

HEART DISEASE IN PREGNANCY 2

Congenital Heart Disease in Pregnancy

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Abstract

Congenital heart disease has become more prevalent in women of childbearing age and represents about 75% of the heart disease seen in pregnancy. Close monitoring by both obstetricians and cardiologists is advisable for women with complex heart disease, and pregnancy should still be considered contraindicated in several types of congenital heart disease. Women should also be advised of the risk that their offspring may be affected.

Women at increased risk for a cardiac event in pregnancy include those with a prior cardiac event or arrhythmia, NYHA functional class > II or cyanosis, left heart obstruction, and systemic ventricular dysfunction. In the absence of adverse predictors, however, women with congenital heart disease can be assured that pregnancy does not pose a significant risk to their health.

Résumé

La fréquence de la cardiopathie congénitale s'est accentuée chez les femmes en âge de procréer et représente environ 75 % des cardiopathies constatées au cours de la grossesse. Il est recommandé que les femmes présentant une cardiopathie complexe fassent l'objet d'un suivi étroit de la part tant d'obstétriciens que de cardiologues; de plus, en présence de plusieurs types de cardiopathie congénitale, la grossesse devrait toujours être considérée comme étant contre-indiquée. Les femmes devraient également être avisées du risque que leurs enfants en soient affectés à leur tour.

Parmi les femmes qui courent des risques accrus de connaître un incident cardiaque pendant la grossesse, on trouve celles qui ont déjà connu un incident cardiaque ou qui présentent une arythmie, une classe fonctionnelle NYHA > II ou une cyanose, une obstruction cardiaque gauche ou un dysfonctionnement ventriculaire systémique. Toutefois, en l'absence de prédicteurs d'issue indésirable, les femmes présentant une cardiopathie congénitale peuvent être assurées que la grossesse ne pose pas un risque important pour leur santé.

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INTRODUCTION

This is the second in a series of five articles reviewing in detail the assessment and management of specific cardiac disorders in pregnancy.

Congenital heart disease has become more prevalent in women of childbearing age. This change is due to increased success in the treatment of young children born with various congenital cardiac defects.¹ Congenital heart disease represents about 75% of the heart disease seen in pregnancy.² Most women present in pregnancy with New York Heart Association (NYHA) class I or II lesions and remain largely asymptomatic.² Women at increased risk for a cardiac event in pregnancy include those with a prior cardiac event or arrhythmia, NYHA functional class > II or cyanosis, left heart obstruction, and systemic ventricular dysfunction. The estimated risk of a cardiac event in pregnancy with none of these predictors was 5%, but was 27% with one predictor and 75% with more than one predictor in a large Canadian cohort. Women without adverse predictors can be reassured that the risk to their health remains low, and their expectations for a successful pregnancy should be high.²

For purposes of organization, specific lesions are separated into acyanotic and cyanotic types. The maternal mortality for congenital and acquired cardiac lesions is listed in Table 1.

Acyanotic Congenital Heart Lesions

Atrial septal defect

Atrial septal defect (ASD) is the most common congenital lesion recognized in adult life.³ Pregnancy is generally well tolerated in this group of patients. Specific treatment is not usually required.^{3,4} In two series, comprising a total of 136 pregnancies in 60 patients, 44 of whom had uncorrected

Table 1. Mortality risk associated with pregnancy

Group 1: Mortality less than 1%
Atrial septal defect, uncomplicated
Ventricular septal defect, uncomplicated
Patent ductus arteriosus, uncomplicated
Pulmonic and tricuspid disease
Corrected tetralogy of Fallot
Porcine valve
Mitral stenosis, NYHA classes I and II
Group 2: Mortality 5% to 15%
Mitral stenosis with atrial fibrillation
Artificial valve
Mitral stenosis, NYHA classes III and IV
Aortic stenosis
Coarctation of aorta, uncomplicated
Uncorrected tetralogy of Fallot
Previous myocardial infarction
Marfan syndrome with normal aorta
Group 3: Mortality 25% to 50%
Pulmonary hypertension
Coarctation of aorta, complicated
Marfan syndrome with aortic involvement

NYHA: New York Heart Association.

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ASD, the live born infant rate was 89%.^{5,6} Other studies suggest that pre-pregnancy correction of ASD can reduce the risk of miscarriage, preterm labour, and cardiac symptoms.⁷ Although most patients with ASD tolerate pregnancy well, secondary pulmonary hypertension may develop in a small subset of these patients by the time they reach adulthood. This, in turn, could lead to reversal of the shunt, cyanosis, and increased morbidity and mortality.⁸

Ventricular septal defects

A ventricular septal defect (VSD) is present in 1.5 to 2.5 of 1000 women with a pregnancy resulting in a live birth.⁹ Many of these defects close spontaneously. Those that do not are often surgically corrected before childbearing. Some patients with a larger uncorrected defect may develop congestive heart failure, arrhythmias, or pulmonary hypertension.¹⁰ Whittemore et al.⁶ described the outcome of 98 pregnancies in 50 patients with ventricular septal defect, most of whom had uncorrected lesions. The liveborn infant rate was 80%. Pregnancy is usually well tolerated in women with corrected VSD, although Jackson et al.⁹ advise a thorough evaluation searching for evidence of secondary pulmonary hypertension.

Patent ductus arteriosus

Although most patients with patent ductus arteriosus (PDA) undergo repair during infancy or childhood, some patients may present with uncorrected lesions during pregnancy.⁸ Those with uncorrected PDA with a small or moderate-sized ductus and normal pulmonary arterial pressure can also expect an uncomplicated pregnancy.⁴ In patients with a significant left-to-right shunt, secondary pulmonary hypertension may occur and result in the increased morbidity and mortality of Eisenmenger syndrome.⁴ Patients who have corrected PDA generally have an uncomplicated course in pregnancy.⁸ Whittemore et al.⁶ report the outcome of 105 pregnancies in 42 women with PDA, all of which had been surgically corrected. The liveborn infant rate was 79%, and there were no maternal complications. Transcatheter closure of a large PDA in pregnancy has been reported.¹¹

Coarctation of the aorta

Coarctation of the aorta has a prevalence of 0.3 to 1 in 1000 in the female population.¹² Many of these patients have had corrective surgery before pregnancy. However, in patients with uncorrected coarctation, pregnancy was once thought to carry such a severe risk to life that termination of

pregnancy and sterilization were recommended.¹³ More recently collected series in patients reveal a low maternal mortality: 0% to 3.5%,¹⁴⁻¹⁶ with good fetal outcome. Deal and Wooley¹⁴ reported that in 185 pregnancies in 83 patients with uncorrected coarctation of the aorta, they found a pregnancy loss rate of 18.9%. Blood pressure during pregnancy in patients with coarctation of the aorta usually follows a similar pattern to that of patients with essential hypertension in pregnancy; that is, it is unchanged or falls in the second trimester, with a return to baseline levels near term.¹⁵

Aortic rupture or dissection is a serious concern in patients with coarctation of the aorta. Correction of coarctation of the aorta in pregnancy with successful maternal and fetal outcomes has been reported.^{12,17} Risks in pregnancy are increased in patients with associated cardiac lesions such as septal defects or bicuspid aortic valves,⁸ as well as in patients with aortic, intervertebral, or circle of Willis aneurysms.¹⁸

It appears that pregnancy is well tolerated in patients who have corrected coarctation of the aorta.⁸

Marfan syndrome

Marfan syndrome is rare, with an incidence of 5/100 000.¹⁹ Although mortality rates as high as 50% have been reported for pregnant patients, those with a documented aortic root diameter of less than 40 mm without an abnormal aortic valve have a mortality rate of less than 5%.²⁰ The risk of aortic dissection during pregnancy or shortly thereafter was 17% in one series of 36 women without cardiac symptoms prior to pregnancy.²¹ Aortic or mitral regurgitation is also seen in 60% of patients with Marfan syndrome and may complicate pregnancy.²² In an attempt to decrease risk of aortic dilatation, some authors recommend use of oral β -blockers.^{2,23} Pregnancies complicated by Marfan syndrome are not associated with poor perinatal outcomes, though some suggest an increased risk of incompetent cervix.^{21,24,25} Patients should be counselled regarding the risk of autosomal dominant inheritance and the need for follow-up for their offspring.

Aortic stenosis

Patients with mild or moderate aortic stenosis (AS) usually tolerate pregnancy well.²⁶ Most are able to increase cardiac output appropriately. With severe disease, however, cardiac output can remain relatively fixed, and even limited exercise may put these patients at risk for sudden cardiac or cerebral hypoxia, resulting in angina, myocardial infarction, syncope, or even sudden death.¹⁰ Therefore, in patients with severe AS, the mainstay of treatment is limitation of activity. Patients reported to be at most risk are those whose aortic valve peak-to-peak systolic gradient is more than 100 mm Hg.¹⁰

As cardiac output increases during pregnancy, aortic valve area, rather than valve gradient, may be a better predictor of the severity of AS.²⁷

In a review of their experience with uncorrected AS of all degrees, Lao et al. described 82 pregnancies in 65 patients.²⁸ Seven women (11%) died; two of these deaths occurred at two and 10 months postpartum. Three fetuses (3.7%) were lost to stillbirth or neonatal death, and four (4.9%) were delivered prematurely. Silversides et al. described their experience with 49 pregnant women with AS, 29 of whom had severe disease. None of the 20 women with mild or moderate AS had cardiac complications during pregnancy. Three (10%) of those with severe AS suffered cardiac complications during pregnancy: two developed pulmonary edema, and one developed persistent arrhythmia. Following delivery, 8% of those with mild or moderate AS required surgical repair, and 41% of those with severe AS underwent surgical repair. The mean time of surgical intervention was 2.6 ± 2 years after delivery. There were no cardiac-related deaths during pregnancy or the follow-up period. Fetal and neonatal complications were not above expected rates.²⁶

Patients with severe AS are at greatest risk during termination of pregnancy or at delivery, as these are times of significant potential for critical falls in cardiac output. Such patients should remain in the left lateral position when possible. In an effort to control preload and avoid the consequences of decreased cardiac output, invasive hemodynamic monitoring for patients with severe AS is recommended.^{8,27}

During labour and delivery, patients with AS are at much greater risk from the consequences of hypovolemia than from pulmonary edema. Clark recommends that pulmonary wedge pressure should be kept at approximately 16 mm Hg in order to maintain a margin of safety.¹⁰ Strict attention should be paid to controlling blood loss in the immediate postpartum period or at the time of Caesarean section. Caesarean section in women with AS should be reserved for obstetrical indications. Successful replacement of the aortic valve in pregnancy has been reported in patients not amenable to medical therapy.^{29,30} Balloon valvuloplasty of severe AS during pregnancy may have a role in the treatment of some patients.³¹

Pulmonic stenosis

Pulmonic stenosis of a mild to moderate nature, i.e., associated with a transvalvular pressure gradient less than 80 mm Hg, is generally well tolerated in pregnancy.^{3,10} Whittemore et al.⁶ describe 46 pregnancies in 24 patients with a livebirth rate of 78%. However, only one of three patients who had a fair to poor functional classification before pregnancy had a liveborn infant. Clark cautions about the risk of right-sided heart failure in patients with severe pulmonic stenosis.¹⁸

Table 2. Given one affected parent, suggested offspring risk for congenital heart defects (%)

Defect	Mother affected	Father affected
Aortic Stenosis	13–18	3
Atrial Septal Defect	4–4.5	1.5
Atrioventricular Canal	14	1
Coarctation of the Aorta	4	2
Patent Ductus Arteriosus	3.5–4	2.5
Pulmonic Stenosis	4–6.5	2
Tetralogy of Fallot	2.5	1.5
Ventricular Septal Defect	6–10	2

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Patients with severe pulmonary stenosis should undergo catheterization or surgical correction before pregnancy.¹⁸

Cyanotic Congenital Heart Lesions

Tetralogy of Fallot

Tetralogy of Fallot is the most common cyanotic heart lesion that permits survival into adulthood.^{4,8} For patients without prior surgical correction, the prognosis is guarded. Meyer et al.³² described a series of 57 pregnancies in such patients with a maternal mortality of 7% and a fetal loss rate of 22%. The increase in maternal mortality and morbidity is due to the decrease in systemic vascular resistance associated with pregnancy and a subsequent increase in the patient's right-to-left shunt. This leads to further cyanosis, a compensatory rise in hematocrit, and a corresponding decrease in arterial oxygen saturation.⁴ A poor prognosis exists for patients whose shunting is of such a degree as to result in a hematocrit 60% or more or an arterial oxygen saturation of less than 80%.³³

Most patients with tetralogy of Fallot undergo surgical treatment during infancy or childhood.^{10,34} For patients entering pregnancy with a corrected lesion, the prognosis is favorable.^{32,35} Singh et al.³⁵ reported the outcomes of 40 pregnancies in 27 patients with surgically corrected tetralogy of Fallot. There were no maternal deaths. One patient required thiazide diuretics for shortness of breath, and one infant was born with pulmonary atresia. Similar results were observed by Zuber et al. in 44 pregnancies in 19 patients who had previous corrective surgery.³⁴

Ebstein's anomaly

Ebstein's anomaly is an uncommon congenital cardiac lesion that may be complicated by cyanosis.⁸ It represents approximately 1% of all congenital cardiac lesions.³⁶ The specific abnormality involves displacement of the tricuspid

valve, resulting in an enlarged right atrium, a small right ventricle, and a regurgitant tricuspid valve. An atrial septal defect, ventricular septal defect, or patent foramen ovale may complicate the lesion and result in right-to-left shunting and cyanosis.³⁷

Two centres have reported their experience with pregnancy and Ebstein's anomaly.^{37,38} Combined, these reviews describe the outcomes of 153 pregnancies in 56 women. There were no maternal deaths and the live birth rate was 79%. Six women required treatment for tachycardia due to Wolff Parkinson White syndrome, associated with Ebstein's anomaly. Nineteen patients (34%) were cyanotic; in the series reported by Connolly et al.³⁷ this was associated with a significantly smaller birth weight. Efforts should be made to control arrhythmias and reduce any degree of cyanosis to minimize maternal and fetal morbidity.

Eisenmenger syndrome

Eisenmenger syndrome is an acquired elevation of pulmonary vascular resistance and pulmonary artery pressure as a result of a left-to-right intracardiac shunt.³⁹ This eventually results in a right-to-left or bidirectional shunt, with subsequent cyanosis and polycythemia. Many reports describe the poor outcome of patients with Eisenmenger syndrome who become pregnant.^{39–44} Gleicher et al.⁴⁴ describe 70 pregnancies in 44 women with confirmed Eisenmenger syndrome. Twenty-three patients (52%) died either during pregnancy or within the first month postpartum. The maternal mortality was 36.1%, 26.7%, and 33.3% for first, second, and third pregnancies, respectively, suggesting that a previous successful outcome is not a valid predictor of outcome in future pregnancies. Death was related to thromboembolism in 43.5% and to hypovolemia in 26.1%. Two patients died before delivery, four patients

died intrapartum, and most of the remainder died within one week of delivery. The perinatal mortality was 28.3%.

Despite many advances in cardiology care over the past 20 years, the mortality rate in women affected with Eisenmenger syndrome has shown little improvement.⁴⁵ Pregnancy should be considered contraindicated in patients with this cardiac disorder. However, should a patient become pregnant, termination of pregnancy appears to offer an improved maternal prognosis, with a mortality rate of 7.1%.⁴⁴

Regardless of risk, some patients will choose to continue pregnancy or may have the diagnosis made during pregnancy. Case reports have described aggressive therapy with inhaled nitric oxide, epoprostenol, sildenafil, and L-arginine as having some success.⁴⁶⁻⁴⁹ Many aspects of the intrapartum care of patients with Eisenmenger syndrome remain unproven and controversial. These include regional anaesthesia,^{39,50,51} invasive hemodynamic monitoring,^{27,33,34} and various methods of delivery.^{44,50}

Primary Pulmonary Hypertension

Primary pulmonary hypertension is uncommon, and there are few reports of pregnancy associated with this condition.^{45,52} The maternal mortality associated with primary pulmonary hypertension ranges from 30% to 56%.^{45,52} Premature delivery is indicated for maternal reasons in the majority of cases, and associated neonatal morbidity and mortality are high.⁴⁵ These patients should be advised not to become pregnant and should be treated in the same way as patients with Eisenmenger syndrome.

Offspring Risk of Congenital Heart Disease

For patients with congenital cardiac disease, a discussion of the increased risk of congenital heart disease in their offspring is an important component of prenatal counselling. The incidence of heart disease is increased in the offspring of patients with almost all forms of congenital heart disease (Table 2). The risk is generally higher if the mother, rather than the father, is affected.³ Fetal echocardiography at 20 to 23 weeks' gestation is recommended for a pregnant patient with a congenital heart defect.

CONCLUSION

Most women who enter pregnancy with congenital heart disease can be reassured that pregnancy will not significantly increase their risk of morbidity or mortality. However, some women with complex heart disease will require very close monitoring by both obstetricians and cardiologists. Understanding the normal physiologic adaptations to pregnancy, especially at the time of delivery, will help predict when these women may decompensate. Despite significant advances in cardiac care, pregnancy

should be considered contraindicated in several types of congenital heart disease and termination of pregnancy advised. Women with any type of congenital heart disease should be advised of the risk that their offspring may also be affected, and fetal echocardiography is recommended.

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