

Maternal Congenital Heart Disease in Pregnancy



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KEYWORDS

- Congenital heart disease • Pregnancy • Tetralogy of Fallot
- Transposition of great arteries • Pulmonary hypertension

KEY POINTS

- Most maternal heart diseases in pregnancy result from congenital heart disease.
- Pregnancy is contraindicated in certain cardiac conditions, such as dilated aortopathy, severe aortic stenosis, primary pulmonary hypertension, and severe mitral stenosis.
- Pregnancy can be safely accomplished in most individuals with careful risk assessment before conception and multidisciplinary care throughout pregnancy and the postpartum period.

INTRODUCTION

Maternal cardiac disease is present in 1% of the pregnant population,¹ most of which originates from congenital heart disease.² Global advances in recognizing and surgically correcting congenital heart disease has resulted in more women living to childbearing ages and the option to pursue future fertility. Despite these advances, pregnancy is often complicated in this population because of the profound physiologic hemodynamic changes associated with pregnancy. Cardiovascular disease has recently been identified as a leading cause of maternal mortality in the United States, although this increase is not primarily related to congenital cardiac disease.³ Pregnancies complicated by cardiac conditions of any cause require coordinated, multidisciplinary care to achieve optimal outcomes. This approach should begin in the preconception period.

MATERNAL PHYSIOLOGIC CHANGES IN PREGNANCY

Normal physiologic alterations in pregnancy often result in significant hemodynamic changes to the cardiovascular system. By 24 weeks of pregnancy, maternal blood

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volume often increases by 40% and is accompanied by marked maternal systemic vasodilation.² In response to these profound changes, heart size expands by 30%, heart rate increases, and cardiac output increases by nearly 50%.²

Delivery poses a particularly challenging situation, as systolic and diastolic blood pressure increases; cardiac output may increase by 25% during active labor and 50% during the second stage with pushing.² Immediately post partum, an auto-transfusion of about 500 mL from the uterus into the systemic circulation occurs, resulting in an incremental increase in cardiac output following delivery of the placenta.^{1,2}

These physiologic stressors are typically well tolerated by most women; however, they can pose a serious challenge for women with cardiac disorders and limited ability to adapt to significant hemodynamic changes.

GENERAL CONSIDERATIONS

Maternal Risk and Risk Stratification

Although most women with structural heart disease will tolerate pregnancy without major complications, this population remains at high risk for maternal, fetal, and neonatal adverse outcomes.⁴ Complications tend to be lower for women with congenital heart disease when compared with acquired heart disease,⁵ and as many as 25% of patients require hospitalization during pregnancy.⁶ The most common cardiac events during pregnancy include atrial arrhythmia, heart failure, and ventricular arrhythmia.² The International Registry of Heart Disease in Pregnancy (ROPAC), with data on structural and ischemic heart disease, found a high rate of maternal heart failure, most often occurring around 31 weeks' gestation.⁷ Cardiac-related medications were used in 32% of women in this registry, most commonly beta-blockers.⁸ Patients with prior cardiac surgery, New York Heart Association (NYHA) classes I and II, and those on no medication tended to experience more favorable outcomes.⁹

Multiple classification and risk prediction models have been applied to maternal cardiac disease to aid clinicians in counseling and clinical management. The Cardiac Disease in Pregnancy (CARPREG) study prospectively enrolled 562 pregnant women with congenital or acquired cardiac disease and created a maternal cardiac risk index based on the history of arrhythmia, prior cardiac event, baseline NYHA functional class, cyanosis, and systemic heart obstruction.¹⁰ The NYHA and World Health Organization (WHO) created 2 commonly used scoring systems in pregnancy. The NYHA risk classification system is based on functional status (**Table 1**), and the WHO risk classification integrates maternal cardiac risk factors with underlying cardiac disease (**Table 2**). Advancing WHO class has been clearly associated with increased maternal and fetal adverse outcomes.⁹ In an analysis of the ROPAC database,

| NYHA Class | Functional Status | Pregnancy Risk Factor |
|-------------------|--|------------------------------|
| I | Asymptomatic | Expect favorable outcome |
| II | Symptoms with greater than normal activity | Expect favorable outcome |
| III | Symptoms with normal activity | Pregnancy not advised |
| IV | Symptoms at rest | Pregnancy not advised |

Data from Simpson LL. Maternal cardiac disease: update for the clinician. *Obstet Gynecol* 2012;119(2 Pt 1):346.

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