

Preconception Counseling for Women With Cardiac Disease

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Opinion statement

All providers who care for reproductive-aged women with cardiac disease should assess these patients' desires and plans for pregnancy at every encounter. For those considering pregnancy, preconception counseling, often performed by a maternal-fetal medicine specialist, can help patients understand the potential implications of pregnancy on their health and estimate the risks of an adverse cardiac event prior to conceiving. There are cardiac conditions, such as pulmonary hypertension and aortic stenosis, in which pregnancy may be contraindicated given the high morbidity and mortality; there are tools available to help quantify a patient's risk. Furthermore, some cardiac lesions may be inherited, which may warrant parental testing or a discussion of strategies to reduce the risk of an affected child, such as the use of assisted reproductive technologies. Preconception counseling is also important to identify other maternal risk factors, such as obesity, hypertension, and tobacco use, which are associated with adverse pregnancy outcomes and develop a strategy to mitigate their potential risks, ideally before pregnancy. For women on medications for their heart disease or other comorbidities, a thorough review of these medications can potentially avoid an exposure to a teratogen during conception and pregnancy. Once pregnant, a patient's obstetrical provider and cardiologist should work together to outline a plan to monitor a patient's cardiac status as the normal physiologic changes of pregnancy, such as increased blood volume and cardiac output, may challenge a patient's functional status and increase the risk for an adverse outcome. Labor and delivery planning are essential to ensure patients with cardiac disease deliver at the appropriate hospital, equipped with the staff and resources to care for women with complex conditions. In summary, preconception counseling aims to stratify a patient's risk in pregnancy, inform patients of possible complications, and discuss strategies to best ensure a healthy mother and baby during pregnancy, labor, and delivery.

Introduction

Preconception counseling is an important first step towards a healthy pregnancy for both the mother and infant. In a sense, preconception counseling is a preventative care that focuses on optimizing the mother's health and identifying potential risks to the mother and fetus prior to conception. All providers caring for reproductive-aged women should consider the possibility of pregnancy in their patients and screen accordingly. This screening can be as simple as the "One Key Question[®]," which simply asks, "Would you like to become pregnant in the next year?" [1]. If a patient is planning a pregnancy or not actively using contraception, she should receive preconception counseling.

Preconception counseling is often performed by maternal-fetal medicine physicians who specialize in high-risk obstetric conditions. It starts by identifying potential medical, psychosocial, and environmental risks through a thorough review of a patient's history.

For example, some common risks include obesity, hypertension, uncontrolled diabetes, teratogenic medication exposure (e.g., angiotensin-converting enzyme inhibitors, warfarin), substance use (e.g., alcohol, tobacco), exposure to secondhand smoke, and housing and/or food insecurity. When risks are identified, providers should help patients reduce the exposure risks, when feasible and not at the expense of the mother's health, and develop a strategy to optimize their health before conception.

Some women with cardiac disease are considered to be "high risk" in pregnancy, and careful planning prior to pregnancy is essential to minimize complications. In the following sections, we discuss the maternal risks of pregnancy, the components to medical optimization prior to conception, and prenatal and intrapartum considerations for women with underlying cardiac disease.

Maternal risks of pregnancy

Physiologic changes of pregnancy include increased cardiac output, increased heart rate, increased plasma volume, reduced systemic vascular resistance, and relative anemia from hemodilution [2]. These normal physiologic changes may not be tolerated by some women with cardiac disease, depending on the condition and their baseline functional status and thus necessitating the need for individualized counseling.

Several tools have been studied to help stratify the risks of pregnancy in these women and aid in preconception counseling. Sui et al. developed and validated the CARPREG score, which was predictive of cardiac events during pregnancy in 546 women (excluding terminations) with congenital (74%) and acquired (22%) heart disease and with arrhythmias (4%) [3]. Each component of the score (a history of a prior cardiac event or arrhythmia, New York Heart Association (NYHA) functional class >II or cyanosis, left heart obstruction, and systemic ventricular dysfunction (ejection fraction <40%)) contributes one point to the score. A score >1 was associated with >60% estimated risk of a cardiac event (pulmonary edema, sustained arrhythmia requiring treatment, stroke, cardiac arrest, or cardiac death). Specifically in patients with congenital heart disease, Drenthen et al. developed the ZAHARA risk score from 1302 pregnancies in 704 women [4]. A score of >3.5 from the components listed in Table 1 was associated with a 70% risk of cardiac complication, defined as an arrhythmia or heart failure requiring treatment, thromboembolic event, myocardial infarction, stroke, and endocarditis within 6 months of delivery.

With the evidence of significant morbidity and mortality of cardiac disease in pregnancy, Thorne et al. classified cardiac conditions into one

Table 1. Patient characteristics contributing to the modified risk score developed in the ZAHARA study. A risk score >3.5 was associated with a 70% chance of cardiac complication in pregnancy [4]

Patient characteristics	Points contributing to risk score
History of arrhythmias	1.50
Cardiac medication before pregnancy	1.50
NYHA class prior to pregnancy ≥ 3	0.75
Left heart obstruction (peak gradient >50 mmHg or aortic valve area <1 cm [2])	2.50
Systemic AV valve regurgitation (moderate or severe)	0.75
Pulmonary AV valve regurgitation (moderate or severe)	0.75
Mechanical prosthetic valve	4.25
Cyanotic heart disease (corrected or uncorrected)	1.00

of four categories, modeled after the World Health Organization's (WHO) classification of risk for contraceptive options [5]. This classification system, shown in Table 2, can be used in combination with risk scores to guide counseling based on specific cardiac conditions. The CARPREG, ZAHARA, and the WHO risk prediction tools were compared prospectively in a cohort of 213 pregnancies by Balci et al. [6]. In their study, 10.3% of women had a cardiac event, including arrhythmias, heart failure, or thrombotic events; this rate was noted to be lower than other reports and was attributed to their overall relatively low-risk cohort. However, of

Table 2. Classification of cardiac conditions into four categories with corresponding risk [5]

	WHO risk category			
	WHO class I	WHO class II	WHO class III	WHO class IV
Pregnancy risk	Average risk	Small increased risk of mortality and morbidity	Significant increased risk of maternal mortality and morbidity	Pregnancy contraindicated: very high risk of maternal mortality and morbidity
Conditions	-Uncomplicated or mild pulmonary stenosis, VSD, PDA, MVP -Successfully repaired simple lesions (ASD, VSD, PDA, total anomalous venous drainage) -Isolated ventricular extrasystoles and atrial ectopic beats	-Unrepaired ASD -Repaired TOF -Most arrhythmias -Mild left ventricular impairment -Hypertrophic cardiomyopathy -Native or tissues valvular heart disease (not considered WHO IV) -Marfan syndrome without aortic dilation -Heart transplantation	-Mechanical valve -Systemic right ventricle -Post-Fontan operation -Cyanotic heart disease -Other complex congenital heart disease	-Pulmonary hypertension -Severe systemic ventricular dysfunction (NHYA III-IV, LVEF <30%) -Previous peripartum cardiomyopathy with residual impairment of EF -Severe left heart obstruction -Marfan syndrome with aortic dilation >40 mm

the three models, the modified WHO classification system provided the most accurate assessment of risk. Women with contraindications to pregnancy (WHO class IV conditions) should be informed of these recommendations prior to conception. Maternal-fetal medicine (MFM) specialists can assist in outlining the maternal and neonatal risks for these extremely high-risk conditions and discuss other alternatives to parenting, such as adoption and surrogacy. Should women with one of these conditions become pregnant, they should be referred immediately to an MFM specialist, to discuss the option of pregnancy termination given the high rates of pregnancy-related mortality.

Medical optimization before pregnancy

Prepregnancy work-up

After a woman is counseled about the potential risks of pregnancy, her providers (cardiologist, obstetrician, primary care provider) should strive to ensure her health is optimized prior to conception. Women with cardiac disease should be screened for common coexisting conditions, including hypertension, diabetes, and obesity, and their possible sequelae. Compared to patients with undiagnosed or poorly controlled hypertension or diabetes, patients with well controlled disease have improved pregnancy outcomes, such as lower rates of preeclampsia, congenital malformations, intrauterine growth restriction, macrosomia, stillbirth, and delivery complications [7, 8, 9•, 10, 11, 12•]. Furthermore, all women, regardless of their cardiac condition, should be counseled that pregnancy outcomes are improved at normal body mass indices (BMI) [13, 14]. For women who are obese, weight loss should be encouraged and nutritional counseling and support strategies should be offered. Ideally, a BMI of less than 30 should be targeted prior to conception.

A patient's cardiac status should be documented prior to conception. This work-up may include an electrocardiogram, prolonged rhythm monitoring, echocardiogram, and/or stress test including cardiopulmonary exercise testing, depending on the individual patient's condition and functional status. These tests help to establish a prepregnancy baseline which can be used to guide counseling and management during pregnancy and for future comparison, in the event the patient experiences a cardiac event during pregnancy.

Medication exposures

Many commonly prescribed medications for women with cardiac disease are potentially teratogenic. Fetal teratogenic exposure can be minimized or avoided if a patient's medications are reviewed and adjusted prior to pregnancy. The greatest risk for medication-related birth defects is from exposure during the first trimester when organogenesis occurs. Common cardiac-related medications with known or suspected teratogenic or adverse fetal effects include warfarin, angiotensin-converting enzyme inhibitors, angiotensin II receptor blockers, phenytoin, atenolol, and amiodarone [15]. For some women, their cardiac disease may not allow for a teratogenic medication to be changed or discontinued throughout the

entire pregnancy; these medications may potentially be temporarily held until the highest risk period passes when embryogenesis is complete. In combination with their cardiologist, the provider performing preconception counseling will discuss the risks and benefits of continuing these medications and develop a plan for each medication in the periconception period and in pregnancy. If a potential teratogenic medication exposure is unavoidable, a screening and management plan for potential fetal malformations will be developed.

When reviewing a patient's medications, all women who are trying to conceive, or not actively contracepting, should take a prenatal vitamin daily that includes folic acid supplementation. Folic acid supplementation has been shown to decrease the risk of neural tube defects and reduce the risk of recurrence of fetal congenital heart disease for women with a previously affected fetus [16, 17, 18•]. The American College of Obstetricians and Gynecologists recommends women to consume at least 0.4 mg of folic acid per day [16]. There is not strong data to suggest that higher doses of folic acid reduce the incidence of fetal cardiac malformations; however, the report of the Canadian Task Force on Preventative Health Care recommends 1 mg of folic acid supplementation per day [19]. Ideally, this supplementation should be started a few months prior to conception to ensure optimal levels during early organogenesis.

Genetics

The possibility of inheritability should be considered in all women with heart disease, particularly those with congenital heart disease. Notably, the risk of an affected fetus is approximately 5% in women with congenital heart disease, nearly a five-fold increase from the baseline population risk [20•, 21–23]. If a genetic condition is suspected, confirmatory maternal testing is important to understand the inheritance pattern and stratify risk. Women with known genetic conditions should be informed of their options for conceiving and fetal testing. The risk of inheritance can sometimes be almost entirely avoided with assisted reproductive techniques utilizing preimplantation diagnosis or egg donation. There is also the option of early postconception screening utilizing amniocentesis, chorionic villi sampling, and ultrasound imaging. A referral to a medical geneticist can aid in diagnosis and counseling for women with a potentially inheritable syndrome.

Pregnancy, delivery, and postpartum considerations

When a woman becomes pregnant, she should be established a prenatal care with an obstetrical provider. In particular, most women with cardiac conditions have consultation with or are primarily cared for by MFM specialists. Collaboration between a patient's primary cardiologist and her obstetrician is essential to ensure her health and pregnancy care are optimized. During preconception counseling and the early stages of pregnancy, the following issues related to pregnancy and delivery should be addressed with the patient and plans made accordingly.

Fetal risk assessment

All women are routinely offered options for prenatal genetic diagnosis in the first and second trimester. For healthy patients, the standard screening includes a risk calculation for trisomy 13, 18, and 21 at the beginning of the second trimester and a fetal anatomy ultrasound between 18 and 20 weeks gestation. Women at risk for conditions not related to common trisomies, such as Marfan or DiGeorge syndromes, are offered fetal diagnostic testing for those specific conditions with either chorionic villus sampling or amniocentesis. Furthermore, a fetal echocardiogram, in addition to the detailed fetal survey, is recommended given the increased prevalence of fetal heart disease in women with congenital heart disease.

Antenatal and delivery management

Patients with cardiac disease often require regular visits with their obstetrical provider and cardiologist during pregnancy, often more frequently than their follow-up outside of pregnancy given the physiologic changes that occur throughout the pregnancy. At the start of pregnancy, women should have a baseline echocardiogram and electrocardiogram. These baseline assessments can be used to stratify a patient's risk of an adverse cardiac event and serve as a basis for comparison for later in pregnancy when the tests are repeated. As pregnancy progresses and in the absence of new or worsening symptoms, a repeat echocardiogram is typically performed around 32 weeks gestation when blood volume levels peak.

An assessment of patient's functional status and risk of an adverse cardiac event should inform delivery planning. Specifically, every patient should have a clearly documented care plan established in the early stages of pregnancy that may evolve if complications arise. Plans should be made for the patient to deliver at an appropriate facility, considering the following: the availability of the appropriate obstetric, anesthesia, cardiac staff; a patient's potential for the need for an intensive care unit, blood transfusions, and extracorporeal membrane oxygenation; and a contingency plan if the patient were to present to another hospital without the necessary resources. Furthermore, a patient's cardiology team and anesthesiologist should guide the plan for cardiac monitoring intra and postpartum, considering the need for invasive or continuous monitoring (e.g., telemetry, arterial line). Other considerations should include the need for subacute bacterial endocarditis prophylaxis, anticoagulation, fluid monitoring, and initiation or changes to a patient's cardiac medications.

Obstetrical providers should seek guidance from a patient's cardiologist on the patient's functional capacity to tolerate the process of labor and delivery. Labor is associated with an additional 30% increase in cardiac output beyond the changes associated with pregnancy. Cardiac output further increases with Valsalva maneuver and with pain [24]. Rapid changes in blood volume also occur at the time of delivery. On average, a woman can lose up to 500 mL of blood during a vaginal delivery and approximately 1000 mL during a cesarean delivery. After delivery, the uterus contracts, resulting in a rapid 500 mL increase in maternal blood volume.

Contrary to popular belief, a vaginal delivery is often the safer option for women with underlying cardiac conditions. Though a cesarean delivery can be

performed in a more controlled setting with anesthesia support at the bedside, the associated rapid fluid shifts and potential for surgical complications should be considered. Much of the risk surrounding vaginal delivery can be minimized with the use of regional anesthesia (e.g., epidural) to decrease pain and an assisted second stage of labor, with the application of forceps or vacuum delivery which minimize pushing and use of the Valsalva maneuver. This plan should be made in collaboration with the patient's cardiologist, delivering obstetrician, and anesthesiologist.

Furthermore, women with cardiac disease should have a predelivery anesthesia consultation. This consult will also allow the anesthesia team to develop an appropriate cardiac monitoring and anesthetic plan for labor and delivery that minimizes the risk to the mother and fetus.

Postpartum planning

After delivery, rapid volume shifts may put some women at risk for pulmonary edema, heart failure, or arrhythmias. A postpartum plan should clearly outline patient-specific risks and management plans that may be different from routine postpartum care. Examples of considerations for patients with cardiac disease include the necessity of intensive care monitoring, a safe protocol for pain management, fluid intake and output goals, and blood pressure targets.

When available, a multidisciplinary team of specialists with expertise in managing pregnant patients (including cardiology, maternal-fetal medicine, anesthesiology, neonatology, and nursing) should collaborate to manage issues that arise during the pregnancy; discuss the plans for labor, delivery, and postpartum care; and anticipate and prepare for any complications that may arise. If such a team and resources do not exist at the patient's local hospital, referral to a tertiary care center with access to these subspecialists and facilities should be considered early in pregnancy.

Summary

In summary, preconception counseling is important for all women with cardiac disease who are contemplating pregnancy. Women with significant or complex cardiac disease should be referred to an MFM specialist, when available, prior to pregnancy. This counseling aims to stratify a patient's risk in pregnancy, inform patients of possible complications, and discuss strategies to best ensure a healthy mother and baby during pregnancy, labor, and delivery.

Compliance with Ethical Standards

Conflict of Interest

Mark A. Clapp and Sarah N. Bernstein each declare no potential conflicts of interest.

Human and Animal Rights and Informed Consent

This article does not contain any studies with human or animal subjects performed by any of the authors.

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