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Pregnancy and Heart Disease

Maternal heart disease has emerged as a major threat to safe motherhood and women's long-term cardiovascular health. In the United States, disease and dysfunction of the heart and vascular system as "cardiovascular disease" is now the leading cause of death in pregnant women and women in the postpartum period (1, 2) accounting for 4.23 deaths per 100,000 live births, a rate almost twice that of the United Kingdom (3, 4). The most recent data indicate that cardiovascular diseases constitute 26.5% of U.S. pregnancy-related deaths (5). Of further concern are the disparities in cardiovascular disease outcomes, with higher rates of morbidity and mortality among nonwhite and lower-income women. Contributing factors include barriers to prepregnancy cardiovascular disease assessment, missed opportunities to identify cardiovascular disease risk factors during prenatal care, gaps in high-risk intrapartum care, and delays in recognition of cardiovascular disease symptoms during the puerperium. The purpose of this document is to 1) describe the prevalence and effect of heart disease among pregnant and postpartum women; 2) provide guidance for early antepartum and postpartum risk factor identification and modification; 3) outline common cardiovascular disorders that cause morbidity and mortality during pregnancy and the puerperium; 4) describe recommendations for care for pregnant and postpartum women with preexisting or new-onset acquired heart disease; and 5) present a comprehensive interpregnancy care plan for women with heart disease.

Background Emerging Trends in Cardiovascular Disease

Cardiovascular disease affects approximately 1–4% of the nearly 4 million pregnancies in the United States each year. The incidence of pregnancy in women with congenital heart disease and acquired heart disease is on the rise (6). In developed countries, maternal morbidity and mortality secondary to congenital heart disease have remained relatively stable at 11% and 0.5% (7), respectively; however, the United States experienced a significant linear increase in maternal congenital heart disease (6.4 to 9.0 per 10,000 delivery hospitalizations) from 2000 to 2010 (8), and maternal deaths due to acquired heart disease remain high. From 2002 to 2011, 22.2% of maternal deaths in Illinois were due to cardiovascular disease, 97.1% of which were related to acquired heart disease (9). This rising trend in maternal deaths related to cardiovascular disease appears to be due to acquired heart disease (10).

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The most common presentations of maternal acquired heart disease during pregnancy and the postpartum periods are heart failure, myocardial infarction, arrhythmia, or aortic dissection (11, 12). Diagnosis can be challenging because the overlap of cardiovascular symptoms with those of normal pregnancy may lead to delays in diagnosis and subsequent care (10). If cardiovascular disease were to be considered in the differential diagnosis by treating health care providers, it is estimated that a quarter or more of maternal deaths could be prevented (10, 13, 14). A recent study of maternal cardiovascular mortality in Illinois found that 28.1% of maternal cardiac deaths were potentially deemed preventable due to health care provider issues, patient features (eg, nonadherence, obesity) (9), and health care system factors related to access. In the United Kingdom, a 2015 report on maternal mortality concluded that substandard health care accounted for more than 50% of cardiac deaths, half of which were considered avoidable (15).

Risk Factors for Cardiovascular Disease Across the Maternity Care Continuum

There are four key risk factors linked to cardiovascular disease-related maternal mortality:

- Race/Ethnicity: Non-Hispanic black women have a 3.4 times higher risk of dying from cardiovascular diseaserelated pregnancy complications compared with non-Hispanic white women independent of other variables (5). Between 2011 and 2013, there were 43.5 pregnancy-related deaths per 100,000 live births for non-Hispanic black women compared with 11.0 and 12.7 pregnancy-related deaths per 100,000 live births for Hispanic and non-Hispanic white women, respectively (5). This disparity can be explained in part by exposure to structural, institutional, and systemic barriers that contribute to a higher rate of comorbidities.
- 2. Age: Age older than 40 years increases the risk of heart disease-related maternal death 30 times the risk for women younger than 20 years (16, 17).
- 3. Hypertension: Hypertensive disorders affect up to 10% of pregnancies and can lead to maternal morbidity and mortality. Severe and early-onset hypertension during pregnancy put women at an increased risk of cardiac compromise during or following delivery (18–20). In pregnancies complicated by hypertension, the incidence of myocardial infarction and heart failure is 13-fold and 8-fold higher, respectively, than in healthy pregnancies (18).
- 4. Obesity: Prepregnancy obesity increases maternal death risk due to a cardiac cause (21), especially if associated with moderate-to-severe obstructive sleep apnea (22). In the United Kingdom from 2006 to

2008, 60% of maternal deaths in which the body mass index (BMI, calculated as weight in kilograms divided by height in meters squared) was known were in overweight or obese women (15).

The presence of one or more of these risk factors should raise the threshold for suspicion that a patient is at-risk for maternal heart disease and pregnancy-related morbidity and mortality (23).

Social Determinants of Disparities in Cardiovascular Disease in Health and Health Care

Increased rates of cardiovascular disease-related complications among women of color are explained, in part, by racial and ethnic bias in the provision of health care and health system processes (24). Patient, physician, and health system-level factors can affect outcomes. Physician implicit and explicit bias and overt racism often can result in missed diagnoses or inappropriate treatment. Health system barriers to efficient triage based on symptom severity, language barriers, and differences in cultural humility are important factors that must be investigated to understand fully the pervasiveness of disparities that women of color face when encountering the health care system (25). Moreover, women of color may have experienced injustice in health care processes, leading to mistrust of the medical system. These factors contribute to a disproportionately higher rate of pregnancyassociated complications among women of color which, in turn, places these women not only at a greater risk of cardiovascular events in the postpartum period but also increase their lifetime risk of cardiovascular disease. Thus, it is important to improve education for these women and their trusted lay sources of information by emphasizing the value of medical care and the importance of healthy dietary habits and regular exercise. Non-Hispanic black women are more likely to develop gestational diabetes mellitus, preeclampsia, and have a preterm delivery or low-birthweight infant compared with non-Hispanic white women (23, 26). These health disparities often are amplified by missed opportunities to identify cardiovascular disease risk factors before pregnancy and limited access to cardiacrelated care algorithms during intrapartum and postpartum care (23, 27). Additionally, the higher rate of obesity among racial and ethnic nonwhite groups independently contributes to disparities in the development of adverse pregnancy outcomes leading to long-term risk of cardiovascular disease. A higher prevalence of postpartum weight retention and persistence of high-glucose levels among women with gestational diabetes mellitus places them at increased risk of cardiovascular disease (28, 29).

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Physiologic Changes in Pregnancy That Affect Cardiovascular Stress

Pregnancy is a natural stress test because the cardiovascular system undergoes structural and hemodynamic adaptations to sustain a high-volume load. An understanding of these physiologic changes is essential for health care providers.

Hemodynamic Changes

Antepartum. Because of increases in estrogen and progesterone and the activation of the renin-angiotensinaldosterone system, pregnancy causes a continuous increase in cardiac output and plasma volume and a decrease in maternal systemic vascular resistance (30). Blood pressure initially decreases but increases in the third trimester (31, 32) (Table 1). Uterine mechanical compression of the inferior vena cava can occur during the second and third trimesters, potentially reducing venous return to the right ventricle, causing a postural hypotensive syndrome (33) and exacerbating lower-extremity edema. These changes are amplified in women with multiple gestations.

Intrapartum and Postpartum. During labor and after delivery, there are dramatic changes in cardiac output, heart rate, blood pressure, and plasma volume (34, 35).

Although heart rate and blood pressure normally decrease within 48 hours postpartum, blood pressure may increase again between days 3–6 due to fluid shifts (36) (Table 1). During this period, clinicians should monitor patients for hypertensive complications and those related to fluid overload (37). Increased hydrostatic pressure and decreased colloid osmotic pressure render women with cardiovascular disease susceptible to pulmonary edema at the time of delivery and immediately postpartum, particularly in women with severe cardiovascular disease and excessive intravenous fluid administration or preeclampsia, or both. Increased maternal plasma atrial natriuretic peptide levels in the first week postpartum allow for postpartum diuresis (38). Maternal hemodynamics generally return to a prepregnancy state 3–6 months after delivery.

Structural Changes

The heart ventricles adapt to the plasma volume increase during pregnancy. Left ventricular end diastolic volume increases by approximately 10% (39) and left and right ventricular mass increase by approximately 50% and 40%, respectively (40). Reports of ejection fraction during pregnancy are varied. Ejection fractions in some women show no change, (39) although others decrease

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Table 1.	Cardiovascular	Changes in a	Normal	Pregnancy
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	First Trimester	Second Trimester	Third Trimester	Stage 1 Labor	Stage 2 Labor	Early Postpartum	3–6 months Postpartum
Cardiac output	↑5–10%	↑↑35−45%		130%	↑↑50%	↑↑↑60–80% immediately, then rapidly decreases within the first hour	Return to prepregnancy values
Heart rate	↑3–5%	10−15%	15−20%	During contrac ↑40–50	tions:	↓5–10% within 24 hours; con- tinues to decrease throughout the first 6 weeks	Return to prepregnancy values
Blood pressure	↓10%	↓5%	↑5%	During contrac ↑ SBP 15 ↑ DBP 10	tions: -25%	\downarrow SBP 5–10% within 48 hours; may increase again between days 3–6 due to fluid shifts	Return to prepregnancy values
Plasma volume	↑	↑↑40-50%)	1	$\uparrow\uparrow$	↑↑↑500 mL due to autotransfusion	Return to prepregnancy values

Abbreviations: SBP, systolic blood pressure; DBP, diastolic blood pressure.

*Hemodynamic changes that occur during pregnancy, labor, and postpartum (compared with prepregnancy) should be understood to identify early interventions (such as blood pressure control and diuresis) that may be needed to prevent clinical deterioration in a woman with cardiovascular disease.

Data from Kuhn JC, Falk RS, Langesaeter E. Haemodynamic changes during labour: continuous minimally invasive monitoring in 20 healthy parturients. Int J Obstet Anesth 2017;31:74–83; Ouzounian JG, Elkayam U. Physiologic changes during normal pregnancy and delivery. Cardiol Clin 2012;30:317–29; Sanghavi M, Rutherford JD. Cardiovascular physiology of pregnancy. Circulation 2014;130:1003–8; Shen M, Tan H, Zhou S, Smith GN, Walker MC, Wen SW. Trajectory of blood pressure change during pregnancy and the role of pre-gravid blood pressure: a functional data analysis approach. Sci Rep 2017;7:6227; Sohnchen N, Melzer K, Tejada BM, Jastrow-Meyer N, Othenin-Girard V, Irion O, et al. Maternal heart rate changes during labour. Eur J Obstet Gynecol Reprod Biol 2011;158:173–8; and Walters BN, Walters T. Hypertension in the puerperium [letter]. Lancet 1987;2:330.

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(41, 42). Importantly, approximately 20% of women have diastolic dysfunction at term, which may be associated with dyspnea on exertion (41, 43). Structural changes of the maternal heart return to baseline before 1 year postpartum.

Hematologic, Coagulation, and Metabolic Changes

Hematologic, coagulation, and metabolic changes in pregnancy are important contributors to cardiovascular risk. Although intensified erythropoiesis in pregnancy increases red blood cell mass by 20-30%, this increase is proportionally lower than the increase in plasma volume, resulting in physiologic anemia from hemodilution. Because severe anemia may be associated with heart failure and myocardial ischemia, hemoglobin or hematocrit levels should be checked each trimester in women with cardiovascular disease. Pregnancy is associated with physiologic and anatomic changes that increase the risk of thromboembolism, including hypercoagulability, venous stasis, decreased venous outflow, compression of the inferior vena cava and pelvic veins by the enlarging uterus, and decreased mobility (44). Pregnancy also alters the levels of coagulation factors normally responsible for hemostasis. The overall effect of these changes is an amplified thrombogenic state with an increased risk of thromboembolism. Certain disorders, such as antiphospholipid antibody syndrome and high-risk thrombophilia and smoking, further increase the risk of thrombosis and embolism during pregnancy. From a metabolic standpoint, pregnancy is a catabolic state that leads to insulin resistance and an atherogenic lipid profile with elevated serum fatty acids.

Signs and Symptoms of Heart Disease

Normal pregnancy and postpartum symptoms and signs can overlap with findings reflective of underlying heart disease (Table 2). Health care providers should become familiar with the signs and symptoms of cardiovascular disease as an important step toward improving maternal outcomes.

Clinical Considerations and Recommendations

What are the prerequisites of pregnancy preparation and prepregnancy counseling for patients with known heart disease?

Whenever possible, optimization of maternal health status should be attempted and achieved before pregnancy. Risk

to a woman's heart and cardiovascular system engendered by pregnancy depends upon the specific type of heart disease and clinical status of the patient. Women with known cardiovascular disease (Table 3) should be evaluated by a cardiologist ideally before pregnancy or as early as possible during the pregnancy for an accurate diagnosis and assessment of the effect pregnancy will have on the underlying cardiovascular disease, to assess the potential risks to the woman and fetus, and to optimize the underlying cardiac condition. A detailed history, including family history and any current cardiovascular symptoms, physical examination, and review of medical records, including prior cardiovascular testing and interventions, should be obtained (45-48). A comprehensive cardiovascular family history should include inquiry about structural, vascular, or rhythm disorders and sudden unexpected death. Clues to a familial cardiac condition may include prior cardiac surgery, myocardial infarction, stroke, aortic dissection, and sudden death. Upon confirmation of family history of cardiovascular disease, health care providers should ask whether genetic testing has been performed. A known gene mutation, such as MYH7 for cardiomyopathy, may have implications for a patient's individual risk of developing cardiomyopathy and may alert the patient and care team to plan postpartum surveillance and to screen offspring (49).

Patients with moderate and high-risk cardiovascular disease should be managed during pregnancy, delivery, and the postpartum period in medical centers with a multidisciplinary Pregnancy Heart Team (Table 4) that includes obstetric providers, maternal-fetal medicine subspecialists, cardiologists, and an anesthesiologist at a minimum. Ad hoc members may include cardiac surgeons, interventional cardiologists, cardiac imaging specialists, electrophysiologists, pulmonary hypertension and heart failure specialists, adult congenital cardiologists, emergency physicians, intensivists, neonatologists, geneticists, mental health specialists, primary care physicians, other medical specialists, advanced practice providers and specialized nurses, midwives, or pharmacists. The members of the Pregnancy Heart Team (Table 4) should work together to assess and counsel the patient regarding the individualized risks of her underlying cardiac condition should she become pregnant, the potential risk of transmission of congenital heart or genetic disease to the child, and the need for increased medical surveillance during the antepartum, parturition, and postpartum phases of pregnancy (Table 3).

A triad of cardiovascular risk screening, patient education, and multidisciplinary team planning has been suggested to optimize outcomes in women with known cardiovascular disease (50). It is imperative to

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Table 2. How to Differentiate Common Signs and Symptoms of Normal Pregnancy Versus Those That Are Abnormal and Indicative of Underlying Cardiac Disease

	ROUTINE CARE	CAUTION* [†]	STOP ^{†‡}
	Reassurance	Nonemergent Evaluation	Prompt Evaluation Pregnancy Heart Team
History of CVD	None	None	Yes
Self-reported symptoms	None or mild	Yes	Yes
Shortness of breath	No interference with activities of daily living; with heavy exertion only	With moderate exertion, new-onset asthma, persistent cough, or moderate or severe OSA [§]	At rest; paroxysmal nocturnal dyspnea or orthopnea; bilateral chest infiltrates on CXR or refractory pneumonia
Chest pain	Reflux related that resolves with treatment	Atypical	At rest or with minimal exertion
Palpitations	Few seconds, self-limited	Brief, self-limited episodes; no lightheadedness or syncope	Associated with near syncope
Syncope	Dizziness only with prolonged standing or dehydration	Vasovagal	Exertional or unprovoked
Fatigue	Mild	Mild or moderate	Extreme
Vital signs	Normal		
HR (beats per minute)	<90	90–119	≥120
Systolic BP (mm Hg)	120–139	140–159	≥160 (or symptomatic Iow BP)
RR (per minute)	12-15	16–25	≥25
Oxygen saturation	>97%	95–97%	<95% (unless chronic)
Physical examination	Normal		
JVP	Not visible	Not visible	Visible >2 cm above clavicle
Heart	S3, barely audible soft systolic murmur	S3, systolic murmur	Loud systolic murmur, diastolic murmur, S4
Lungs	Clear	Clear	Wheezing, crackles, effusion
Edema	Mild	Moderate	Marked

Abbreviations: BP, blood pressure; CVD, cardiovascular disease; CXR, chest x-ray; HR, heart rate; JVP, jugular venous pressure; OSA, obstructive sleep apnea; RR, respiratory rate.

*If unclear, any combination of factors in the yellow column that add up to 4 or more should prompt further evaluation.

[†]Data in this column from Afshan B. Hameed, Christine H. Morton, and Allana Moore. Improving Health Care Response to Cardiovascular Disease in Pregnancy and Postpartum. Developed under contract #11-10006 with the California Department of Public Health, Maternal, Child and Adolescent Health Division. Published by the California Department of Public Health, 2017. Available at https://www.cmqcc.org/resources-toolkits/toolkits/improving-health-care-response-cardiovascular-disease-pregnancy-and.

[‡]History of CVD or signs and symptoms in the red column should lead to urgent evaluation by the Pregnancy Heart Team.

§Should raise concern about heart failure and should promptly be evaluated.

Modified from Thorne S. Pregnancy and native heart valve disease. Heart 2016;102:1410-7.

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Table 3. Modified World Health Organization Pregnancy Risk Classification for Women With Preexisting Cardiovascular Disease

Modified WHO Pregnancy Risk Classification		
(Risk of Pregnancy by medical condition)		Pregnancy Care
Suggested follow-up*	Specific Cardiac Lesions	Delivery Location
mWHO Risk Class I No detectable increased risk of maternal mortality and no or mild increase in morbidity (2–5% risk of maternal cardiac event rate) Follow-up: Cardiology evaluation once or twice during pregnancy	 Uncomplicated, small, or mild Pulmonary stenosis Patient ductus arteriosus Mitral valve prolapse Successfully repaired simple lesions (atrial or ventricular septal defect, patent ductus arteriosus, anomalous pulmonary venous drainage) Atrial or ventricular ectopic beats, isolated 	 Prepregnancy/pregnancy counseling Care at local hospital Delivery at local hospital*
mWHO Risk Class II Small increased risk of maternal mortality or moderate increase in morbidity (6–10% maternal cardiac event rate) Follow-up: Cardiology, every trimester	 Unoperated atrial or ventricular septal defect Repaired Tetralogy of Fallot or aortic coarctation Most arrhythmias (supraventricular arrhythmias) Turner syndrome without congenital cardiac disease 	 Prepregnancy/pregnancy counseling Pregnancy Heart Team[*] consultation/ counseling Care at local hospital Delivery at local hospital[*]
mWHO Risk Classes II and III Intermediate increased risk of maternal mortality or moderate to severe increase in morbidity (11–19% maternal cardiac event rate) Follow-up: Cardiology, every trimester	 Mild left ventricular impairment (EF >45%) Hypertrophic cardiomyopathy Native or bioprosthetic valve disease not considered mWHO Risk Class I or IV (mild mitral stenosis, moderate aortic stenosis) Marfan or other HTAD syndrome without aortic dilation Aorta <45 mm in bicuspid aortic valve pathology Repaired coarctation without residua (non-Turner) Atrioventricular septal defect 	 Prepregnancy/pregnancy counseling Pregnancy heart team[*] consultation/ counseling Care at an appropriate level hospital (critical members of the Pregnancy Heart Team[*] available depending on cardiac disease) Delivery at an appropriate level hospital^{*†}
 Pre-mWHO Risk Class III Significantly increased risk of maternal mortality or severe morbidity (20–27% maternal cardiac event rate) Follow-up: Cardiology, every 1–2 months 	 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 Uncomplicated Fontan circulation, Unrepaired cyanotic heart disease Other complex heart disease Moderate mitral stenosis Severe asymptomatic aortic stenosis Moderate aortic dilation (40-45 mm in Marfan syndrome or other HTAD; 45-50 mm in bicuspid aortic valve; Turner syndrome ASI 20-25 mm/m²; Tetralogy of Fallot <50 mm) Ventricular tachycardia 	 Prepregnancy/pregnancy counseling Pregnancy Heart Team[*] consultation/ counseling Care at an appropriate level hospital[†] Delivery at an appropriate level hospital^{*†}
		(continued)

(continued)

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Table 3. Modified World Health Organization Pregnancy Risk Classification for Women With Preexisting Cardiovascular Disease (continued)

Risk Classification			
(Risk of Pregnancy by medical condition)		Pregnancy Care	
Suggested follow-up*	Specific Cardiac Lesions	Delivery Location	
mWHO Risk Class IV	• Pulmonary arterial hypertension	• Pregnancy Heart Team [*] consultation/	
Pregnancy contraindicated	 Severe systemic ventricular dysfunction (EF <30%, NYHA III-IV) 	 counseling Care at an appropriate level hospital[‡] (critical members of the Pregnancy Heart Team[*] 	
Discuss induced abortion	Previous peripartum cardiomyopathy with any residual left ventricular dysfunction	 available depending on cardiac disease) Delivery at an appropriate level hospital*[‡] 	
Extremely high risk of maternal	 Severe mitral stenosis 		
mortality or severe morbidity	 Severe symptomatic aortic stenosis 		
(>27% maternal cardiac event rate)	 Systemic right ventricle with moderate to severely decreased ventricular function 		
Follow-up: Cardiology follow-up every month (minimum)	 Severe aortic dilation (>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 Fontan circulation with any complication 		

Modified WHO Pregnancy

Abbreviations: ASI, aortic size index; EF, ejection fraction; HTAD, hereditary thoracic aortic disease; mWHO, modified World Health Organization; NYHA, New York Heart Association.

*Pregnant women with a positive cardiac history or findings, or both, should receive prenatal, intrapartum, and postpartum care in a hospital setting that represents an appropriate maternal level of care that is at Level II or higher depending upon the specific cardiac lesion(s) that are present. "The goal of regionalized maternal care is for pregnant women at high risk to receive care in facilities that are prepared to provide the required level of specialized care, thereby reducing maternal morbidity and mortality in the United States." (Levels of maternal care. Obstetric Care Consensus No. 2. American College of Obstetricians and Gynecologists. Obstet Gynecol 2015;125:502–15).

[†]**mWHO Risk Class III.** Critical members of the Pregnancy Heart Team available depending on cardiac disease. For example: A mechanical valve patient requires care at a center with cardiologist/maternal–fetal medicine team who monitor and adjust anticoagulation weekly, delivery at a center with obstetric anesthesia, and advance cardiac care options including access to emergency cardiac surgery should acute prosthetic valve thrombosis necessitate emergency intervention.

[†]**mWHO Risk Class IV.** For example, a severe pulmonary hypertension patient 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 ventricular assist device and extracorporeal membrane oxygenator management.

Adapted from Thorne S, MacGregor A, Nelson-Piercy C. Risks of contraception and pregnancy in heart disease. Heart 2006;92:1520-5).

identify cardiac conditions associated with significantly increased maternal mortality or severe morbidity. Pregnancy is not recommended for women in modified World Health Organization (WHO) pregnancy risk category IV (Table 3) (51–53). Discussion of cardiovascular disease with the woman should include the possibilities that 1) pregnancy can contribute to a decline in cardiac status that may not return to baseline after the pregnancy; 2) maternal morbidity or mortality is possible; and 3) fetal risk of congenital heart or genetic conditions, fetal growth restriction, preterm birth, intrauterine fetal demise, and perinatal mortality is higher when compared with risk when cardiovascular disease is not present (54–56). Approximately one third of cardiac patients will require medication during pregnancy (57), and special emphasis should be placed on agents to be avoided, and when feasible, switching to safer alternatives before pregnancy (see Table 5). Certain medications, such as angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, and aldosterone antagonists should be avoided if possible because of the risk of potential fetal adverse effects (58). However, there may be cardiac conditions that are controlled only by medications or interventions that have potential teratogenic effects that must be used during pregnancy despite known risk to the fetus, such as warfarin in a patient with a mechanical valve prosthesis (57). In these circumstances, the

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_	Modified WHO Pregnancy Risk Classification I	Modified WHO Pregnancy Risk Classification II	Modified WHO Pregnancy Risk Classifications III and IV
Pregnancy Heart Team Members	Obstetrician, family medicine practitioner, internist Cardiologist consultation	Obstetrician, family medicine practitioner, internist Maternal–fetal medi- cine subspecialist Cardiologist consultation	Obstetrician, family medicine practitioner, maternal-fetal medicine subspecialist, internist, obstetric anesthesiologist, cardiology sub- specialists in adult congenital/aortopathy [*] , heart rhythm [*] , heart failure [*] , pulmonary hypertension [*] , and cardiac imaging [*] Interventional cardiologist [*] Cardiac surgeon [*] Neonatologist [*] Geneticist [*] Mental health specialist [*] Pharmacist [*]

Abbreviation: WHO, World Health Organization.

*Ad Hoc members of a Pregnancy Heart Team

specialists who constitute the Pregnancy Heart Team (Table 4) should review the risks, benefits, and alternative therapeutic options with the patient and document in the medical record a summary of what is discussed and recommended. Patients should be encouraged not to stop any medications until they have reviewed management options with their care team.

Although the goal of prepregnancy counseling is to identify and modify risks to improve pregnancy outcome, the individual's choices will be conditional upon her values and preferences, and patient autonomy must be ensured. A collaborative discussion with shared decision making should take place between the Pregnancy Heart Team (Table 4), the patient, and her family. A personalized approach estimating the maternal and fetal hazards related to the patient's specific cardiac disorder and the patient's pregnancy plans can provide anticipatory guidance to help support her decision making. For some patients, the prepregnancy evaluation may suggest a pregnancy risk that is unacceptable (Table 3). For those women, reproductive alternatives, such as surrogacy or adoption, and effective contraceptive methods should be discussed (58).

Why is risk assessment indicated, what types are recommended, and which patients should be referred to centers with a high level of care?

A key area of competence and expertise for obstetric care providers is the ability to differentiate between

common symptoms of pregnancy and those suggestive of cardiovascular disease. Maternal mortality reviews indicate that most women who die from cardiovascular disease had either undiagnosed cardiovascular disease or new-onset cardiovascular disease of pregnancy, specifically peripartum cardiomyopathy. Therefore, all women should be assessed for cardiovascular disease in the antepartum and postpartum periods using the California Improving Health Care Response to Cardiovascular Disease in Pregnancy and Postpartum toolkit algorithm (Fig. 1). Use of this algorithm could have identified individuals as high risk requiring further cardiac evaluation and referral in 88% of maternal deaths (50). Patients with concerning symptoms or signs of cardiovascular disease should undergo consultation with a Pregnancy Heart Team (Table 4).

Risk Assessment of the Pregnant or Postpartum Patient With Known Cardiovascular Disease

Risk assessment can be accomplished using one of the several available risk stratification models, such as the Canadian Cardiac Disease in Pregnancy risk index (CARPREG II) (a comprehensive scoring system that incorporates general cardiac factors, specific cardiac lesions, and process of care factors), the Zwangerschap bij Aangeboren HARtAfwijkingen (ZAHARA) (a weighted risk score for congenital heart disease patients), and the modified World Health Organization (WHO) classification of maternal cardiovascular risk (54–56, 59). Among these, the modified WHO

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Drug	Teratogenic	Fetal Effects	Breastfeeding
Inotropic Agents			
Dopamine	No	No adverse fetal effects	Probably compatible, may inhibit prolactin release
Dobutamine	No	No adverse fetal effects	Probably compatible
Epinephrine	No	No adverse fetal effects when used acutely	Probably compatible
Vasodilators		-	
Nitroprusside	No	Potential for fetal cyanide toxicity with high doses	Possibly hazardous
Hydralazine	No	Relatively safe for the fetus	Probably compatible
Nitroglycerin	No	No adverse fetal effects	Possibly hazardous
		Observe for risks of methemoglobinemia	
Ephedrine sulfate	No	No adverse fetal effects when used acutely	Possibly hazardous with chronic use
Angiotensin Converting En	zyme Inhibitors and Ai	ngiotensin Receptor Blockers	
	Yes	Contraindicated Associated with fetal renal failure, growth restriction, malformations and death	Probably compatible No published information
Beta-blockers			
Propranolol	No	May increase risk of growth restriction	Probably compatible
Labetalol	No	No adverse fetal effects	Probably compatible
Atenolol	No	May increase risk of growth restriction	Probably compatible Limited information
Metoprolol	No	May increase risk of growth restriction	Probably compatible
Esmolol	No	May cause beta blockage in fetus	Probably compatible No published information
Carvedilol	Limited Information	May increase risk of growth restriction	Probably compatible No published information
Calcium Channel Blockers			
Verapamil	No	No adverse fetal effects	Probably compatible
Nifedipine	No	No adverse fetal effects	Probably compatible
Diltiazem	No	No adverse fetal effects	Probably compatible Limited information
Amlodipine	No	No adverse fetal effects	Probably compatible
·		Limited human information, animal data suggest risk	Limited information
Antiarrhythmic Agents		55	
Lidocaine	No	No adverse fetal effects	Probably compatible
Procainamide	No	Limited human information	Probably compatible
	Limited human information		Limited information
Phenytoin	Yes	Potential for early hemorrhagic disease of the newborn	Probably compatible

Table 5. Cardiac Medications With Potential Pregnancy and Lactation Influence

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(continued)

Drug	Teratogenic	Fetal Effects	Breastfeeding
Amiodarone	No	May be associated with fetal thyroid toxicity	Hazardous
Flecainide	Yes	Limited human information	Probably compatible
	Limited human information		Limited information
Sotalol	No	Human data suggest fetal risk	Possibly hazardous
	Limited human information		
AV Node Blocking Agents			
Adenosine	No Information	No adverse fetal effects	Probably compatible No published information
Digoxin	No	No adverse fetal effects	Probably Compatible
Anticoagulants and Anti-Thro	ombotics		
Warfarin	Yes	Risk of fetal hemorrhage	Probably compatible
Low-molecular-weight heparin	No	No adverse fetal effects Does not cross placenta	Probably compatible
Unfractionated heparin	No	No adverse fetal effects Does not cross placenta	Probably compatible
Clopidogrel	No	Limited human information	Probably compatible
	Limited human information		No published information
Direct Factor Xa Inhibitors (I	rivaroxaban or apix	aban)	
	No	Product labeling warns about abnormal bleeding risk	Possibly hazardous
		Crosses placenta	No published information
Diuretics			
Hydrochlorothiazide	No	No adverse fetal effects	Probably compatible
Furosemide	No	No adverse fetal effects	Probably compatible No published information

Table 5. Cardiac Medications With Potential Pregnancy and Lactation Influence (continued)

* For additional information on an individual medication's risk with breastfeeding, see https://toxnet.nlm.nih.gov/lactmed.htm. Data from Hale TW. Hale's medications and mothers' milk: a manual of lactational pharmacology. 18th ed. New York (NY): Springer; 2019 and Briggs GG, Freeman RK, Towers CV, Forinash AB. Drugs in pregnancy and lactation. 11th ed. Philadelphia (PA): Wolters Kluwer; 2017.

risk assessment model is most widely accepted and validated in pregnant women with known cardiovascular disease (Table 3). The modified WHO pregnancy risk classification stratifies cardiovascular disease into 5 groups and informs the health care provider of the frequency of cardiology evaluation recommended. All pregnant and postpartum women with known or suspected cardiovascular disease should proceed with further evaluation by a Pregnancy Heart Team (Table 4) consisting of a cardiologist and maternal-fetal medicine subspecialist, or both, and other subspecialists as necessary. The goal is to

establish a multidisciplinary comprehensive plan of care for the pregnancy, delivery, and postpartum periods. A mechanism for local, regional, and high-level facility referral should be in place for all labor and delivery units, particularly those with limited resources, in the event the need for consultation or emergency transfer arises. Referral to a hospital setting that represents an appropriate maternal level of care dependent upon the specific cardiac lesion (Table 3) is recommended for all pregnant patients with moderate- to high-risk cardiac conditions (modified WHO risk classes III and IV) because outcomes are

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significantly better for women in these facilities (8, 60). Complex congenital heart disease patients should be managed, to the extent possible, at advanced care centers with congenital heart disease expertise.

What are the indicated tests and how should these tests be interpreted for the pregnant patient with possible heart disease?

Testing of maternal cardiac status is warranted during pregnancy or postpartum in women who present with symptoms such as shortness of breath, chest pain, or palpitations and known cardiovascular disease whether symptomatic or asymptomatic, or both. Factors linked to cardiovascular disease, such as family history and underlying medical conditions, play an important role in assessing the risk of cardiovascular disease (Fig. 1). The type of testing and urgency of evaluation depends on the underlying cardiac condition and symptoms at the time of presentation (Table 2; Fig. 1).

Natriuretic Peptides

Brain natriuretic peptide (BNP) and N-terminal probrain natriuretic peptide (NT-proBNP) are natriuretic peptides (referred to collectively as BNP in this document). Elevated levels can be suggestive of heart failure. Although BNP reference ranges vary among laboratories, assays, age, gender, and BMI, in general a BNP level of greater than 100 pg/mL and an NTproBNP level greater than 450 pg/mL suggest the diagnosis of heart failure in nonpregnant patients (61). Brain natriuretic peptide levels in healthy women increase twofold during pregnancy (62) with a further increase early after delivery, (63) but values remain within normal range. Levels of BNP increase significantly in pregnant women with shortness of breath related to heart failure from left ventricular systolic dysfunction, (64) diastolic dysfunction, (65) and hypertensive disorders, including preeclampsia. (66)

Natriuretic peptides should be measured in the presence of new clinical symptoms or suggestive signs of heart failure to prevent delayed diagnosis. It may be helpful to obtain a baseline BNP level during pregnancy in women at high risk of or with known heart disease, such as dilated cardiomyopathy and congenital heart disease (Fig. 1). Serial determinations of BNP levels throughout each trimester and in the early postpartum period may assist in clinical decision making. Normal or low BNP levels are useful in excluding cardiac decompensation during pregnancy (67–69), and increasing BNP levels from the second trimester of pregnancy appear to predict adverse events (67, 70).

Cardiac Troponin I, Troponin T, and "High-Sensitivity" Troponin

Cardiac troponin I, troponin T, and "high-sensitivity" troponin are specific and sensitive biomarkers of myocardial injury (71). The diagnosis of acute coronary syndrome associated with pregnancy is similar to that in the general adult population, including comparable symptoms, electrocardiogram abnormalities, and elevations in biomarkers such as troponin (72). All pregnant and postpartum patients with chest pain should undergo standard troponin testing and an electrocardiogram to evaluate for acute coronary syndrome. Cardiology consultation should be obtained as clinically indicated. It should be noted that troponin I may be mildly elevated in the early postpartum period (73) in women with preeclampsia with severe features and in other noncardiac conditions, such as acute pulmonary embolisms or chronic renal disease (74).

Electrocardiogram

An electrocardiogram should be performed in pregnant women presenting with chest pain, shortness of breath, or palpitations to assess for features of ischemia, infarction, or arrhythmias. Normal pregnancy-related physiologic changes in maternal heart rate and chest wall shape cause benign nonpathologic electrocardiogram changes (75). Nonspecific ST-wave and T-wave abnormalities are found in up to 14% of pregnancies, usually occur in the left precordial leads, resolve after delivery, and may recur with subsequent pregnancies. Any rhythm abnormalities noted on electrocardiogram should prompt further evaluation.

Chest Radiograph

A chest radiograph with abdominal shield (76) should be considered as an important early test in pregnant or postpartum women presenting with shortness of breath to evaluate cardiac or pulmonary etiology.

Echocardiogram

An echocardiogram should be performed in pregnant or postpartum women with known or suspected congenital heart disease (including presumed corrected cardiac malformations), valvular and aortic disease, cardiomyopathies, and those with a history of exposure to cardiotoxic chemotherapy (eg, doxorubicin hydrochloride). Women with pulmonary hypertension or unexplained oxygen desaturation should have an echocardiogram before pregnancy, when pregnancy is confirmed, and during and after pregnancy. If there is doubt about the etiology as well as presence and severity of pulmonary hypertension, cardiac catheterization should be performed (52). The frequency of clinical and echocardiographic follow-up during pregnancy and postpartum is individualized. Cardiac chamber enlargement,

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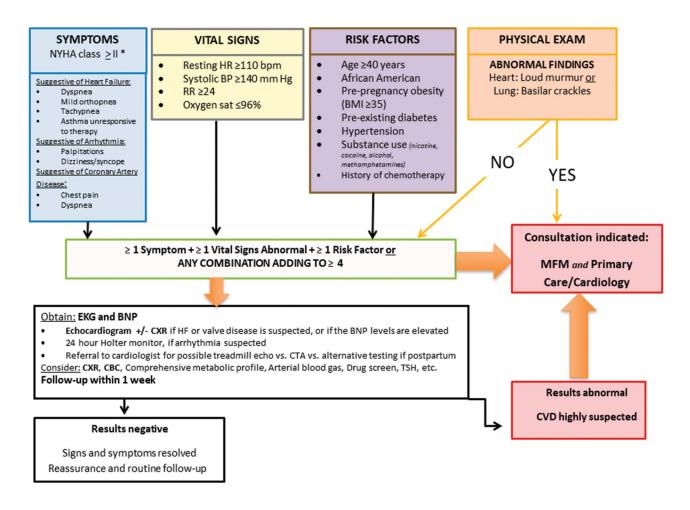


Figure 1. Cardiovascular Disease Assessment in Pregnant and Postpartum Women. *The NYHA Functional Classification is available at http://www.heart.org/HEARTORG/Conditions/HeartFailure/AboutHeartFailure/Classes-of-Heart-Failure_UCM_306328_ Article.jsp. Abbreviations: BMI, body mass index; BNP, brain natriuretic peptide; BP, blood pressure; CBC, complete blood count; CVD, cardiovascular disease; CXR, chest x-ray; EKG, electrocardiogram; HR, heart rate; MFM, maternal–fetal medicine; TSH, thyroid stimulating hormone; NYHA, New York Heart Association; RR, respiratory rate. (Modified from California Department of Public Health, 2017; supported by Title V funds. Developed in partnership with California Maternal Quality Care Collaborative Cardiovascular Disease in Pregnancy and Postpartum Taskforce. Visit www.CMQCC.org for details.)

concentric cardiac remodeling, diastolic dysfunction, valvular annular dilatation with regurgitation, and small asymptomatic pericardial effusion are frequent normal echocardiogram findings during late gestation. (41, 77–79)

Exercise Stress Test

An exercise stress test is an important predictor of a woman's ability to tolerate pregnancy. An exercise stress test provides an objective assessment of maternal functional capacity and facilitates the identification of exercise-induced arrhythmias (52). An exercise stress test should be performed in patients with known heart disease who plan pregnancy (80). International guidelines recommend submaximal exercise testing (80% of predicted maximal heart rate) in asymptomatic patients with suspected heart disease if already pregnant (80).

Computed Tomography

Computed tomography should be performed in pregnant or postpartum women presenting with chest pain when pulmonary embolism or acute aortic dissection is suspected. Iodinated contrast materials are not teratogenic or carcinogenic but cross the placenta and can produce transient depressive effects on the developing fetal thyroid gland. It is recommended that contrast agents be used only when absolutely required to obtain additional diagnostic information that will affect care. Less than 1% of iodinated contrast administered to a lactating woman is excreted into breast milk and absorbed through the infant's gastrointestinal tract. Therefore, breastfeeding can be continued without interruption after administration of iodinated contrast (81).

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Magnetic Resonance Imaging

Magnetic resonance imaging is used rarely in the urgent or emergent evaluation of cardiovascular concerns during pregnancy because imaging is less available and is more time consuming than computerized tomography. However, it is the preferred imaging modality in pregnant women to assess aortic dimension and for assessment of ventricular function and wall motion when echocardiography is nondiagnostic. When elective cross-sectional imaging is needed during pregnancy, a discussion with a cardiac imaging specialist to assist with choosing the most appropriate study and protocol is recommended to evaluate the patient optimally. There are no reported adverse maternal or fetal effects from magnetic resonance imaging during pregnancy (82). Reference values for cardiac magnetic resonance imaging indices during normal pregnancy and the postpartum state have been reported (40). Gadolinium, the contrast agent used for magnetic resonance imaging, should be limited in pregnant patients. It may be used as a contrast agent only if it significantly improves diagnostic performance and is expected to improve fetal or maternal outcome. Breastfeeding should not be interrupted after gadolinium contrast is administered (81).

Holter Monitor or Prolonged Cardiac Monitoring Device

A Holter monitor (24-hour to 48-hour ambulatory electrocardiogram monitoring) or a prolonged cardiac monitoring device (such as wireless patch cardiac monitor) is helpful for assessing symptoms of palpitations, lightheadedness, and syncope during pregnancy (83).

D-dimer

D-dimer is not recommended as part of routine evaluation of cardiac disease in pregnancy or the postpartum period (44).

Which types of preexisting maternal cardiac disease have the greatest effect on pregnancy and the postpartum period?

Evidence of underlying or overt cardiovascular disease can present initially either during pregnancy or in the first days, weeks, and months postpartum. Women with any high-risk cardiovascular disease, such as pulmonary hypertension, congenital heart disease, noncongenital valvular disease, dilated hypertrophic or peripartum cardiomyopathy, aortic disorders, or coronary artery disease should be monitored during pregnancy and the postpartum period by a cardiologist with expertise in the management of such patients or a Pregnancy Heart Team (Table 4) if institutionally available. A plan for management during pregnancy, labor, and postpartum should be decided and recorded in the medical and prenatal records.

Pulmonary Arterial Hypertension

Pulmonary arterial hypertension is defined as a mean pulmonary arterial pressure more than 25 mm Hg at rest. It can be either idiopathic or caused by various disorders. Pulmonary arterial hypertension carries an increased risk of maternal mortality, reported to range from 9% to 28% (84-86). Despite improved prognosis in women with pulmonary arterial hypertension, lowrisk patients might not be identified easily. Therefore, all women with severe pulmonary arterial hypertension should be advised against pregnancy. Health professionals caring for women with pulmonary arterial hypertension should ensure that women who are at risk of pregnancy understand these hazards and receive effective contraception. Induced abortion should be discussed if pregnancy occurs (80, 87). If a woman with severe pulmonary hypertension elects to proceed with or continue pregnancy, medical therapy for pulmonary hypertension can be initiated or modified during pregnancy (Table 5).

Congenital Heart Disease

Congenital heart disease encompasses multiple cardiac structural lesions. Many patients with congenital heart disease require additional specialized care while pregnant. Regular follow-up is required, the frequency of which depends on the type of the disease and the patient response to pregnancy (Table 3). Patients with high-risk lesions, such as those associated with pulmonary hypertension (eg, Eisenmenger syndrome), severe left-sided heart obstruction, severe ventricular dysfunction, cyanosis, failing Fontan circulation, and lesions associated with complex arrhythmias are counseled to avoid pregnancy or to proceed with surgical correction before pregnancy to allow for a lower-risk future pregnancy. The implications of maternal congenital heart disease on the fetus, including potential inheritance, should be discussed. In addition, certain genetic disorders are associated with congenital heart disease (eg, Noonan syndrome, Down syndrome, Holt-Oram syndrome, 22q11 microdeletion) and, therefore, prepregnancy genetic consultation and testing is recommended. Congenital heart disease in the woman should prompt fetal echocardiography, and conversely, identification of congenital heart disease in a fetus or neonate may prompt screening for parental congenital heart disease.

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Noncongenital Valvular Disease

Noncongenital valvular disease, (examples include rheumatic valvular disease, mitral valve prolapse, bioprosthetic valve prosthesis, or valve disease related to infective endocarditis), requires specialized evaluation. A transthoracic echocardiogram and an exercise stress test generally are recommended for patients with moderate-to-severe valve disease (such as valve stenosis or severe regurgitation), associated ventricular dysfunction, or pulmonary hypertension. Women with asymptomatic valve disease should be monitored by a cardiologist and may require additional testing or care during pregnancy. The frequency of monitoring necessary is indicated in the patient's modified WHO classification (Table 3). Ideally, symptomatic severe valve disease should be treated before pregnancy.

Mechanical Valve Prostheses

During pregnancy, mechanical valve prostheses and some cardiac lesions require therapeutic anticoagulation, which carries an increased risk for the woman and fetus. A detailed discussion about anticoagulation options and risks, frequency, and type of monitoring is best performed and documented before pregnancy. Regular monitoring and medication adjustment to confirm therapeutic levels is required (80, 88, 89). All pregnant patients with mechanical and bioprosthetic valves should be maintained on daily low-dose (81 mg) aspirin during pregnancy (90). Endocarditis prophylaxis should be administered around the time of delivery in high-risk patients (see "Intrapartum Management Principles") (88, 91).

Preexisting Dilated Cardiomyopathy

Prepregnancy assessment will include a baseline BNP level, transthoracic echocardiogram to assess ejection fraction, and hemodynamics, as well as an exercise stress test to assess functional capacity. The cause of the cardiomyopathy should be evaluated. Prepregnancy genetic consultation is recommended for patients with familial dilated cardiomyopathy. Cardiomyopathy related to prior unrecognized peripartum cardiomyopathy also should be considered. Women with preexisting dilated cardiomyopathy have a high rate (25-40%) of major adverse cardiovascular events, mainly heart failure, during pregnancy (92, 93). Patients should be counseled to avoid pregnancy or consider induced abortion if they have severe heart disease, including an ejection fraction less than 30% or class III/IV heart failure, severe valvular stenosis, Marfan syndrome with aortic diameter more than 45 mm, bicuspid aortic valve with aortic diameter more than 50 mm, or pulmonary arterial hypertension (Table 3) (80). Furthermore, women with ejection fractions between 30% and 45% also should be counseled regarding an increased risk of adverse cardiac events during pregnancy, such as heart failure or arrhythmia (94). Once pregnancy occurs, medication changes (Table 5) and follow-up frequency are dependent on cardiac and functional status.

Hypertrophic Cardiomyopathy

Hypertrophic cardiomyopathy is the most common genetic cardiac disease, with a prevalence of 2%. An analysis of pregnancy outcomes in such patients reported that cardiovascular complications are common and can be predicted by prepregnancy status, facilitating prepregnancy counseling and targeted antenatal care (95). Prepregnancy cardiovascular and genetic consultations are recommended for patients with hypertrophic cardiomyopathy.

Aortic Aneurysmal Disease and Dissection

Aortic aneurysmal disease and dissection in women of childbearing age generally are triggered genetically and are familial, syndromic, congenital, or inflammatory. Before pregnancy, a thorough cardiovascular specialty consultation to assess the cause, size, and location of the aneurysm is recommended. This consultation should include imaging with echocardiography and either computerized tomography or magnetic resonance imaging to evaluate the entire aorta. Although most dissections in young patients occur in the ascending aorta, the descending thoracic or abdominal aorta also can be affected. The cause, location, and size of the aortic aneurysm will influence counseling before and management during pregnancy. For example, all patients with vascular Ehlers-Danlos syndrome are advised to avoid pregnancy. The risk of aortic dissection associated with these conditions is increased during pregnancy and postpartum because of hormonal and hemodynamic changes on the aorta. No aortic dimension guarantees a safe pregnancy in a patient with aortopathy. The aortic size threshold for intervention before pregnancy depends on the cause of aortic aneurysmal disease (Table 6) (6, 80, 96). Even after ascending aorta replacement, aortic dissection can affect the remaining native aorta, so patients with prior operative intervention also should be monitored closely. During pregnancy, patients with aortic aneurysmal disease often are treated with beta-blocker therapy and should be seen regularly with repeat aortic imaging. The frequency of follow-up and imaging depends on the underlying disorder and aortic aneurysm location and dimension (Table 6). Surgical or percutaneous intervention for aortic aneurysm or dissection during

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pregnancy or postpartum rarely is needed and should occur only for an aortic emergency. Type and timing of invasive maternal interventions and the preferred mode of delivery should be made by the Pregnancy Heart Team (Table 4).

Atrial Arrhythmias

Atrial arrhythmias that cause palpitations are a common indication for cardiac evaluation during pregnancy. Any pregnant woman who presents with an arrhythmia should undergo evaluation to assess the cause and the possibility of underlying structural heart disease. The most common arrhythmias during pregnancy are premature atrial beats and paroxysmal supraventricular tachycardia, usually atrioventricular-nodal reentrant tachycardia that can be successfully treated with medication. Atrial fibrillation and flutter during pregnancy often occur in women with structural heart disease. Management is individualized depending on the effect of the arrhythmia and the presence of underlying cardiac disease (55).

Ventricular Arrhythmias

Ventricular arrhythmias are rarely encountered during pregnancy. If detected, a search for a cause and underlying structural heart disease is appropriate. The most common type of ventricular tachycardia that occurs in the absence of structural heart disease is right ventricular outflow tract ventricular tachycardia. This form of ventricular tachycardia initially may be identified during pregnancy because it is catecholamine sensitive, and it often can be treated successfully with beta-blockers or verapamil. Women with the long QT syndrome are at risk of ventricular tachycardia, especially in the postpartum period. Treatment with beta-blocker therapy throughout pregnancy and postpartum is appropriate. Acute treatment of sustained ventricular arrhythmias in pregnant women is similar to that in nonpregnant women. In women with structural heart disease and ventricular tachycardia, the risk versus benefit of antiarrhythmic drug therapy, an implantable cardioverterdefibrillator, and ablation should be reviewed with a Pregnancy Heart Team (Table 4) in conjunction with an electrophysiologist with expertise in managing patients with arrhythmias during pregnancy (80, 97).

How should women at high risk of peripartum cardiomyopathy be identified, assessed, and managed?

Peripartum cardiomyopathy occurs in 25–100 per 100,000 live births in the United States (98). It is characterized as a nonischemic cardiomyopathy presenting late in pregnancy or the first few months postpartum (99, 100) with a decrease in the left ventricular ejection fraction to less than 45% and no previous history of

Table 6. Management Strategies in Pregnant Women With Aortopathy

Surveillance Frequency	Suggested Mode of Delivery	
Normal-sized aorta Each trimester		
4–6 weeks	Vaginal	
4 weeks	Cesarean	
Ascending aorta 40–45 mm 4 weeks Ascending aorta >45 mm Prophylactic aortic surgery before or during pregnancy for rapid growth		
Surveillance Frequency	Suggested Mode of Delivery	
4–6 weeks	Vaginal	
4 weeks	Cesarean	
Aortic surgery before or during pregnancy for rapid growth	Cesarean	
	Each trimester 4–6 weeks 4 weeks Prophylactic aortic surgery before or during pregnancy for rapid growth Surveillance Frequency 4–6 weeks 4 weeks Aortic surgery before or during pregnancy	

Modified from Elkayam U, Goland S, Pieper PG, Silversides CK. High-risk cardiac disease in pregnancy: part II. J Am Coll Cardiol 2016;68:502–16.

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cardiac disease. The etiology remains uncertain. Although an autoimmune pathogenesis has been postulated (101), recent work has focused on vascular (102) and genetic etiologies (103).

Most women eventually recover myocardial function. For the remainder, chronic cardiomyopathy and heart failure persist. The overall rate of death or cardiac transplantation for women presenting with peripartum cardiomyopathy is 5–10% by 1 year postpartum (104, 105). Peripartum cardiomyopathy disproportionately affects non-Hispanic black women as evidenced by an increased incidence (106) and a lower rate of complete myocardial recovery (104, 107–110). Other risk factors for peripartum cardiomyopathy include increased maternal age, multifetal pregnancies, gestational hypertension, and preeclampsia. Women with a history of peripartum cardiomyopathy have a risk as high as 20% of experiencing a recurrence during subsequent pregnancies (111–113).

Pregnant or postpartum women who present with shortness of breath, chest discomfort, palpitations, arrhythmias, or fluid retention should be evaluated for peripartum cardiomyopathy. An echocardiogram is generally the most important diagnostic test. This evaluation also applies to women who are thought to have a hypertensive disorder of pregnancy. Consultation with a cardiologist is recommended to assist in management of peripartum cardiomyopathy, and referral to an appropriate level facility should be considered to allow multidisciplinary care by a Pregnancy Heart Team (Table 4). Medical management of peripartum cardiomyopathy follows the same general principles as management of heart failure with a reduced ejection fraction. Treatment with bromocriptine to improve myocardial recovery in peripartum cardiomyopathy remains investigational and requires further study (98, 114, 115). Breastfeeding should not be discouraged in women with peripartum cardiomyopathy because there are no data to suggest it negatively affects maternal cardiac status.

For women with peripartum cardiomyopathy who are pregnant at the time of peripartum cardiomyopathy diagnosis, timing and mode of delivery should be individualized, weighing the maternal risks of continuing pregnancy against the perinatal morbidity and mortality associated with preterm birth, and documented by a Pregnancy Heart Team (Table 4). Women presenting with shock (hypotension, tachycardia, or end-organ compromise) should be transferred to an appropriate level facility for consideration of a ventricular assist device support and transplant options. Vaginal delivery is a reasonable consideration for many women with peripartum cardiomyopathy because vaginal delivery results in less maternal morbidity and improved neonatal outcomes (116). Predicted outcomes of women with peripartum cardiomyopathy can be stratified by the severity of left ventricular dysfunction at presentation because women with a lower left ventricular ejection fraction have poorer outcomes (117). In the North American Registry Investigations of Pregnancy-Associated Cardiomyopathy (104), women with an initial ejection fraction less than 30% had less myocardial recovery and higher rates of left ventricular assist device implantation, cardiac transplantation, and death. In contrast, nearly 90% of women with an initial ejection fraction of more than 30% had complete myocardial recovery.

How should acute coronary events, including maternal cardiac arrest, be managed during pregnancy?

Acute Myocardial Infarction and Acute Coronary Syndrome

Ischemic heart disease complicates 8 per 100,000 hospitalizations for pregnancy and postpartum care (118). Maternal death occurs in 5–11% of affected patients with the highest risk in the peripartum period, a rate that is 3–4 times more than that of nonpregnant agematched women (17, 119).

Acute coronary syndrome implies suspicion of myocardial oxygen deprivation culminating in myocardial injury and necrosis. The spectrum of myocardial ischemia includes stable angina, unstable angina, and myocardial infarction. Increased cardiac output, enhanced stroke volume, and hypercoagulability favor the development or unmasking of underlying coronary artery disease. Risk factors for acute coronary syndrome during pregnancy (120) include traditional and pregnancy-specific features (see Box 1).

Box 1. Risk Factors for Acute Coronary Syndrome During Pregnancy

- Maternal age more than 30 years
- Non-Hispanic black race
- · Elevated body mass index
- Diabetes mellitus
- Tobacco use
- Hyperlipidemia
- Strong family history of cardiovascular disease
- · Hypertensive disorders of pregnancy
- · History of coronary artery dissection
- Blood transfusion
- Peripartum infection

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Acute coronary syndrome can be caused by coronary atherosclerosis, dissection, embolism, spasm, arteritis, and coronary artery occlusion related to aortic dissection. The differential diagnosis also should include takotsubo (stress) cardiomyopathy (119, 120). Coronary artery dissection is the most common cause of pregnancy-associated acute coronary syndrome and, although it can happen at any time during pregnancy, typically occurs in the early postpartum period (119, 121, 122). Coronary angiography remains the standard for diagnosis in patients with ST-segment elevation myocardial infarction. The noninvasive approach, however, is preferred in stable patients with preserved global left ventricular function because of the risk of complications, such as iatrogenic coronary dissection associated with coronary angiography and other interventions (119, 122, 123).

Every pregnant or postpartum patient with chest pain or cardiac symptoms should have consideration of acute coronary syndrome. Patients who have an acute coronary syndrome can present with typical (chest pain or shortness of breath) or atypical (vomiting, reflux, or diaphoresis) symptoms that mimic physiological changes of pregnancy or a pregnancy-related condition such as preeclampsia, or both. Some patients present with hemodynamic compromise, arrhythmia, or cardiogenic shock. Elevated troponins have sensitivity and specificity for myocardial damage. Electrocardiographic changes revealing STsegment elevations or depression are pathological and suggest acute myocardial infarction or ischemia. The differential diagnosis includes pericarditis, pulmonary embolism, and electrolyte abnormalities.

Acute coronary syndrome during pregnancy is best managed by a medical team such as a Pregnancy Heart Team (Table 4). Management of the maternal condition should receive priority. While maternal evaluation and initial therapy are proceeding, an unstable patient should be placed in a left lateral tilt ranging from 30-90 degrees. Fetal monitoring and corticosteroids to enhance fetal lung maturation are recommended for appropriate gestational ages. Initial medical management usually includes oxygen supplementation, nitrates, aspirin, intravenous unfractionated heparin, and betablocker therapy. If symptoms persist, coronary angiography is the preferred test and should be performed without delay. The type of intervention should be individualized based on the etiology of acute coronary syndrome, patient characteristics, and facilities available at the presenting medical center. The goal is to restore coronary blood flow promptly to accomplish tissue reperfusion, which is best accomplished by percutaneous coronary intervention if the cause is atherosclerotic coronary disease. The results of percutaneous coronary intervention in women with coronary dissection are, however, suboptimal and associated with high risk of propagation of the existing dissection. For this reason, a conservative approach is recommended in stable patients with coronary artery dissection (123).

When a patient with acute myocardial infarction presents to a medical center that does not have interventional cardiac catheterization facilities, options include emergent transfer to a center that has these capabilities or emergent thrombolysis in patients with ST-elevation myocardial infarction, or both, with subsequent planned transfer. Complications of maternal acute coronary syndrome include heart failure, cardiogenic shock, ventricular arrhythmias, recurrent myocardial infarction, and death. Data regarding timing and mode of delivery are limited.

Maternal Cardiac Arrest

Although maternal cardiac arrest occurs infrequently, the health care provider should be prepared to manage this situation in any health care facility (124). Maternal cardiac arrest etiologies include pregnancy-related and nonpregnancy-related conditions. The American Heart Association recommends the use of an alphabetical categorization for the differential diagnosis of maternal cardiac arrest that underscores the importance of a broadbased approach (125) (see Box 2).

Among the various etiologies for maternal cardiac arrest in patients admitted for delivery, hemorrhage is the most common (38.1%), followed by amniotic fluid embolism (13.3%) (126). Approximately 10% of pregnant or postpartum women with acute coronary syndrome and 4% with venous thromboembolism experience a maternal cardiac arrest (126).

An obstetric care provider is among the members of a multidisciplinary team that should be assembled immediately with the announcement of a facility alert "maternal code" (125). A health care facility that deals with obstetric patients should have 24-hour access to an experienced maternal code team. Management of cardiac arrest in the pregnant or postpartum patient requires familiarity with the physiologic adaptations of pregnancy that affect the execution of interventions dictated by basic and advanced cardiac life support. There are six key concepts to emphasize for the pregnant cardiac arrest patient:

1. Increased oxygen demand coupled with alteration in pharyngeal/laryngeal landmarks and a greater tendency toward aspiration upon loss of consciousness necessitate prioritization of bag mask ventilation with 100 percent oxygen and early intubation with a small endotracheal tube by an experienced health care provider (6–7 mm) (125).

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- 2. Aortocaval compression by a uterus larger than 20 weeks of gestation should be reduced with a one-handed or two-handed manual left uterine displacement maneuver very early in the resuscitation process while the patient remains in the full supine position on a backboard to maximize cardiac compression efforts (127, 128).
- 3. Simultaneous concurrent interventions are recommended in contrast to a sequential approach used in nonpregnant populations (128) (See Fig. 2).
- 4. Preparations for fetal delivery should be initiated in parallel with maternal resuscitative efforts.
- 5. Perform high-quality chest compressions on a backboard at a rate of 100–120 per minute using the same landmarks over the mid-lower sternum as left lateral uterine displacement is accomplished.
- Oxygenation remains a primary goal using a ratio of 30:2 chest compressions/ventilation efforts initially supplied by bag mask ventilation with 100% oxygen.

Otherwise intervention is similar to management of cardiac arrest in the nonpregnant state. Defibrillation pads are placed to enable rhythm analysis. Use of an automated external defibrillator may facilitate rhythm analysis when rescuers are less acquainted with this task. Use of an automated external defibrillator, however, does not obviate the requirement for resuscitation skill training (128). Although there is only a theoretical risk of electrocution from defibrillation, fetal monitors should be removed to allow maternal status to guide resuscitation interventions. Prompt biphasic defibrillation should be performed for appropriate shockable rhythms with reassessment of rhythm/pulse every 2 minutes, taking care to minimize interruptions in chest compressions. Although there can be a reluctance to use medications during pregnancy, the gravity of maternal cardiac arrest is such that medications should be used in resuscitation. Epinephrine is the vasopressor of choice and should be administered by intravenous or intraosseous access above the diaphragm. A timekeeper should keep the resuscitation team aware of the time that has transpired since cardiac arrest (125).

Perimortem Cesarean Delivery/ Resuscitative Hysterotomy

When initial interventions are unsuccessful, the American Heart Association recommends timely consideration of perimortem cesarean delivery or resuscitative hysterotomy (129) when the uterus is sized 20 weeks of gestation or more. Because achieving the shortest time from cardiac arrest to delivery clearly enhances maternal and neonatal outcomes, efforts should be made to facilitate delivery as

Box 2. Alphabetical Categorization for the Differential Diagnosis of Maternal Cardiac Arrest

- A (anesthetic complications, accidents)
- B (bleeding)
- C (cardiovascular disorders)
- D (drugs such as magnesium sulfate)
- E (embolism including venous thromboembolism and amniotic fluid embolism)
- F (fever including sepsis)
- G (general including metabolic and electrolyte)
- H (hypertensive disorders including stroke)

rapidly as possible from cardiac arrest, with the target to deliver within a 4-5-minute window. When return of spontaneous circulation is very unlikely, or arrest is unwitnessed, postponing delivery 4-5 minutes is not necessary (128, 130). Preparations to undertake resuscitative hysterotomy should begin immediately during the first minute of maternal cardiac arrest or apparent rapidly declining maternal cardiac function. Health care providers should be aware that there is no obvious threshold for either death or damage at 4 minutes. Instead there is a progressive decrease in the likelihood of injury-free survival for the woman and fetus with lengthening time since cardiac arrest (131). Survival curves for women and newborns have shown 50% injury-free survival rates with perimortem cesarean as late as 25 minutes after maternal cardiac arrest (131); therefore, delivery may be of benefit even if it does not occur within 4 minutes.

Ideally, perimortem cesarean delivery should occur at the site of the arrest because transport compromises cardiopulmonary resuscitation and also leads to further time delay (124). Initiation of perimortem cesarean delivery requires a scalpel, which usually is contained in the code cart's perimortem cesarean delivery kit (125). A vertical skin incision may be fastest to accomplish and provides more options for further exploratory surgery. If return of cardiac function has not occurred with perimortem cesarean delivery, alternatively open-chest direct cardiac massage can be attempted (128). Cardiopulmonary bypass and extracorporeal membrane oxygenation have been successfully employed for etiologies requiring timelimited cardiopulmonary support, such as local anesthetic drug toxicity, acute cardiac decompensation related peripartum cardiomyopathy, and acute respiratory distress syndrome (128).

The infrequency of maternal cardiac arrest underscores the need for regular team training and practice of

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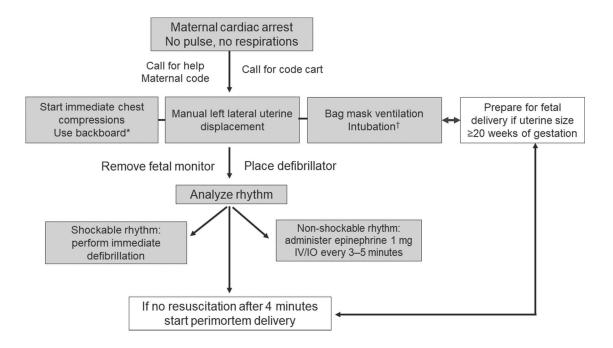


Figure 2. Maternal Resuscitation Algorithm. Abbreviations: IO, Intraosseous; IV, intravenous. *High-quality chest compressions on a backboard are performed at a rate of 100–120 per minute. [†]Prioritization of bag mask ventilation with 100 percent oxygen. Oxygenation remains a primary goal using a ratio of 30:2 (chest compressions/ventilation efforts). (Modified from Zelop CM, Einav S, Mhyre JM, Martin S. Cardiac arrest during pregnancy: ongoing clinical conundrum. Am J Obstet Gynecol 2018;219:52–61.)

resuscitation skills and scenarios through simulation training (128).

What are the general approaches to pregnancy management antepartum, intrapartum, and postpartum for the patient with cardiovascular disease?

Antepartum Management Principles

Pregnant women with cardiac disease should give birth at a hospital with the appropriate maternal level of care (60). The resources needed to minimize maternal and fetal complications should be anticipated, outlined, and documented before delivery. A comprehensive plan of care for the pregnancy, delivery, and postpartum periods should be available readily in the medical record and easily accessible to all health care providers involved with the woman's care. Women with complex congenital or noncongenital heart disease should be treated by a Pregnancy Heart Team (Table 4) (52, 80, 132) and should undergo comprehensive cardiac diagnostic evaluation as directed by the team and the diagnosis. In women with congenital heart disease, screening fetal echocardiogram is indicated at 18–22 weeks of gestation because the risk of congenital heart defect in the fetus is estimated at 4-10% (133, 134). Fetal growth assessment by either serial clinical examination or ultrasonography should be considered because fetal growth restriction occurs in many types of maternal congenital and acquired cardiac lesions (133, 135).

Women with chronic medical conditions, such as pregestational diabetes or chronic hypertension, can develop cardiac and other vascular complications of their disease (46, 47). Daily low-dose aspirin prophylaxis is recommended in women at high risk of preeclampsia and should be initiated between 12-28 weeks of gestation and continued until delivery. Similar prophylaxis should be considered for women with more than one of several moderate risk factors for preeclampsia (136). The precise blood pressure level at which antihypertensive therapy is indicated during pregnancy in women with cardiovascular disease continues to be debated. The use of blood pressure-lowering medications is recommended for secondary prevention of recurrent cardiovascular disease events in nonpregnant patients with clinical cardiovascular disease (defined as coronary heart disease, congestive heart failure, and stroke) and an average systolic blood pressure of 130 mm Hg or higher or an average diastolic blood pressure of 80 mm Hg or higher (137). Few clinical trials on this topic have been conducted in pregnancy and the evidence is limited (47). Prompt treatment of severe hypertension (systolic blood pressure more than 160 mm Hg and diastolic blood pressure more than

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110 mm Hg) is recommended to prevent complications (47, 138). Left ventricular hypertrophy with impairment of diastolic function may develop in the setting of long-term hypertension. This scenario may place the pregnant woman at risk of cardiogenic pulmonary edema due to the baseline volume increase in pregnancy and after intravenous fluid boluses. Pulmonary edema in the patient with preeclampsia may be cardiogenic or noncardiogenic in origin or a combination of both. Echocardiography can help differentiate between the two entities. An echocardiogram should be performed in any pregnant or post-partum patient with pulmonary edema possibly due to peripartum cardiomyopathy or preeclampsia.

In general, regular physical activity during pregnancy and postpartum improves or maintains physical fitness, helps with weight management, reduces the risk of gestational diabetes in obese women, and enhances psychologic well-being. During pregnancy complicated by cardiac disease, the woman should be carefully evaluated by a Pregnancy Heart Team (Table 4) before recommendations are made regarding physical activity participation (139) to ensure that a patient does not have a cardiac reason to avoid exercise.

Intrapartum Management Principles

A detailed delivery plan should be determined between 20-30 weeks of gestation and recorded in the medical record. An individualized plan through shared decision making with the patient and the Pregnancy Heart Team (Table 4) is recommended. This strategy should include management of induction, delivery, and postpartum concerns and a surveillance plan. Women with stable cardiac disease can undergo a vaginal delivery at 39 weeks of gestation, with cesarean delivery reserved for obstetric indications (140). Some patients with very high-risk cardiac conditions may not be able to tolerate the fluctuations in cardiac output or Valsalva efforts that occur during vaginal delivery. For many of these patients, regional anesthesia during labor may provide sufficient pain relief (thereby minimizing catecholamine release and resultant cardiac output fluctuations) to render a vaginal delivery feasible. A Pregnancy Heart Team (Table 4) should determine which patients are not candidates for vaginal delivery or require assisted second stage of labor during pregnancy. In the absence of spontaneous onset of labor or indicated delivery before term, scheduled induction of labor for pregnant women with cardiac disease between 39-40 weeks of gestation may be considered with input from the Pregnancy Heart Team.

Anticoagulation must be carefully reviewed and managed by the Pregnancy Heart Team during pregnancy and adjusted appropriately at the time of neuraxial anesthesia and delivery. For women who are receiving prophylactic low-molecular-weight heparin, discontinuation is recommended at least 12 hours before scheduled induction of labor or cesarean delivery. A 24-hour interval is recommended for patients on an adjusteddose regimen (44, 141, 142). For unfractionated heparin doses of 7,500 units subcutaneously twice a day or more, a 12-hour interval as well as evaluation of coagulation status with laboratory testing are recommended. Women receiving anticoagulation therapy may be converted from warfarin or low-molecular-weight heparin to the shorter half-life unfractionated heparin in anticipation of delivery, depending upon the institution's protocol. An alternative may be to stop anticoagulation and induce labor within 24 hours, if clinically appropriate. If conversion to unfractionated heparin is planned, timing should be based upon the likelihood of spontaneous labor with the goal of minimizing the time without anticoagulation coverage. This approach is especially important in a patient with a mechanical valve prosthesis (44, 88, 119).

The most common intrapartum cardiac complications include pulmonary edema or arrhythmias (54, 59, 133). These patients require a high level of surveillance and care. For women with a history of arrhythmias and for those who develop an arrhythmia during pregnancy, intrapartum cardiac monitoring is recommended. (52). Pulmonary edema usually can be prevented by maintaining a meticulous fluid balance. Expert consensus is that antibiotic prophylaxis administered at the time of delivery is reasonable for the subset of patients at increased risk of developing infective endocarditis, such as those with a history of previous infective endocarditis, and for patients at high risk of experiencing an adverse outcome from infective endocarditis (88, 91).

Obstetric Anesthesia Principles

Cardiac disease patients may require an elevated level of monitoring and anesthetic care for all obstetric procedures (eg, dilation and curettage or evacuation or cerclage) as well as vaginal or cesarean delivery. Consultation with an anesthesiologist should be performed antepartum for anesthetic, cardiac, and obstetric risk assessment and planning.

Under the direction of an anesthesiologist, cardiac disease patients undergoing vaginal delivery should be offered epidural labor analgesia, and cardiac disease patients undergoing cesarean delivery should have neuraxial anesthesia, if possible. Cardiovascular events (usually arrhythmia) are significantly decreased with epidural use (143). Exceptions for neuraxial anesthesia include the usual anesthetic contraindications and patients receiving pharmacologic anticoagulation as noted above (141, 142, 144). Consideration also should be

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given to modifying neuraxial anesthesia management for patients at risk of cardiovascular decompensation related to reduction of systemic vascular resistance. Such patients include those with left ventricular outflow tract obstruction or cyanotic congenital heart disease.

Immediate Postpartum Management Principles

The postpartum period is a time of heightened risk of cardiovascular disease-related maternal morbidity and mortality (80) as evidenced by a threefold increase in the rate of postpartum hospitalizations for chronic heart disease in the past decade (14). Among cardiovascular disease-related mortality, peripartum cardiomyopathy (25–100 per 100,000 live births) is identified as the leading (23%) cause of late postpartum death (10, 144). Aortic dissection and acute coronary syndromes typically are diagnosed in the early postpartum period and are associated with a high risk of maternal mortality (15, 145–147). The incidence of acute coronary syndrome is estimated at 2.7-8.1 per 100,000 deliveries, a rate known to be threefold to fourfold higher during the pregnancy and postpartum periods compared with nonpregnant women matched for age (15, 17, 118, 119, 148). Cardiac disease is particularly linked to late maternal death as long as 1 year postpartum (10).

Women with cardiac disease are at high risk of immediate complications during the early puerperium (first 7 days after delivery) and as long as 6 months postpartum (26). This risk is compounded by the common concurrence of immediate postpartum obstetric complications, such as hypertensive disorders, hemorrhage, and infection. An elevated level of care or a prolonged period of monitoring may be necessary, particularly for patients at risk of cardiogenic pulmonary edema and arrhythmias or in the setting of concurrent obstetric or surgical complications. Consideration should be given to careful and frequent monitoring of the signs and symptoms of cardiovascular disease (Table 2) using pulse oximetry, lung auscultation, the recording of fluid balance, and for the development of shortness of breath or cough. Cardiovascular testing may be appropriate and individualized to presenting features. Early consultation with a cardiologist and possible transfer of the patient to a facility with a higher level of care should be expedited if maternal complications related either to known disease or to new-onset, acquired maternal heart disease develop at any time during the course of care.

Each facility should review the available venous thromboembolism risk assessment protocols and adopt and implement one of them in a systematic way to reduce the incidence of venous thromboembolism in the post-

partum period (44). Cesarean delivery, particularly when complicated by postpartum hemorrhage or infection, as well as medical factors or pregnancy complications, increases the risk of venous thromboembolism. Although current evidence is insufficient to recommend universal adoption of pharmacologic prophylaxis for venous thromboembolism after cesarean delivery, for selected high-risk patients in whom significant risk factors persist after delivery, prophylaxis may be considered (44). If thromboprophylaxis is considered, evidence suggests that in women with a BMI of 35 or more, weight-based dosage (0.5 mg/ kg enoxaparin every 12 hours) compared to fixed dosage will achieve significantly higher anti-Xa concentrations within the adequate prophylaxis range (P < .01) (149, 150). However, the optimal dose, route, and duration of thromboprophylaxis need further evaluation. In the absence of clear, randomized controlled trial evidence, practitioners can rely on consensus-derived clinical practice guidelines or recommendations from national and international societies (44).

Pharmacologic Considerations

Health care providers should be aware of cardiac medications with obstetric implications (Table 5) as well as obstetric medications with cardiac implications (Table 7). Obstetrician–gynecologists and other health care providers should consult lactation pharmacology resources for current information on individual medications because inappropriate advice often can lead women to discontinue breastfeeding unnecessarily (151).

► How should in-hospital postpartum care be altered for women with or at risk of cardiovas-cular disease?

Postpartum Considerations After Delivery Hospitalization

Complications are frequently encountered in the days, weeks, and months after delivery in women with known cardiovascular disease and in those with latent cardiovascular disease. Women with multiple risk factors for cardiovascular disease (See Box 3) may be particularly at risk of manifesting symptoms for the first time during their postpartum course. A postpartum follow-up visit (early postpartum visit) with either the primary care provider or cardiologist is recommended within 7–10 days of delivery for women with hypertensive disorders or 7–14 days of delivery for women with heart disease/cardiovascular disorders. Ideally, future pregnancy intentions and commensurate contraceptive needs should be discussed before delivery or hospital discharge and reassessed at each postpartum visit.

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Drug	Cardiovascular Side Effects	Cardiac Conditions Contraindicated	Special Considerations
Corticosteroids (Betamethasone or Dexamethasone)	Fluid retention Electrolyte disturbance Hypertension	Use with caution in patients with heart failure or hypertension	Recent history of myocardial infarction; risk of left ventricular free wall rupture
Hydroxyprogesterone	Fluid retention Electrolyte disturbance Hypertension	Use with caution in patients with cardiac dysfunction	
Prostaglandin (PGE ₂)	None reported		
Misoprostol	Rare		
Oxytocin	Arrhythmias Hypotension		Titrate carefully and avoid rapid intravenous bolus
Magnesium Sulfate	Hypotension Vasodilation Syncope	Caution in patients with heart block	Titrate carefully in hypertrophic obstructive cardiomyopathy and stenotic valvular lesions especially aortic stenosis
Terbutaline	Tachycardia Hypotension Arrhythmias Myocardial ischemia	Hypertrophic obstructive cardiomyopathy Patients at risk of arrhythmias or ischemia Stenotic valvular lesions especially mitral stenosis	Do not use beyond 48–72 hours
Methylergonovine	Coronary artery vasospasm Hypertension Arrhythmias	Coronary artery disease or risk for ischemia Aortopathies	Do not give intravenously
Carboprost	Hypertension	Pulmonary hypertension	Can cause bronchospasm
Tromethamine	Palpitations Tachycardia Vasodepressor syncope Pulmonary hypertension	Cyanotic congenital heart disease Pulmonary edema	Do not give intravenously
Tranexamic Acid			Use with caution in uncorrected cardiovascular disease due to thrombosis

Table 7. Obstetric Medications With Cardiac Influences

Data from Facts & Comparisons. St. Louis (MO): Wolters Kluwer Health, Inc; 2019. Available at: http://fco.factsandcomparisons. com/lco/action/home. Retrieved January 22, 2019.

Optimal care for women with known cardiovascular disease during this critical period requires a team-based approach, such as with a Pregnancy Heart Team (23, 47, 138), and a cardiovascular disease risk assessment by a maternal care provider (Fig. 1). Mortality reviews indicate that cardiovascular disease signs and symptoms are not recognized readily by the patient, family, or the health care provider and that there are delays in access

to health care related to transportation or other financial barriers (10). All postpartum women with cardiovascular disease and those identified as at high risk of cardiovascular disease should be educated on their individual risk. They should be instructed when and how to seek medical care and be provided with phone numbers and a printed or electronic copy of their discharge summary, including an explanation of signs and symptoms that should

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prompt timely assessment. These women benefit from an early outpatient visit within 7–14 days after delivery to facilitate overall assessment of well-being and symptoms or functional status, or both. To facilitate patient adherence to appointments, it is important to address barriers to care, such as socioeconomic variability, insurance status, access to health care, and physical distance to the nearest hospital.

Contraceptive options, including immediate postpartum placement of long-acting reversible contraceptive methods, should be discussed in the prenatal period, and plans to execute should be implemented before hospital discharge to minimize the risk of short-interval recurrent pregnancy.

Breastfeeding has important short-term and long-term health benefits for the woman. Cardiac patients should be encouraged to breastfeed during the postpartum hospital stay and in the outpatient setting because most medications are considered safe (Table 5) (152). Breastfeeding has favorable effects not only on hypertension through positive effects on the maternal vasculature but fosters a favorable lipid and hormonal milieu along with improved mother-infant bonding (153). Women whose cumulative lifetime duration of breastfeeding is 6–12 months are 10% less likely to develop cardiovascular disease (154).

It is important to emphasize that the overwhelming majority of cardiovascular disease mortality occurs beyond the conventional postpartum period, including the first 42 days after delivery (10). Thus, a long-term care plan is crucial. Women identified as high risk (Fig. 1) should be evaluated at 3 months in a comprehensive cardiovascular postpartum visit. Payment models that provide health care coverage for the 3-month visit for these high-risk patients should be developed. This 3-month comprehensive cardiovascular postpartum visit with the Pregnancy Heart Team, the obstetrician-gynecologist, or other primary care provider should be individualized to each patient and should include a history of pertinent symptoms, a physical examination, an assessment of height and weight (BMI), waist circumference, heart rate, respiratory rate, blood pressure, and oxygen saturation. Laboratory testing, including fasting blood glucose or hemoglobin A_{1c}, and a complete lipid profile should be considered. Patients should have a yearly follow-up with their primary care physician. Health care providers should establish and maintain an ongoing partnership with a cardiologist or primary care physician, or both, who will be available for future care. Bundled payments for maternity care should be expanded to include this intensive classification (as many as three visits in the first 3 months postpartum) for a more individualized approach to these women. Ongoing collaborative care of the woman with cardiovascular disease or at risk of future cardiovascular disease is essential to reducing morbidity and mortality, optimizing the woman's health in preparation for future pregnancies, and promoting long-term cardiovascular health (26, 139).

What are the contraceptive options and considerations for women with heart or cardiovascular disease, or both?

Contraception Considerations

Decisions regarding the most appropriate contraceptive option for a woman require discussion of her future pregnancy desires and personal preferences, as well as critical assessment of the patient's underlying disease and the relative risks and benefits of the contraceptive option considered. The Centers for Disease Control and Prevention and the World Health Organization have established a four-tier scale related to medical eligibility criteria for contraceptive use that provides clinicians an assessment of the relative risks and benefits of contraceptive methods in various medical settings (155-157). Clinicians can access this detailed clinical guidance at https://www. cdc.gov/reproductivehealth/contraception/pdf/summarychart-us-medical-eligibility-criteria_508tagged.pdf. See also the American College of Obstetricians and Gynecologists' For More Information web page.

Intrauterine devices are the recommended nonpermanent option for women with high-risk cardiovascular conditions (155, 158). Intrauterine devices are highly effective and reliable long-acting reversible contraception. Multiple intrauterine device options (copper and progestin containing) are available based on patient preference, contraindications, and desire for future fertility. Annual failure rates with intrauterine devices use are less than 1%, and duration of action ranges from 3 to 10 years depending on the device used. Intrauterine device placement can be undertaken in the clinician's office and poses minimal risk for women with underlying cardiac disease (155, 158). Although expulsion rates are increased (10-27%) with placement at the time of delivery, immediate postpartum intrauterine device placement after delivery of the placenta is also a consideration for women with high-risk cardiac disease to ensure there is no gap in contraceptive protection (159). Women should be counseled about the increased expulsion risk as well as signs and symptoms of expulsion (159).

Progestin-only contraceptives (oral, depot medroxyprogesterone acetate injection, or implant) are potentially effective alternatives for women with cardiac disease. The progestin-only pill is limited primarily to use in the immediate postpartum period in lactating women. This option, however, has lower efficacy (more than 9% failure rate) for pregnancy prevention (155, 160, 161). Intramuscular depot medroxyprogesterone acetate is a highly

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Box 3. Risk Factors for Maternal Cardiovascular Disease

- Non-Hispanic black race
- Older age (more than 40 years)
- Obesity
- Hypertensive disorders of pregnancy (preeclampsia, eclampsia, or hemolysis, elevated liver enzymes, and low platelet count syndrome)
- Chronic disease (chronic hypertension or pregestational diabetes mellitus)
- Obstructive sleep apnea (moderate to severe)
- History of preterm delivery
- · Strong family history of heart disease
- Exposure to cardiotoxic drugs

effective contraceptive modality and appears to be a safe option for women with valvular heart disease, cardiomyopathy, and well-controlled hypertension (155, 162). For women receiving therapeutic anticoagulation, depot medroxyprogesterone acetate injections theoretically can increase risk of hematoma formation. Reversible bone loss, diminution of protective high-density lipoprotein, and increased triglycerides have been noted secondary to the hypoestrogenic effect of depot medroxyprogesterone acetate (163, 164). The progestin implant is highly efficacious and appears to be a safe option for most women with hypertension or known cardiac disease. Use in women with current or previous ischemic heart disease or cerebrovascular accident is limited secondary to increased concern for thrombosis (155). There also may be risk of hematoma formation at the time of insertion or removal, or both, in women who are anticoagulated.

Combined hormonal contraception (eg, oral, ring, or patch), although effective, may pose significant risk for women depending on the patient's underlying cardiac condition because of the estrogen component. The use of combined hormonal contraception in women with poorly controlled hypertension, aged more than 35 years, who are smokers, or who have migraine with aura, is associated with increased risks for exacerbation of high blood pressure, cardiovascular events, such as stroke and acute myocardial infarction, and thromboembolic events (155, 161, 162, 165-169). For women with valvular heart disease, especially those with complicated valvular pathology, combined hormonal contraception may increase the risk of arterial thrombosis and other adverse cardiovascular consequences. Use of combined hormonal contraception in the setting of cardiomyopathy can be associated with fluid retention, which can exacerbate heart failure (170). Because of these concerns, alternative contraceptive options should be considered in women with prothrombogenic states, uncontrolled hypertension, ischemic heart disease, and complicated valvular heart disease (155).

Barrier, fertility awareness-based, and other nonhormonal methods used to lessen the risk of fertilization, although safe, have high risk of contraceptive failure. Therefore, these methods are suboptimal for women who do not desire further childbearing or who have significant cardiovascular disease in which pregnancy is ill-advised or contraindicated. Estimated annual failure rates vary according to the method used. The fertility-awareness method has a failure rate of 24%; withdrawal, 22%; spermicide use, 28%; male condom, 18%; female condom, 21%; sponge, 12–24%; and diaphragm, 12% (155, 160, 171).

Emergency contraception is available for women with contraindications to use of combined hormonal contraception (155, 161). The presence of cardiovascular disease is not a contraindication to the use of emergency contraception (155, 161). Progestin-only emergency contraceptive methods are generally better tolerated and are more efficacious than combined regimens and may be preferred in the setting of cardiovascular disease. Insertion of a copper intrauterine device is an effective method of emergency contraception when inserted within 5 days after unprotected intercourse. The copper intrauterine device provides ongoing contraception and should be made available to patients at high risk of pregnancy morbidity and mortality (158).

Permanent sterilization is one of the most effective contraceptive options for reproductive-aged women who have completed childbearing, especially for women with high-risk cardiac conditions or cardiovascular disease. Paternal vasectomy is a highly effective approach for male sterilization with low complications and failure rates of less than 1% (155, 172, 173). Limitations of vasectomy include the potential for pregnancy in the setting of a nonmonogamous relationship or a sexual relationship with a new partner. Female sterilization may be performed by several approaches (eg, laparoscopy, minilaparotomy, and in combination with cesarean delivery) (172). Although laparoscopy is an effective and safe approach for sterilization, the need for general anesthesia and pneumoperitoneum (with resultant increased intraperitoneal pressure) can alter cardiac and pulmonary function and thereby impose challenges for women with certain critical cardiac abnormalities (174, 175). Low-pressure laparoscopy does not appear to mitigate these operative physiologic effects (176). Minilaparotomy with tubal ligation can be performed under regional anesthesia and may minimize intraoperative risks in women with cardiac disease (172).

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What are the long-term considerations and implications after pregnancy for women with cardiovascular disease?

There are immediate and long-term continuity of care considerations for women with congenital or acquired heart and cardiovascular disease. Specific and immediate considerations include the following:

- Ensure proper cardiology follow-up is initiated during pregnancy or postpartum.
- Acknowledge the effect of a chronic diagnosis and possible need for long-term medication use. Consider 3-month prescriptions (or longer) if clinically appropriate (177).
- Refer patients with cardiovascular disease to lactation services when breastfeeding presents challenges, which often arise because of preterm delivery (178).
- Be mindful of the mental health implications of cardiovascular disease during the postpartum period and beyond. Preterm birth also is associated with maternal depression, anxiety, and posttraumatic stress disorder (179). Of note, most medications used to treat these disorders are compatible with breastfeeding, even in conjunction with cardiac medications. Mobilize all available resources to support the patient and her family during this time as indicated.
- Discuss future pregnancy intentions and provide a commensurate form of contraception.
- Screen patients routinely at postpartum follow-up visits for depressive symptoms and evidence of posttraumatic stress disorder and refer to social services or psychologic services, or both, as indicated (179).

These are priorities early in the puerperium because many women lose health insurance beyond the first 42 days postpartum. These steps are especially relevant in the postpartum period when women with cardiovascular disease are focused on newborn care and are less likely to prioritize their own health.

Continuity of Care Considerations for Women With Cardiovascular Disease Risk Factors

Acute (gestational hypertension, preeclampsia) and chronic hypertensive disorders of pregnancy are important identifiers of patients at risk of cardiovascular disease (23). Gestational hypertension and preeclampsia increase the risk of future cardiovascular disease by severalfold, and the risk is even higher in women with recurrent preeclampsia, preterm birth at less than 37 weeks of gestation, or intrauterine growth restriction (29, 180–186). Not only do women with hypertensive disorders of pregnancy have a substantially higher risk of future cardiovascular disease, they also have a threefold to fourfold increase in the risk of chronic hypertension, a 4.2-fold increase in the risk of heart failure, an 81% increase in the risk of stroke, a 5-fold to 12-fold increased risk of developing end-stage renal disease, and double the risk of atrial arrhythmias, coronary heart disease, and mortality when compared with women with normotensive pregnancies (184, 187). Exposure to severe maternal preeclampsia is an independent risk factor for long-term cardiovascular morbidity in offspring born at term (188).

The presence of gestational complications reliably identifies women with underlying, often unrecognized, cardiovascular risk factors (189, 190). Because approximately 20% of women have one or more of these complications (191), risk screening is recommended (192) within the first year postpartum (191). Cardiovascular assessment and follow-up at 3 months postpartum is recommended for women with the following conditions:

- Hypertension, chronic/essential or hypertensive disorder of pregnancy (ie, gestational hypertension, preeclampsia, eclampsia, hemolysis, elevated liver enzymes, and low platelet syndrome, chronic hypertension [with or without superimposed preeclampsia])
- · Gestational diabetes mellitus
- Intrauterine fetal growth restriction (particularly less than the 5th percentile for gestational age or less than 2,500 g at term)
- Idiopathic preterm birth
- Placental abruption
- Obesity/excessive pregnancy weight gain/postpartum weight retention
- Sleep disorders/moderate-to-severe obstructive sleep apnea (193–197)
- Maternal age older than 40 years

Cardiovascular risk screening within 3 months postpartum includes a detailed medical history (including history of cardiovascular disease), postpartum medication monitoring (such as antihypertensive medication), a physical examination, and basic biochemical testing (see Box 4).

After cardiovascular screening is complete, women should be counseled with regard to their identified risk factors. The goal of targeted cardiovascular risk assessment and patient education is to promote patient selfawareness and self-initiation of preventive actions. The American Heart Association's Life's Simple 7 describes

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Box 4. Postpartum Cardiovascular Risk Screening

Medical history

- Smoking (number of cigarettes per day, number of years smoked)
- Physical activity (times per week, duration)
- Breast feeding (how long)
- History of hypertension, diabetes, or cardiovascular disease
- First degree family history of cardiovascular disease, hypertension, or diabetes

Physical examination

- · Resting blood pressure and heart rate
- Body mass index and waist circumference

Biochemical testing

- Cholesterol/lipid profile
- Fasting glucose (or oral glucose tolerance testing if patient had gestational diabetes)
- Urine protein assessment (protein:creatinine ratio)

Nutrition assessment

seven steps to achieve a healthy lifestyle (198). Tests for borderline or elevated blood pressure or lipid abnormalities, or both, should be repeated after 6–12 months of lifestyle modification and, if persistently elevated, initiation of pharmacologic treatment should be considered.

Ongoing Postpartum Care After the 3-Month Cardiovascular Assessment Visit

Continuing follow-up as indicated after the 3-month comprehensive cardiovascular postpartum evaluation provides the opportunity for counseling, planning, and intervention to optimize underlying medical conditions to improve future pregnancy outcomes and cardiovascular health. If not already managed, contraceptive needs can be considered, managed, or modified as needed. In addition to the usual prepregnancy topics such as folic acid usage, restoration to prepregnancy weight should be emphasized because not achieving it increases the risk of future pregnancy complications (199). Weight management strategies include referral to a registered dietitian, peer support, improved access to opportunities for physical activity, and programs that provide child care at no or low cost. Women with pregnancy complications, such as preeclampsia and gestational diabetes, should be counseled regarding the risks of future cardiovascular disease and overt diabetes,

respectively. In any future pregnancy, patients with a history of prior preeclampsia should be considered for lowdose aspirin prophylaxis (136). For those who have previous gestational diabetes mellitus, early screening in the next pregnancy is recommended (200). Finally, given the benefits for the infant and the cardiometabolic benefits for the woman (201), breastfeeding should be recommended, and community support identified, to increase breastfeeding success after future pregnancies. During the postpartum period, health care providers may include a primary care provider and various other specialists, and communication across the clinical team should continue. However, because coordinated care can be challenging among many different specialists and subspecialists (202), the patient must be educated about her individualized cardiovascular risk, and a recommended plan of care for future pregnancies should be developed in collaboration with cardiologist colleagues. During postpartum care, opportunities should be developed to expand shared decision making whereby clinicians can understand their patients' goals, values, and preferences for health care and to facilitate a mutually suitable evaluation and management plan for future pregnancies (202).

For More Information

The American College of Obstetricians and Gynecologists has identified additional resources on topics related to this document that may be helpful for ob-gyns, other health care providers, and patients. You may view these resources at www.acog.org/More-Info/PregnancyAndHeartDisease.

These resources are for information only and are not meant to be comprehensive. Referral to these resources does not imply the American College of Obstetricians and Gynecologists' endorsement of the organization, the organization's website, or the content of the resource. The resources may change without notice.

Summary of Recommendations and Conclusions

The following recommendations and conclusions are based on limited or inconsistent scientific evidence (Level B):

Referral to a hospital setting that represents an appropriate maternal level of care dependent upon the specific cardiac lesion is recommended 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.

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- ► It may be helpful to obtain a baseline BNP level during pregnancy in women at high risk of or with known heart disease, such as dilated cardiomyopathy and congenital heart disease.
- All pregnant and postpartum patients with chest pain should undergo standard troponin testing and an electrocardiogram to evaluate for acute coronary syndrome.
- Patients should be counseled to avoid pregnancy or consider induced abortion if they have severe heart disease, including an ejection fraction less than 30% or class III/IV heart failure, severe valvular stenosis, Marfan syndrome with aortic diameter more than 45 mm, bicuspid aortic valve with aortic diameter more than 50 mm, or pulmonary arterial hypertension.

The following recommendations and conclusions are based primarily on consensus and expert opinion (Level C):

- Health care providers should become familiar with the signs and symptoms of cardiovascular disease as an important step toward improving maternal outcomes.
- ► Women with known cardiovascular disease should be evaluated by a cardiologist ideally before pregnancy or as early as possible during the pregnancy for an accurate diagnosis and assessment of the effect pregnancy will have on the underlying cardiovascular disease, to assess the potential risks to the woman and fetus, and to optimize the underlying cardiac condition.
- Patients with moderate and high-risk cardiovascular disease should be managed during pregnancy, delivery, and the postpartum period in medical centers with a multidisciplinary Pregnancy Heart Team that includes obstetric providers, maternalfetal medicine subspecialists, cardiologists, and an anesthesiologist at a minimum.
- ► Discussion of cardiovascular disease with the woman should include the possibilities that 1) pregnancy can contribute to a decline in cardiac status that may not return to baseline after the pregnancy; 2) maternal morbidity or mortality is possible; and 3) fetal risk of congenital heart or genetic conditions, fetal growth restriction, preterm birth, intrauterine fetal demise, and perinatal mortality is higher when compared with risk when cardiovascular disease is not present.
- A personalized approach estimating the maternal and fetal hazards related to the patient's specific cardiac disorder and the patient's pregnancy plans can provide anticipatory guidance to help support her decision making. For some patients, the prepregnancy evaluation may suggest a pregnancy risk that

is unacceptable. For those women, reproductive alternatives, such as surrogacy or adoption, and effective contraceptive methods should be discussed.

- All women should be assessed for cardiovascular disease in the antepartum and postpartum periods using the California Improving Health Care Response to Cardiovascular Disease in Pregnancy and Postpartum toolkit algorithm.
- All pregnant and postpartum women with known or suspected cardiovascular disease should proceed with further evaluation by a Pregnancy Heart Team consisting of a cardiologist and maternal–fetal medicine subspecialist, or both, and other subspecialists as necessary.
- Testing of maternal cardiac status is warranted during pregnancy or postpartum in women who present with symptoms such as shortness of breath, chest pain, or palpitations and known cardiovascular disease whether symptomatic or asymptomatic, or both.
- An echocardiogram should be performed in pregnant or postpartum women with known or suspected congenital heart disease (including presumed corrected cardiac malformations), valvular and aortic disease, cardiomyopathies, and those with a history of exposure to cardiotoxic chemotherapy (eg, doxorubicin hydrochloride).
- Congenital heart disease in the woman should prompt fetal echocardiography, and conversely, identification of congenital heart disease in a fetus or neonate may prompt screening for parental congenital heart disease.
- ► Women with asymptomatic valve disease should be monitored by a cardiologist and may require additional testing or care during pregnancy. The frequency of monitoring necessary is indicated in the patient's modified WHO classification.
- ► Any pregnant woman who presents with an arrhythmia should undergo evaluation to assess the cause and the possibility of underlying structural heart disease.
- Pregnant or postpartum women who present with shortness of breath, chest discomfort, palpitations, arrhythmias, or fluid retention should be evaluated for peripartum cardiomyopathy. An echocardiogram is generally the most important diagnostic test.
- Every pregnant or postpartum patient with chest pain or cardiac symptoms should have consideration of acute coronary syndrome.
- ► Although maternal cardiac arrest occurs infrequently, the health care provider should be prepared to manage this situation in any health care facility.

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- The infrequency of maternal cardiac arrest underscores the need for regular team training and practice of resuscitation skills and scenarios through simulation training.
- ► Women with complex congenital or noncongenital heart disease should be treated by a Pregnancy Heart Team.
- ► Women with stable cardiac disease can undergo a vaginal delivery at 39 weeks of gestation, with cesarean delivery reserved for obstetric indications.
- Health care providers should be aware of cardiac medications with obstetric implications as well as obstetric medications with cardiac implications.
- A postpartum follow-up visit (early postpartum visit) with either the primary care provider or cardiologist is recommended within 7–10 days of delivery for women with hypertensive disorders or 7–14 days of delivery for women with heart disease/cardiovascular disorders.
- All postpartum women with cardiovascular disease and those identified as at high risk of cardiovascular disease should be educated on their individual risk.
- Decisions regarding the most appropriate contraceptive option for a woman require discussion of her future pregnancy desires and personal preferences, as well as critical assessment of the patient's underlying disease and the relative risks and benefits of the contraceptive option considered.
- Intrauterine devices are the recommended nonpermanent option for women with high-risk cardiovascular conditions.

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The MEDLINE database, the Cochrane Library, and the American College of Obstetricians and Gynecologists' own internal resources and documents were used to conduct a literature search to locate relevant articles published between January 2010-February 2019. The search was restricted to articles published in the English language. Priority was given to articles reporting results of original research, although review articles and commentaries also were consulted. Abstracts of research presented at symposia and scientific conferences were not considered adequate for inclusion in this document. Guidelines published by organizations or institutions such as the National Institutes of Health and the American College of Obstetricians and Gynecologists were reviewed, and additional studies were located by reviewing bibliographies of identified articles. When reliable research was not available, expert opinions from obstetrician-gynecologists were used.

Studies were reviewed and evaluated for quality according to the method outlined by the U.S. Preventive Services Task Force:

- I Evidence obtained from at least one properly designed randomized controlled trial.
- II-1 Evidence obtained from well-designed controlled trials without randomization.
- II-2 Evidence obtained from well-designed cohort or case–control analytic studies, preferably from more than one center or research group.
- II-3 Evidence obtained from multiple time series with or without the intervention. Dramatic results in uncontrolled experiments also could be regarded as this type of evidence.
- Opinions of respected authorities, based on clinical experience, descriptive studies, or reports of expert committees.

Based on the highest level of evidence found in the data, recommendations are provided and graded according to the following categories:

Level A-Recommendations are based on good and consistent scientific evidence.

Level B-Recommendations are based on limited or inconsistent scientific evidence.

Level C—Recommendations are based primarily on consensus and expert opinion.

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