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Review Article

Pregnancy in Women with Complex Congenital Heart Disease

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Abstract

More women with complex congenital heart disease are now surviving into childbearing age. Most of these women have residual hemodynamic lesions that may impair cardiovascularadaptation to pregnancy related physiologic changes. The second and third trimesters of pregnancyare associated with significant increase in preload, and heart rate with slight decrease in afterload. Labor is associated with abrupt increase in cardiac output mostly due to increase in heart rate and contractility in the first stage and increase in preload in the second stage of labor. These physiologic changes are tolerated differently based on the type and complexity of cardiac lesions. Antenatal cardiovascular assessment is necessary to determine lesion-specific risk for the mother and the fetus. Multi-disciplinary approach is required at all stages of pregnancy in patients with significant risk of cardiovascular complications.

INTRODUCTION

Congenital heart disease (CHD) is the most common birth defect [1]. Although the birth prevalence of severe/complex CHD has remained unchanged over time [2,3]. more patients with severe/complex CHD are surviving into adulthood because of improvement in surgical and postoperative care.[4]In the current era, women with complex CHD are now growing into childbearing age and are now getting pregnant.Physiologic changes that occur during pregnancy result in significant hemodynamic burden even in women with structurally normal hearts. Most women with severe/complex CHD have residual hemodynamic lesions either due to palliative procedure or sequelae of surgical correction. Physiologic changes associated with pregnancy that otherwise would have been tolerated in a woman with a normal cardiovascular system can result in deleterious effects on the health of both the mother and her offspring in the CHD population.

PHYSIOLOGIC CHANGES ASSOCIATED WITH PREGNANCY, LABOR AND PUERPERIUM

The Starling forces, preload, afterload, heart rate and contractility, regulate the cardiovascular system. Pregnancy result is significant changes in all the above factors.There is a reduction in afterload (peripheral vascular resistance) during the first and second trimesters mostly due to low resistance placental vascular bed as well as the effect of progesterone on vascular tone [5]. Systolic and diastolic arterial pressures both fall, but there is a larger proportional fall in diastolic pressure and consequently an increase in pulse pressure. There is also significant increase in preload (plasma volume) due to activation of the renin-angiotensin-aldosterone system and reduction of plasma atrial natriuretic peptide concentration [6]. The above changes in preload and afterload result in a 50 percent increase in cardiac output by the end of the second trimester. During the third trimester, an increase in heart rate is primarily responsible for the increase in cardiac output. Compression of the inferior vena cava by the fetus can result in as much as 25-30% decrease in the stroke volume and cardiac output [7].

During the first stage of labor, cardiac output increase by 30-50% above baseline due stimulation of sympathetic nervous system resulting increase contractility and heart rate. After second stage of labor, relief of inferior vena cava compression resulting in increased preload further augments cardiac output. At the end of the third stage of labor (delivery of the placenta), splanchnic vasoconstriction occurs, divertingblood from the uteroplacental circulation back into the maternal circulation [8]. Postpartum, there is increased diuresis and natriuresisleading torestoration of the cardiovascular system back to pre-pregnancy baseline within 1-3 months postpartum [9].

MATERNAL RISK

Women with congenital heart disease are at risk of cardiovascular complications during pregnancy. These complications include heart failure, arrhythmias, stroke and cardiac death. A prospective multicenter study by Siu et al [10] determined the outcomes of 599 pregnancies not ending in miscarriage. Pulmonary edema, arrhythmia, stroke, or cardiac death complicated 13% of pregnancies. Prior cardiac

events or arrhythmia, poor functional class or cyanosis, left heart obstruction, and left ventricular systolic dysfunction independently predicted maternal cardiac complications [10]. A literature review that pooled 2491 pregnancies in women with CHD estimated that adverse cardiac events occurred in 11% of pregnancies. These cardiac events were mostly heart failure (4.8%) and arrhythmias (4.5%) [10]. Khairy et al [11] reported 19.4% risk of maternal cardiac complication in a cohort of 53 women aged 27 +/- 6 years. Cardiac risk assessment was improved by including decreased subpulmonary ventricular systolic function and/or severe pulmonary regurgitation in a previously proposed risk index developed in pregnant women with acquired and congenital heart disease [11].

In addition to cardiac complications, women with CHD are also at significant risk for obstetric complications during pregnancy [10-14]. Ouyang *et al.* recently assessed obstetrical outcomes in 112 pregnancies in 65 women with congenital heart disease [13]. In this study,33% of pregnancies were associated with adverse obstetrical events, such as preterm labor (21%), postpartum hemorrhage (14%), premature rupture of membranes (10%), pregnancy-induced hypertension or preeclampsia (3%), placental abruption (3%) and intra-uterine fetal demise (2%) [13]. Pregnancy-related hypertensive disorders are more common in subsets of patients with certain CHD diagnoses such as aortic stenosis, pulmonary stenosis, aortic coarctation and transposition of the great arteries [12]. Preeclampsia is also more frequent in women with aortic coarctation (5%), pulmonary stenosis (5%) and transposition of the great arteries (10%) [12].

NEONATAL/FETAL RISK

Neonatal/fetal complications such as premature birth, small-for-gestational-age birthweight, respiratory distress, intraventricular hemorrhage, intra-uterine fetal demise and neonatal death complicate 15–39% of pregnancies in women with congenital heart disease [9-16] Khairy et al reported adverse neonatal outcomes in 27.8% of pregnancies in women with CHD. These neonatal complications comprised of preterm delivery (20.8%), small for gestational age (8.3%), respiratory distress syndrome (8.3%), intraventricular hemorrhage (1.4%), intrauterine fetal demise (2.8%), and neonatal death (1.4%) [11]. A subaortic ventricular outflow tract gradient of 30 mm Hg independently predicted an adverse neonatal outcome [11]. Sui et al also reported neonatal adverse event rate of 20% [10]. They identified poor functional class/cyanosis, left heart obstruction, anticoagulation, smoking, and multiple gestations as independent risk factors [10]. Offspring of women with congenital heart disease are at higher for congenital heart disease compared to the general population. The overall recurrence risk is estimated at about 2–5% for overall the CHD population with significantly higher risk in patients with left-heart obstruction (7-8%) and deletion syndromes such as 22q11 [16-19].

LESION SPECIFIC RISK OF SELECTED COMPLEX CONGENITAL HEART DISEASES

Transposition of great arteries

Majority of women with history of transposition of great arteries who are of childbearing age underwent atrial switch procedure (Mustard or Senning procedure). Although this procedure achieves 'physiologic' correction and eliminates cyanosis, it leaves the morphologic right ventricle as the systemic ventricle placing the patient at risk for systolic dysfunction and inadequate contractile reserve during pregnancy. Additionally, these patients are at very high risk for arrhythmia because of multiple atrial suture lines. Drenthen et al studied 69 pregnancies in 28 women and reported that the most common cardiovascular complication in this population was arrhythmia (22% of cases), followed by heart failure and decline in functional class [20]. Obstetrical complications in this cohort include premature rupture of membranes and postpartum hemorrhage. Combined fetal and neonatal mortality was 11.8% [20]. In another series of 70 pregnancies in 40 women with atrial switch procedures, there were 16 (23%) fetal losses. Maternal cardiac complications observed in this series include arrhythmias, hemoptysis, heart failure, and decline in functional class [21]. Atotalof39%ofinfantswere delivered prematurely [21].

Tetralogy of fallot

Most women with tetralogy of Fallot (TOF) will have pulmonary regurgitation and/or pulmonic stenosis as the most common residual hemodynamic lesion. Physiologic adaptations of pregnancy such as increased plasma volume (preload) will poorly tolerated in the setting of significant pulmonary regurgitation. Also severe pulmonic stenosis will result in fixed stroke volume limiting the ability to increase cardiac output to meet increased metabolic demand of pregnancy. Other residual problems in this population include diastolic dysfunction, left ventricular dysfunction and arrhythmia [22]. Veldtman et al showed that apart from high incidence of abortion and miscarriage (29%), pregnancy in the TOF population had low risk of cardiac, obstetrical neonatal complications [20].

Fontan physiology

The Fontan physiology directs systemic venous return to the pulmonary artery bypassing the pulmonary right ventricle. Absence of pulmonary ventricle results in systemic venous hypertension and limited preload to the systemic ventricle. Patients with Fontan physiology are typically sensitive to preload changes and are at risk for edema, ascites, arrhythmias, thromboembolism, hepatic dysfunction, protein-losing enteropathy, and worsening cyanosis during pregnancy [23,24]. Subfertility or infertility and menstrual disorders are also common.[25]A small series that looked at 10 pregnancies in six patients showed 60% fetal loss (5 miscarriages and one aborted ectopic pregnancy) [26]. Of the 4 successful pregnancies, 2 were complicated by a decline in functional status and one was complicated by atrial arrhythmias. Canobbio et al [27] studied 33 pregnancies in women with Fontan physiology. In this series, 14 mothers had 15 live births (often preterm), with 13 miscarriages, five elective terminations.

CONCLUSION

With improved survival in the CHD population, pregnancy in women with CHD will remain an increasingly important and daunting clinical and public health problem. Multi-disciplinary approach is very important in achieving optimal outcome in this

population. Delivery and peripartum management should be performed in a center with significant experience in the medical, obstetric, and anesthesiologic care of these patients.

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