history of endocarditis (36).

**Cardiac Arrhythmias**

Premature atrial and/or ventricular complexes frequently occur during pregnancy, but are not usually associated with adverse maternal or fetal outcomes and often do not require antiarrhythmic therapy. Antiarrhythmic therapy should only be initiated in symptomatic or hemodynamically unstable patients. Atrial fibrillation and
atrial flutter are infrequently seen during pregnancy and rate control can be achieved with digoxin and beta-blockers. Electrical cardioversion can be safely performed during any stage of pregnancy in hemodynamically important or life threatening situations (37).

Resumen

Se presenta una revisión de hallazgos sobresalientes de enfermedades cardiovasculares que pueden ocurrir en asociación al embarazo. Se describen particularmente, varias enfermedades congénitas, tanto anormales como cianóticas y desórdenes que afectan las válvulas mitral, aórtica y tricuspidea.

References


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