Seminars in Perinatology 38 (2014) 260–272

Available online at www.sciencedirect.com

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www.elsevier.com/locate/semperi

Adult congenital heart disease and pregnancy

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ARTICLE INFO

Keywords:
adult congenital heart disease
pregnancy

ABSTRACT

Adults with congenital heart disease now form the largest group of women with cardiac disease becoming pregnant in the developed world. This is both a mark of impressive steps forward in the management of congenital heart disease and also a challenge to the medical community to develop systems of care that will best serve these women and their babies. Each woman with congenital heart disease presents a unique pattern of challenges for the cardiologist, obstetrician, and anesthesiologist, and their care should be tailored to deal with their individual circumstances. As this population of patients continues to grow, we must continue to learn and improve our diagnostic tools and management strategies to refine their care. This review intends to focus on reviewing the outcomes in this set of patients and also an approach to the assessment and the management of these patients, primarily for an audience of obstetricians, pediatricians, and anesthesiologists.

Introduction

The last 4 decades have seen a rapid improvement in the early diagnosis of fetal and neonatal cardiac anomalies, extraordinary evolution in techniques and success of childhood cardiac surgery, and improvements in postoperative and long-term pediatric cardiac care, which have resulted in a huge increase in the number of patients who survive to adulthood with congenital heart disease (ACHD). Nearly 90% of infants born with ACHD today can expect to survive to adulthood, and most with good functional status.[1] Over 1 million adults with CHD are alive today in the USA, more than the number of children with CHD.[2] As a consequence, many affected women reach childbearing age and request guidance regarding pregnancy options. Patients with repaired congenital heart disease now form the largest proportion of women becoming pregnant with significant cardiac issues.[3,4] Pregnancy is well known to impose significant hemodynamic changes on normal women. In general, most women with ACHD will have uneventful pregnancies though almost all are at some increased risk (albeit slight in many) of cardiac, obstetric, and fetal complications.[5] Some lesions continue to pose a high risk of adverse outcome, and pregnancy remains truly contraindicated in a small number of lesions.[6] Obstetricians, anesthesiologists, and pediatricians must be aware of the types of CHD (both repaired and unrepaired) encountered in adults and the potential risks to the mother, fetus, and neonate during pregnancy. A multidisciplinary approach to managing pregnancy in these women is essential, and with over 30 different types of CHD, the involvement of an ACHD cardiologist is critical as these patients develop unique patterns of complications specific to each lesion. This review will focus on the assessment and management of patients with ACHD who wish to become pregnant and on their cardiac care through pregnancy. We will review the cardiac physiology of pregnancy, general management strategies, management of specific cardiac complications, and lesion-specific considerations for an audience of obstetricians,

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http://dx.doi.org/10.1053/j.semperi.2014.04.015
0146-0005/

pediatricians, and anesthesiologists involved in the care of these patients.

Hemodynamic changes of pregnancy

The cardiovascular changes noted during pregnancy occur to meet the rising metabolic demands of the mother and the fetus. Each trimester is characterized by specific physiologic changes[7] (Fig. 1). During the first trimester, there is a 40–70% decline in total peripheral vascular resistance due to growth of the low-resistance vascular bed of the placenta and uterus. The first trimester is notable for increase in cardiac output (CO) primarily due to increased stroke volume.[8] This increase in CO peaks during the second trimester at 25–50% above pre-pregnancy levels as the heart rate also begins to increase.[9] Plasma volume increases by 30–50% until the third trimester.[9] Pregnancy also causes a relative anemia, as the increase in plasma volume is greater than the increase in red blood cell mass. Mean arterial pressure also falls due to a decline in diastolic blood pressure, reaching a nadir early in the second trimester.[9] At the end of the third trimester peripheral vascular resistance returns to baseline and may be increased by full term. In the third trimester, the supine position can lead to reduced CO due to compression of the IVC by the enlarged uterus. The left lateral position is thought to prevent compression of the inferior vena cava and can improve stroke volume and cardiac output. During labor and delivery, uterine contraction can cause a temporary increase in circulating blood volume of up to 500 ml, increase in stroke volume, and increased cardiac output up to 30–50% higher than at the onset of labor.[10] These hemodynamic changes may not be tolerated by some women with ACHD and may result in cardiac complications such as congestive heart failure and arrhythmias at different stages of pregnancy, labor, and delivery. These hemodynamic shifts may also have complications that impact fetal and neonatal development.

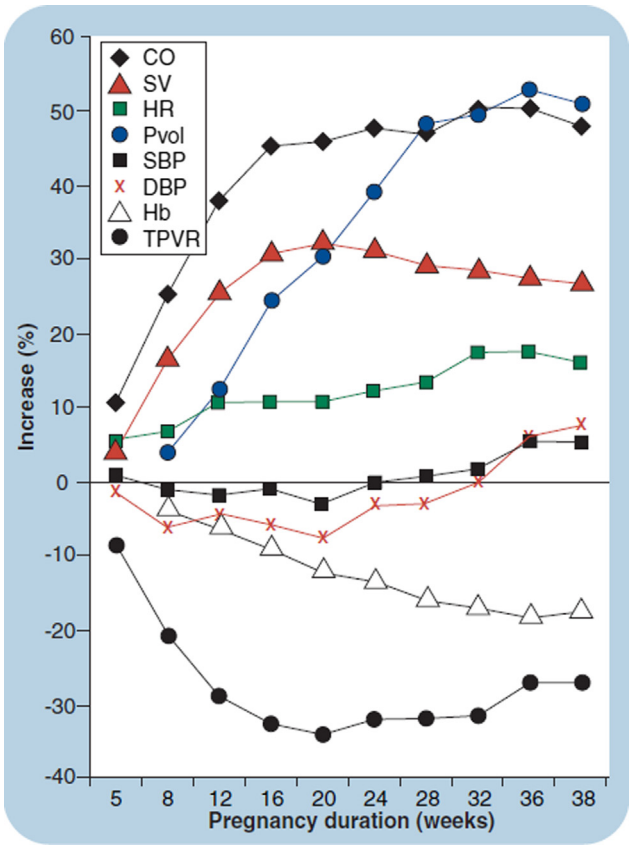

Fig. 1 – Hemodynamic changes in pregnancy: Changes in CO, PV, HR, SV, TPVR, SBP, DBP, and Hb during pregnancy. CO = cardiac output, DBP = diastolic blood pressure, Hb = hemoglobin concentration, HR = heart rate, PV = plasma volume, SV = stroke volume, SBP = systolic blood pressure, TPVR = total peripheral vascular resistance. (Adapted with permission from Karamermer and Roos-Hesselink.[7])

Given that delivery and early postpartum is the time of highest hemodynamic stress, various interventions have been designed to ameliorate some of these burdens. In general, vaginal delivery is preferred, though an assisted second stage can reduce some of the large rises in output associated with uterine contraction. Epidural anesthesia can reduce pain and distress that contribute to the acute rise in cardiac output.[11] Caesarian section can reduce some of the hemodynamic stressors of labor and delivery but is associated with greater blood loss and fluid shifts, and it is controversial whether this, overall, places less demand on the mother's cardiovascular system.[3] Currently, most authorities agree obstetric indications should prevail for determining whether a caesarian delivery is appropriate in the mother with cardiovascular disease, except where there is a risk of aortic dissection.[3]

General management of pregnancy in the woman with ACHD

Baseline clinical assessment

Whenever possible, preconception assessment is recommended for a woman with ACHD. Ideally, a visit with a high-risk obstetrician should be obtained well in advance of conception. Additionally, given that the types of CHD are so varied, a visit with an experienced ACHD cardiologist can prove valuable not only for the general management of their cardiac disease but also in ensuring the best possibility of a safe and full-term pregnancy. A thorough review of a patient's previous history, prior imaging findings, surgical records, current symptoms, and medication history is key. Medications may need to be changed where appropriate to avoid potentially teratogenic agents. A complete cardiovascular examination is important, paying attention to oxygen saturation, signs of congestive heart failure, and any residual cardiac lesions. In general, most patients would benefit from a 12-lead electrocardiogram and transthoracic echocardiogram, occasionally from stress testing or cardiac MRI, and rarely from cardiac catheterization.

Cardiac risk assessment

After obtaining appropriate clinical data, a general assessment of the risk cardiovascular complications of pregnancy should be performed. Most available risk assessment tools for