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Managing congenital heart disease in the obstetric patient

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ABSTRACT

Objective: Cardiovascular disease is the major cause of pregnancy-related maternal mortality in the United States, and congenital heart disease (CHD) is the most common form of structural heart disease affecting women of childbearing age. Most females born with CHD will reach childbearing age and consider pregnancy. Adult CHD and maternal-fetal medicine (MFM) specialists managing women with CHD should provide preconception counseling, cardiovascular risk assessment prior to pregnancy that estimates maternal and fetal risk, management during pregnancy, and in the peripartum period and also know the potential complications and special circumstances that may occur in the post-partum period.

This chapter will review the population at risk, patient risk prior to pregnancy, management during pregnancy, management in the peripartum and post-partum periods, and outline specific cardiovascular complications. The chapter will also briefly review some common or high-risk congenital cardiovascular lesions commonly encountered.

Conclusion: Management of patients with most forms of CHD encountered during pregnancy requires a multidisciplinary approach and careful team-based care to facilitate safe and appropriate management and pregnancy success.

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Introduction

Congenital heart disease (CHD) is heart disease present from before birth. Congenital heart disorders are often divided into simple and complex lesions. However, pregnancy risk is not determined by CHD complexity alone, but also depends on comorbid conditions. The care of patients with complex CHD and those with comorbidities related to CHD during pregnancy requires a specialized care team, with adult congenital heart disease (ACHD), maternal-fetal medicine (MFM) specialists as well as anesthesia and occasionally other team members.

Cardiovascular physiologic and hemodynamic changes during pregnancy

The maternal blood volume increases during pregnancy, with peak around 32 weeks of gestation. A physiologic anemia occurs related to plasma volume increase which exceeds red cell mass.[1] Maternal heart rate rises by 10–20 beats per minute during pregnancy with a peak heart rate occurring during the late second or early third trimester. Combined these changes cause a cardiac output rise of 30–50% for a single pregnancy. Maternal posture can affect cardiac output; the lateral recumbent position provides the most favorable hemodynamic option.[2,3] The systemic and pulmonary vasculature dilates to accommodate increased blood volume with associated decrease in systemic and pulmonary vascular resistance. Blood pressure generally decreases slightly during pregnancy. Additional increase in heart rate, blood volume, cardiac output, and blood pressure occur during labor and delivery. These hemodynamic changes can precipitate cardiovascular symptoms in patients with known or previously undetected cardiovascular disease.

After delivery, inferior vena cava compression is relieved with associated increase in venous return. Spontaneous diuresis occurs early after delivery; however, it takes up to

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https://doi.org/10.1053/j.semperi.2017.11.008
0146-0005/© 2018 Published by Elsevier Inc.

4 weeks for hemodynamic values to return to baseline after vaginal delivery, and up to 8 weeks after cesarean delivery.

Pre-pregnancy assessment

Preconception counseling

Women with CHD should be counseled regarding pregnancy, including the importance of pregnancy planning. When pregnancy is contraindicated, counseling should provide the patient and her partner a clear understanding of the potential maternal and fetal risks of pregnancy and contraception options.

Diagnostic evaluation

Women with complex CHD must be evaluated and cared for by a multidisciplinary care team, including an ACHD cardiologist and MFM specialist experienced in the management of CHD patients.[4,5] For CHD patients contemplating pregnancy, a comprehensive evaluation is needed to adequately determine maternal risk. The impact of pregnancy on maternal cardiac status should also be reviewed. The preconception evaluation of a patient with CHD includes a comprehensive history, assessment for cardiovascular symptoms, a thorough family history, including assessment for possible genetic predisposition for CHD, and a review of all medications for pregnancy risk. A thorough physical examination including measurement of arterial oxygen saturation should be performed. A review of medical records should include analysis of information on the type of CHD, surgical history, presence of comorbidities, and any residua or sequelae associated with the cardiac lesions.

The pre-pregnancy diagnostic evaluation will depend on the complexity of CHD and prior assessments. For a new or complex CHD patient, the preconception assessment should include laboratory testing, electrocardiogram, chest radiograph, echocardiogram, and assessment by a stress or cardiopulmonary exercise test to obtain objective assessment of functional capacity and to determine the presence of exercise-induced arrhythmias.[6] Advanced testing with computerized tomography, magnetic resonance imaging, and cardiac catheterization may be recommended in patients with incompletely assessed anatomy, or suspected pulmonary hypertension. Preconception ACHD and MFM evaluation should assess a patient's pregnancy risk, identify which patients should avoid pregnancy and determine whether therapy is available and necessary to reduce pregnancy risk.[7,8]

Medications

The United States Food and Drug Administration updated pregnancy and lactation drug safety categories in 2015 (Table 1).[9] Generally, medications should be used only if absolutely necessary at the lowest possible dose and duration.[10]

Assessing maternal and fetal risk

Risk stratification scores that predict maternal and fetal risk during pregnancy are available and include the Canadian Cardiac Disease in Pregnancy (CARPREG) score, the Zwangerschap bij Aangeboren HARtAfwijkingen (ZAHARA; translated in English "Pregnancy and Congenital Heart Disease") score, and World Health Organization (WHO) classification.[11,12] The assessment and recommendation for each patient should be individualized. A preconception visit with a MFM specialist is also recommended, particularly for women at high risk for complications during pregnancy or delivery.

Fetal risk is accentuated by maternal CHD; there is an increased risk of miscarriage, spontaneous abortion, intrauterine death, and CHD in the fetus.[13] Fetal echocardiography is recommended between 18 and 22 weeks gestation in patients with CHD. Premature birth and neonatal events such as small for gestational age, respiratory distress, and neonatal death occur with increased frequency, especially in patients with the most severe types of CHD, such as Eisenmenger syndrome.

When to avoid pregnancy

Pregnancy should be avoided in the setting of certain cardiac conditions, and if pregnancy occurs in these circumstances, termination should be considered (Table 2). When pregnancy risk is unacceptable, counseling regarding appropriate and safe contraception is vital.

Genetic counseling

Genetic counseling and testing may be appropriate for couples with a suspected or known genetic disorders associated with CHD such as Holt-Oram syndrome, 22q11.2 deletion or a high risk of CHD recurrence in offspring, such as a family or personal history of CHD. The risk for recurrence of CHD varies widely from 3% to 50%, depending on the type of parenteral CHD.[14]

Pregnancy management

Care during pregnancy

The frequency and type of cardiovascular follow-up for patients with CHD depends on the underlying and comorbid conditions. Low-risk patients may be followed and delivered at local centers. High-risk patients require co-ordinated care at a referral center. The comprehensive care team should discuss the frequency of evaluation during pregnancy, anticipated delivery plan, including type of delivery and anesthesia, and post-partum care plan.

Peripartum care

The MFM and anesthesia team members manage the peripartum period; it is rare that high-level cardiovascular care is required during delivery.[15] Stable CHD patients should be able to have a normal vaginal delivery. High-risk patients may need electrocardiographic or invasive cardiac monitoring during delivery. The highest-risk patients may need to be delivered in a cardiac surgical operating room with staged cardiac surgical intervention, or the availability of cardiopulmonary bypass should acute intervention be needed.

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