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Peripartum Consideration for Women with Cardiac Disease

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Abstract

Purpose of review: The aim of this review of cardiac disease in pregnancy is to delineate current best practices and highlight emerging themes in the literature.

Recent findings: Cardiovascular disease is the leading cause of death among pregnant women in the United States. Many clinicians and institutions have developed care pathways to approach care in these high-risk patients including highly coordinated multidisciplinary teams. The diagnosis of pulmonary hypertension is the greatest risk factor for an adverse event in pregnant women. Vaginal delivery, with good neuraxial anesthesia, is usually the preferred mode of delivery in women with cardiac disease, although the rate of cesarean delivery is higher among women with heart disease.

Summary: The leading cause of morbidity and mortality in pregnant women is cardiac disease. Preconception counseling is useful for optimizing patients for pregnancy and setting appropriate expectations about care and outcomes. Ensuring that women are cared for in centers with appropriate multidisciplinary resources is key for improving outcomes for cardio-obstetric patients.

Keywords

Cardiovascular disease; Pregnancy; Obstetric anesthesia

Introduction

Cardiovascular disease is the leading cause of death among pregnant women in the United States. The prevalence of cardiac disease among pregnant women is expected to increase

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Conflicts of interest
None

with improved survival among women with congenital heart disease, delay in timing of motherhood, and the associated comorbidities with advancing maternal age.[1] It is difficult to conduct randomized controlled trials in cardiovascular disease and pregnancy; however, there are new observational studies and pertinent clinical reviews published on this topic in the year 2020. The aim of this review is to describe the major themes in the literature and highlight current clinical practice and research.

New Statements/Care pathways

The pregnant cardiac patient has become a national focus as cardiac conditions are a leading cause of maternal morbidity and mortality. In 2020, investigators at Columbia University Irving Medical Center conducted a secondary analysis of a four-year observational cohort study of the Eunice Kennedy Shriver National Institute of Child Health and Human Development (NICHD) Maternal-Fetal Medicine Units (MFMU) performed at 19 academic medical centers between 1999 and 2002. They assessed baseline factors associated with critical care interventions during delivery admission. 73,257 women were included in this secondary analysis of a multicenter observational registry of pregnancy after prior uterine surgery and primary cesarean delivery. Heart disease [aOR = 10.05, CI = 6.97 – 14.49], renal disease [aOR = 2.78, CI = 1.49 – 5.18], connective tissue disease [aOR = 3.27, CI = 1.52 – 6.99], and hypertensive disorders of pregnancy [aOR = 2.04, CI = 1.31 – 3.17] were associated with the greatest odds of critical care intervention [$p < .01$].[2] The authors concluded providers should create a multidisciplinary approach to assess maternal risk and determine appropriate level of care.

Recent consensus statements address maternal cardiovascular disease, describe the prevalence of cardiac disease in pregnancy and provide guidance on identifying and modifying risk factors and diagnosing and managing cardiac disease in the pregnant women. [3, 4] In 2017, California Maternal Quality Care Collaborative (CMQCC) published the Cardiovascular Disease in Pregnancy and Postpartum Toolkit; in 2019, American College of Obstetrics and Gynecology published practice bulletin No. 212; and in 2020, the American Heart Association published a statement about cardiovascular considerations in pregnant patients.[3–5]

For example, as more women born with congenital heart disease approach child bearing age, contraceptive needs should be addressed in collaboration with cardiologists.[6] If a patient with cardiac disease wishes to become pregnant, she should be referred to a pregnancy heart team for evaluation. The ACOG Practice Bulletin recommends delivery in a pregnancy heart center for all pregnant patients with moderate- to high-risk cardiac conditions (modified WHO risk classes III and IV), because outcomes are significantly better for women in these facilities.[3] Many clinicians and institutions have developed care pathways to approach care in these high-risk patients including highly coordinated multidisciplinary teams.

At Montefiore Medical Center, a combined inpatient and outpatient multidisciplinary program was organized to optimize care for these high-risk patients[7]. In the outpatient care, a multidisciplinary office located in the cardiology care rooms was equipped with obstetric examination equipment allowing for same room interviews with both

maternal fetal medicine and cardiology specialists. Obstetric anesthesia consultations and laboratory testing were performed in the same location. The inpatient care consisted of monthly meetings including the following specialties: labor and delivery nursing, intensive care, obstetric and cardiac anesthesia, maternal fetal medicine, cardiology, neonatology, perfusion specialists, blood bank, cardiothoracic surgery, and neonatal intensive care unit. Coordination of highly specialized care is key to the successful management of these women with maternal cardiac disease.

Outcomes for Women with Cardiac Disease

The common theme across the literature is patients with cardiac disease who become pregnant are at risk for serious cardiac complications and pregnancy in these women can lead to adverse long-term outcomes. In 2007, the EURObservational Research Programme (EORP) of the European Society of Cardiology established the Registry of Pregnancy and Cardiac disease (ROPAC) to provide updated information on the impact of cardiac diseases on maternal and fetal mortality and morbidity and to identify areas of where management can be improved. The diagnosis of pulmonary hypertension was identified as the greatest risk factor for an adverse event in pregnant women. Valvular heart disease, hypertensive disorders of pregnancy, and the increasing age of repaired congenital heart lesions also are among the highest risk factors for adverse events in pregnant women. Within 5 years of delivery, hypertensive disorders of pregnancy were associated with all-cause mortality (hazard ratio [HR] 2.21; 95% CI, 1.61-3.03), coronary heart disease (HR, 3.79; 95% CI, 3.09-4.65), and stroke (HR, 3.10, 95% CI, 2.09-4.60).[8] In a retrospective cohort study of patients with heart disease, a multivariate analysis found cesarean delivery (OR 2.68; 95% CI 1.05-6.86) and gestational age <27weeks at delivery (OR, 0.39; 95% CI, 0.22-0.67) were associated with maternal-fetal adverse outcomes, and modified world health organization (mWHO) risk classification had an acceptable predication of adverse outcomes. [9] Although there was an increase in pregnancy among women with high-risk cardiac disease, the incidence of heart failure and/or mortality decreased over the decade.[10]

Delivery mode

Although the rate of cesarean delivery is higher among women with heart disease, vaginal delivery, with good neuraxial anesthesia, is usually the preferred mode of delivery in women with cardiac disease. In a randomized controlled study, maternal and perinatal outcomes were compared in pregnant women with underlying heart disease with favorable Bishop scores at 38 weeks. These women were randomized to induction of labor versus expectant management. There was no difference in the rate of maternal complications, no cardiac complications reported in the induction of labor group, and no reported maternal or neonatal deaths. More women undergoing induction of labor delivered during the daytime which was thought to minimize maternal and fetal complications as multidisciplinary teams were readily available.[11] The decision to undergo an induction of labor requires a multidisciplinary team approach that calls for input from the patient, obstetricians, cardiologists, anesthesiologist, neonatologist, and any other subspecialties that may be necessary in order to ensure a safe delivery. If the recourses necessary to conduct an

induction of labor are not available at a delivery center, then early transfer of pregnancy care to a Maternal Level 3 or 4 Center should be considered.

Interventions/surgery

At times, medical management is not sufficient for pregnant women with cardiac disease and cardiac interventions have to be considered. Although the risk for maternal complications is similar to the nonpregnant state, fetal mortality rate is higher when maternal interventions are required.[12] When possible, percutaneous interventions offer less risk to mother and fetus, but if surgery requiring cardiopulmonary bypass is necessary, modifications to perfusion protocols should be considered: higher flow rates, normothermic perfusion, and intraoperative external fetal monitoring.[13] When a pregnant patient experiences cardiovascular and/or severe respiratory failure, extracorporeal membrane oxygenation (ECMO) is a life saving measure with a survival rate as high as 70% varying by etiology of disease.[14] ECMO should be considered an appropriate therapy for the acutely decompensated mother with reversible cardiorespiratory failure.

Specific disease

Congenital heart disease

Pregnant women with any severity of congenital heart disease when compared to average pregnant women have more non-cardiovascular co-morbidities, increased risk for preterm labor, adverse short-term perinatal outcomes (i.e. low birthweight), and cesarean delivery. [15–17] The risk of pregnancy in women with congenital heart disease and expected complications will vary per lesion type and by the current status of maternal health.[18]

The three most commonly used risk classification models are CARPREG II (Cardiac disease in pregnancy study), ZAHARA (Pregnancy and congenital heart disease), and modified WHO classification (mWHO). In studies by Denayer et al and Wang et al, it was determined that all risk models overestimated maternal cardiac risk and had at best, moderate discrimination for women with congenital heart disease.[19, 20] Although these models can serve as a starting point for risk stratification and coordination of patient care, more research in this area is needed to accurately risk stratify patients. According to the 2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy, mWHO is the most accurate way to classify risk of pregnancy in women with cardiac disease.[1] [Table 1]

A history of uneventful pregnancy can be reassuring in terms of future uneventful pregnancies. In a tertiary single-center retrospective study in Gothenburg, Sweden, maternal cardiac complications in 307 women with congenital heart disease were analyzed. 571 deliveries and 9 miscarriages were reviewed, and if CARPREG I and mWHO scores remained unchanged from the first pregnancy to the second pregnancy, the odds ratio of having an uneventful second pregnancy if the first was uneventful was 5.47 (95% CI 1.76-16.94). 14.6% of the pregnancies experienced maternal cardiac complications with arrhythmia (5.7%) and heart failure (4.4%) being the most common complications. [21] Risk stratification by any of the models above is important to identify women with congenital

heart disease that may require tertiary care as opposed to delivering in a community setting. A tertiary care center can provide additional interventions such as real time transthoracic echocardiography, invasive monitors such as a pulmonary artery catheter, or prophylactic extracorporeal membrane oxygenation cannulation.

In general, women with mWHO lesions in Class I and II may be cared for at Maternal Level I or II centers. Women who require subspecialty care and a cardiologist should, at a minimum, be cared for at Level III centers and, should there be a possible need for peripartum cardiac surgery or extracorporeal membrane oxygenation (ECMO), a woman should be transferred to a hospital with cardiac surgery capabilities, which is likely to be a Level IV center. [22]

Congenital vascular malformations and connective tissue disease—Congenital aortic diseases include coarctation of the aorta and transposition of the great vessels. Of the 303 patients with coarctation of the aorta in the ROPAC database, of which 9.6% were unrepaired, only 4.3% had a major adverse cardiac event, notably new onset heart failure with no previous risk factors, suggesting pregnancy in this population to be safer than previously anticipated.[23]

Transposition of the great vessels may accompany congenital cardiac disease. Even when isolated, the diagnosis does appear to confer increased obstetric risk. Infants born to a mother who had undergone an atrial or arterial switch operation were found to have higher risk of preterm labor and be small for gestational age.[24]

Marfan syndrome, a genetic connective tissue disorder, is associated with increased risk of aortic root aneurysm and dissection during pregnancy. A 21-year single institution review of 30 patients with Marfan syndrome showed overall maternal mortality was 35.7% and fetal mortality was 45.9% despite varying medical and surgical treatment.[25] Risk of a catastrophic outcome likely varies depending on the severity of disease. A retrospective study of 35 patients suggested rate of aortic root and aortic dilation was similar between pregnant and nonpregnant women with diagnosed with Marfan syndrome.[26] Cottrell et al published a case series of 4 patients with Marfan Syndrome and evidence-based suggestions for the pregnant woman with Marfan Syndrome[27]. Specific risk factors include an aortic root diameter greater than 4.0cm, an increase of 0.5cm per year over serial echocardiograms, or history of aortic dissection. Beta adrenergic receptor blockade to prevent wall stress in the aorta beginning from second trimester has been recommended. Regardless of disease severity, with the physiologic changes of pregnancy and the post-partum period (connective tissue changes, increased blood volume and sheer stress to the aorta), the risk of aortic dissection is still high and these women should be monitored closely. Women with aortic dimensions or aortopathies considered to be in mWHO classification III or IV should deliver at Maternal Level 3 or 4 Care Centers.

Valvular heart disease

Valvular heart disease represents one third of the cardiac disease in pregnant women. Rheumatic heart disease is the most common form of valvular heart disease in low-income countries. Valvular heart disease in pregnancy increases both maternal and fetal risk.[28]

The risk varies depending on the type and severity of the lesion, but generally, stenotic lesions are less tolerated than regurgitant lesions in pregnancy. Women with moderate to severe mitral and aortic stenosis are at highest risk of adverse events in pregnancy and should receive preconception counseling.[29] Vaginal delivery is recommended for most women with valvular disease, but mode of delivery should be chosen based on obstetric and fetal indications specific to each pregnancy. Advances in minimally invasive treatments of valvular heart disease present options for interventions that do not require cardiopulmonary bypass and may therefore pose less risk to the fetus. These interventions which may be considered in pregnancy if the benefits outweigh risk.

The pregnancy-induced hypercoagulable state increases the risk of mechanical prosthetic heart valve thrombosis and currently there is no optimal anti-coagulation therapy as all management options have advantages and disadvantages. Warfarin has been the anticoagulant of choice in this setting as it has the best anti-coagulation efficacy, but it is linked to fetal demise and teratogenicity. If anti-coagulation can be achieved with low-dose warfarin (5mg/day) then it is usually the regimen of choice as it provides effective anti-coagulation and minimizes fetal risk even during the first trimester.[30] Low-molecular weight heparin is an acceptable alternative option when appropriate dosing and monitoring of peak and trough anti-Xa levels can be achieved.[31] The decision to temporarily hold anticoagulation for delivery and/or neuraxial blockade requires a multidisciplinary team discussion, and providers should adhere to national SOAP guidelines to ensure a safe neuraxial anesthesia placement.[32]

Heart transplant

There is limited information in the literature about maternal and fetal outcomes among orthotopic heart transplant recipients. A recent case series followed pregnancies of 5 women who had a history of heart transplant. Preconception counselling, immunosuppressant titrations, regular monitoring for common complications of pregnancy are necessary to avoid graft rejection and teratogenic complications in this population.[33]

Pulmonary Hypertension

The different etiologies and varying severity of pulmonary hypertension makes this rare but deleterious disease in pregnancy challenging to study without multicenter registries. Pulmonary hypertension is a mWHO class IV lesion, so these women are advised to avoid pregnancy. ACOG Practice Bulletin 212 states that a patient with severe pulmonary hypertension requires care and delivery at a center with maternal–fetal medicine, obstetric and cardiac anesthesia, a pulmonary hypertension specialist, and advanced heart failure care options, such as extracorporeal membrane oxygenator management.[3] Despite improvements in pulmonary vasodilators, heightened awareness of the role of extracorporeal membrane oxygenation and improved knowledge of appropriate cardiology, anesthesia and obstetric care of women with pulmonary hypertension who become pregnant, maternal morbidity and mortality remain high in this population, yet perhaps not prohibitive in women with mild disease.[34] A series of 59 women with pulmonary hypertension in pregnancy revealed a 20.8% mortality rate in women with severe PH (PAH>79mmHg

n=24) with no mortalities in the women with mild or moderate PH, suggesting that with contemporary care, pregnancy may be safe in women with mild PH.[35]

Peripartum cardiomyopathy/Heart Failure

Heart failure occurs in pregnancy both in women with known cardiac disease and women without a history of cardiac disease. Diagnosis of heart failure in pregnancy can be challenging as signs and symptoms of heart failure overlap with those of pregnancy. Clinical observation of functional status, measurement and monitoring of BNP levels during each trimester, and transthoracic echocardiography can assist in predicting potential cardiac decompensation during pregnancy.[3] Pregnancy can unmask underlying heart disease or heart disease can develop de novo in pregnancy, due to cardiomyopathy but also as a result of preeclampsia. Jayasooriya et al examined the etiology of heart failure in 120 pregnancies managed in a Canadian tertiary care center, identifying preeclampsia (28%), cardiomyopathy (22%), and valvular heart disease (18%) as the most common causes of heart failure. The majority of women received neuraxial anesthesia for both vaginal and cesarean delivery. In the majority of cases, symptoms and signs of heart failure were present: dyspnea (88.3%), respiratory crackles (81.7%) and peripheral edema (81.7%).[36] Sliwa et al examined the 739 women with peripartum cardiomyopathy in the ROPAC database and found conversely that peripheral edema and pulmonary rales were absent in 42% and 41% of women with peripartum cardiomyopathy, respectively.[37] This retrospective cohort study revealed symptom onset most commonly occurred within one month of delivery (44%) and 6-month outcomes included thromboembolism (7%), readmission (10%), and mortality (6%).[37] Symptoms of heart failure in women who are at higher risk for heart failure (older women, women with preeclampsia or hypertensive disorders, or history of cardiac disease) should prompt BNP testing and or echocardiography.

Arrhythmias

The most common cardiac complication among women with cardiac disease is arrhythmias. Atrial fibrillation is the most common arrhythmia in pregnant women. Manolis et al summarizes management options for arrhythmias in pregnancy. Antiarrhythmic medications may be warranted depending on the clinical picture but should be avoided in the first trimester if possible.[38] If electrical cardioversion is necessary, it is considered safe for both mother and fetus.[38] For refractory arrhythmia, interventional procedures can be performed when needed with minimal radiation exposure including reduction of fluoroscopy dose by the interventionalist and additional maternal lead.[38]

Broader Implications of Cardiovascular Disease on Maternal Health

While the risk of complications is highest among women with pre-existing cardiac disease, women without a prior cardiac diagnosis may also be at risk for adverse cardiac events in the post-partum period. The physiologic stresses of pregnancy (mostly the increase in blood volume and need for augmented cardiac output) may unmask pre-existing cardiovascular disease, or, pregnancy may precipitate cardiovascular decompensation.

Risk factors for cardiovascular disease during and after pregnancy include race (specifically, African American and American Indians/Alaskan Native race), hypertension, obesity,

advanced maternal age, and low socioeconomic status. These risk factors are correlated with a higher risk of cardiovascular disease during and after pregnancy.[39] The risk for cardiac readmission among women without a cardiac diagnosis was examined in the Nationwide Readmission Database. Postpartum readmission rates for a cardiac condition, namely arrhythmia, heart failure, and pulmonary heart disease, occurred in 26.8 per 10,000 women.[40] The following factors were all associated with increased risk of 9-month postpartum cardiac admissions: hypertensive diseases of pregnancy (aRR 2.19, 95% CI 2.09, 2.30), severe maternal morbidity at delivery (aRR 2.06, 95% CI 1.79, 2.37), chronic hypertension (aRR 2.52, 95% CI 2.31, 2.74), lupus (aRR 4.62, 95% CI 3.82, 5.60), venous thromboembolism during delivery (aRR 3.72, 95% CI 2.75, 5.02), Medicaid insurance (aRR 1.57, 95% CI 1.51, 1.64), Medicare insurance (aRR 3.06, 95% CI 2.70, 3.46), and maternal ages 35-39 and 40-54 years (aRR 1.24, 95% CI 1.17, 1.32, aRR 1.74, 95% CI 1.60, 1.90, respectively).[40] Close post-partum follow-up, especially within the first 30 days after discharge in women with the aforementioned risk factors, may improve the diagnosis and treatment of cardiac events that may arise after delivery.

Regardless of whether cardiac disease is diagnosed before pregnancy or not, women who suffer cardiovascular complications of pregnancy face elevated lifelong risk for cerebrovascular complications including stroke and atherosclerotic cardiovascular disease. Hypertensive disorders and low birth weight were associated with atherosclerotic cardiovascular disease later in life and delaying treatment of hypertensive disorders of pregnancy was associated with increased maternal cardiac risk.[41, 42] In a prospectively cohort, 3.6% of pregnancies were complicated by serious cardiac events and almost one-half of these were preventable based on The Harvard Medical Study criteria.[43] This leads to the topic of identifying risk factors that may predict maternal complications.

The ability to identify risk factors in this high-risk cohort allows for care planning to improve long term outcomes. Knowledge of these risk factors and prior pregnancy history can be important for the anesthesiologists during a standard pre-anesthetic evaluation.

Conclusion

The leading cause of morbidity and mortality in pregnant women is cardiac disease. Preconception counseling is useful for optimizing patients for pregnancy and setting appropriate expectations about care and outcomes. A multidisciplinary team approach is key for successful management of the cardio-obstetric patient.

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Key points:

- Cardiovascular disease is the leading cause of death among pregnant women in the United States.
- Many clinicians and institutions have developed care pathways to approach care in these high-risk patients including highly coordinated multidisciplinary teams.
- The diagnosis of pulmonary hypertension is the greatest risk factor for an adverse event in pregnant women.
- Vaginal delivery, with good neuraxial anesthesia, is usually the preferred mode of delivery in women with cardiac disease, although the rate of cesarean delivery is higher among women with heart disease.

Table 1

Modified World Health Organization classification of maternal cardiovascular risk

	mWHO I	mWHO II	mWHO II–III	mWHO III	mWHO IV
Diagnosis (if otherwise well and uncomplicated)	<ul style="list-style-type: none"> Small or mild <ul style="list-style-type: none"> pulmonary stenosis patent ductus arteriosus mitral valve prolapse Successfully repaired simple lesions <ul style="list-style-type: none"> atrial or ventricular septal defect patent ductus arteriosus anomalous pulmonary venous drainage Atrial or ventricular ectopic beats, isolated 	<ul style="list-style-type: none"> Unoperated atrial or ventricular septal defect Repaired tetralogy of Fallot Most arrhythmias (supraventricular arrhythmias) Turner syndrome without aortic dilatation 	<ul style="list-style-type: none"> Mild left ventricular impairment (EF >45%) Hypertrophic cardiomyopathy Native or tissue valve disease not considered WHO I or IV (mild mitral stenosis, moderate aortic stenosis) Marfan or other HTAD syndrome without aortic dilatation Aorta <45 mm in bicuspid aortic valve pathology Repaired coarctation Atrioventricular septal defect 	<ul style="list-style-type: none"> Moderate left ventricular impairment (EF 30–45%) Previous peripartum cardiomyopathy without any residual left ventricular impairment Mechanical valve Systemic right ventricle with good or mildly decreased ventricular function Pontan circulation if otherwise the patient is well and the cardiac condition uncomplicated Unrepaired cyanotic heart disease Moderate mitral stenosis Severe asymptomatic aortic stenosis Moderate aortic dilatation Severe asymptomatic aortic stenosis 40–45 mm in Marfan syndrome or other HTAD 45–50 mm in bicuspid aortic valve Turner syndrome, ASI 20–25mm/m² tetralogy of Fallot <50 mm Ventricular tachycardia 	<ul style="list-style-type: none"> Pulmonary arterial hypertension Severe systemic ventricular dysfunction (EF <30% or NYHA class III–IV) Previous peripartum cardiomyopathy with any residual left ventricular impairment Severe mitral stenosis Severe symptomatic aortic stenosis Systemic right ventricle with moderate or severely decreased ventricular function Severe aortic dilatation >45 mm in Marfan syndrome or other HTAD >50 mm in bicuspid aortic valve Turner syndrome, ASI >25 mm/m² tetralogy of Fallot >50 mm Vascular Ehlers–Danlos Severe (re)coarctation Pontan with any complication
Risk	No detectable increased risk of maternal mortality and no/mild increased risk in morbidity	Small increased risk of maternal mortality or moderate increase in morbidity	Intermediate increased risk of maternal mortality or moderate to severe increase in morbidity	Significantly increased risk of maternal mortality or severe morbidity	Extremely high risk of maternal mortality or severe morbidity
Maternal cardiac event rate	2.5–5%	5.7–10.5%	10–19%	19–27%	40–100%
Counselling	Yes	Yes	Yes	Yes: expert counselling required	Yes: pregnancy contraindicated: if pregnancy occurs, termination should be discussed
Care during pregnancy	Local hospital	Local hospital	Referral hospital	Expert center for pregnancy and cardiac disease	Expert center for pregnancy and cardiac disease
Minimal follow-up visits during pregnancy	Once or twice	Once per trimester	Bimonthly	Monthly or bimonthly	Monthly
Location of delivery	Local hospital	Local hospital	Referral hospital	Expert center for pregnancy and cardiac disease	Expert center for pregnancy and cardiac disease

ASI = aortic size index; EF = ejection fraction; HTAD = heritable thoracic aortic disease; mWHO = modified World Health Organization classification; NYHA = New York Heart Association; WHO = World Health Organization.