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Review

Congenital Heart Disease and Women's Health Across the Life Span: Focus on Reproductive Issues

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ABSTRACT

From adolescence to older age, women with congenital heart disease (CHD) face unique challenges. In this review we explore the ways in which CHD affects women's sexual and reproductive health and, in turn, how their sexual and reproductive history affects the course of their CHD. In adolescence, special attention must be paid to menstrual irregularities and concerns of developing sexuality and self-image. Discussions about sexuality and reproduction are an important part of transition planning and must be done with an awareness of the adolescent's developing understanding and maturity. Pregnancy imposes a hemodynamic load on the heart which may lead to cardiac, obstetric, and fetal/neonatal complications in women with CHD. Pre-pregnancy counselling must include an assessment of maternal and

RÉSUMÉ

De l'adolescence à un âge plus avancé, les femmes atteintes d'une cardiopathie congénitale font face à des défis uniques. Nous explorons ici les répercussions de la cardiopathie congénitale sur la santé des femmes en matière de sexualité et de reproduction et, inversement, les répercussions des antécédents des femmes en matière de sexualité et de reproduction sur la cardiopathie congénitale. À l'adolescence, il faut accorder une attention particulière à l'irrégularité du cycle menstruel et aux questions relatives à la sexualité et à l'image de soi en développement. Des discussions au sujet de la sexualité et de la reproduction constituent un volet important de la planification de la transition et doivent tenir compte du degré de compréhension et de maturité de l'adolescente. La grossesse impose un fardeau

Women with congenital heart disease (CHD) are increasingly surviving into adulthood and older age. As these women age, issues of sexual health and reproduction are increasingly coming to the forefront. The experience of women with CHD through the life span differs from both their male counterparts and healthy peers. From the outset, female patients with CHD appear to have poorer outcomes after high-risk cardiac surgery.[1] In addition, as girls enter into adolescence a new host of health issues arise involving menstruation, sexual activity, and contraception. As they start to consider a family, the effect of their heart condition on their ability to carry a pregnancy to term, as well as the enduring influence of pregnancy on long-term health and life span need to be considered. In many developed countries, cardiovascular disease is now the leading cause of nonobstetrical death during pregnancy. There is also an increasing recognition of a link between the complications of pregnancy and cardiovascular events in later life, prompting calls for action.[2,3] In a study of 2 large North American centers, CHD made up 64% of pregnancies in women with heart conditions.[4] These women have higher rates of complications during pregnancy, and the effect on their long-term health is still poorly understood. Although there has been a large body of research directed toward understanding the risk during pregnancy in women with CHD, relatively little has been published on women's health in adolescence and young adulthood or on their postpartum experiences. In the present review, we examine issues related to sexual and reproductive health of women with CHD across the life span, with a focus on pregnancy (Fig. 1; Table 1).

Adolescence

Adolescents with CHD represent a challenge for health professionals as they are in the process of changing from pediatric to adult care. The transition has been defined as the "purposeful planned movement of adolescents with chronic medical conditions from child-centered to adult-oriented health care."[5] The American Academy of Pediatrics recommends starting transition care at the age of 12 years and finishing at the age of 18-21 years with transfer to adult care.[5] The Canadian Pediatric Society does not give an age to start transition but suggests it is an "ongoing process that may begin as early as the time of diagnosis and ends sometime after transfer."[6] The American Heart Association guidelines suggest "providing age-appropriate counselling

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fetal risk according to several well developed models. Counselling should also include discussions about fertility and alternatives to pregnancy when appropriate. Recommendations for contraception must be made according to the patient's cardiac lesion. In caring for women with CHD during pregnancy, a multidisciplinary cardio-obstetrics team is recommended to optimize care. More research is needed into the long-term impact of pregnancy on the prognosis of patients with CHD. As women with CHD increasingly survive into old age, more attention will need to be directed toward the treatment of menopause and acquired heart disease in this population.

hémodynamique au cœur et peut mener à des complications cardiaques, obstétricales et fœtales/néonatales chez les femmes atteintes d'une cardiopathie congénitale. Le counselling préalable à la conception doit comprendre une évaluation des risques pour la mère et pour l'enfant fondée sur plusieurs modèles bien établis. Il doit aussi comprendre des discussions sur la fertilité et sur les solutions de rechange à la conception, s'il y a lieu. Des recommandations quant à la contraception doivent être formulées en fonction des lésions cardiaques de la patiente. Afin d'optimiser les soins, on recommande de confier la patiente atteinte d'une cardiopathie congénitale aux soins d'une équipe multidisciplinaire formée de spécialistes en cardiologie et en obstétrique durant la grossesse. D'autres recherches s'imposent pour examiner le retentissement à long terme d'une grossesse sur le pronostic des patientes atteintes d'une cardiopathie congénitale. Comme les femmes atteintes d'une cardiopathie congénitale vivent de plus en plus longtemps, il faudra également accorder plus d'attention au traitement des symptômes de la ménopause et des maladies cardiaques acquises au sein de cette population.

that includes discussion of the anticipated effects of pregnancy on maternal heart disease and the importance of pregnancy planning, including effective contraception options." Despite this recommendation, many women are entering adulthood with inadequate information concerning their sexuality and reproductive health. In one study, 66% of adolescent girls had never discussed the risks of pregnancy with a physician before transferring to adult care.[7] Similarly, in another study of young women transferring to adult care, patients expressed that they received sometimes conflicting and incomplete information on contraception and pregnancy.[8] In a study of 536 women with CHD, 43% had not been counselled about contraception.[9] A study by our group found that only 37% of women with CHD had ever been told that they were at increased risk in pregnancy.[10] In addition, many women who were at low risk regarding pregnancy were left with the impression that they were at high risk and sometimes avoided pregnancy as a result. Ensuring that accurate information is delivered before conception should be of utmost priority to those caring for young women with CHD.

Menstruation

Little has been published regarding menstruation patterns in women with CHD. For women with acyanotic heart disease, menarche occurs at the age of 13 years, somewhat later than the mean reported in the general population (12.3 years). In cyanotic women, menarche occurs even later, at 13.9 years of age, and they have a higher frequency of menstrual irregularities.[9] Delayed menarche in women with cyanotic or complex heart disease is due to a high incidence of primary amenorrhea.[11]

Special consideration must be given to patients on anticoagulation for prosthetic mechanical heart valves, arrhythmias, or prophylaxis (in single ventricles), because there is a nearly 3-fold risk of menorrhagia among such patients.[12] Adolescents need to be warned about the potential for heavy bleeding and menstrual irregularity. In fact, one study reported a general increase in menorrhagia in women with CHD compared with the general population.[13] Also in that study, the age at menarche in complex cyanotic heart disease was higher than previously reported, at 14.5 years. These menstrual irregularities may also play a role in infertility and difficulty conceiving among patients with complex CHD.

Sexuality

The data on sexual behaviour among adolescents with CHD is conflicting. The National Health and Nutrition Examination Survey found no difference between patients with CHD and control subjects in age at first sexual intercourse, lifetime sexual partners, or reported condom use.[14] In contrast, other studies have found that reported rates of sexual intercourse in adolescents and young adults with CHD were significantly lower than those of their healthy peers.[15-17] However, more than one-third of young adults who were sexually active still reported engaging in potentially risky behaviour. One theory is that the late onset of sexual activity may involve a delay in psychosocial development among adolescents with CHD.[16] Indeed individuals with CHD may be treated as "asexual" by their families.[18] Other studies have reported increased distress during sexual intercourse or fears of rejection and embarrassment regarding their scar.[17,19,20]

Women with CHD have been shown to have higher levels of sexual dysfunction. A study of 254 patients demonstrated that women with CHD had more sexually related distress and dysfunction.[17] Women with CHD have also been reported to have impaired psychosocial adjustment and lower self-esteem compared with their men with CHD.[21,22] Health professionals should therefore consider assessing the level of knowledge, readiness, beliefs, and feelings of patients concerning sexuality before discussions about contraception and pregnancy and address those issues on an individual basis. A Japanese study suggested that counselling for young women is more effective when patients have a better understanding of what it means to live with CHD.[23] This should serve as a reminder to pediatric specialists to start raising early awareness among adolescents about their disease and its impact on their life and future.

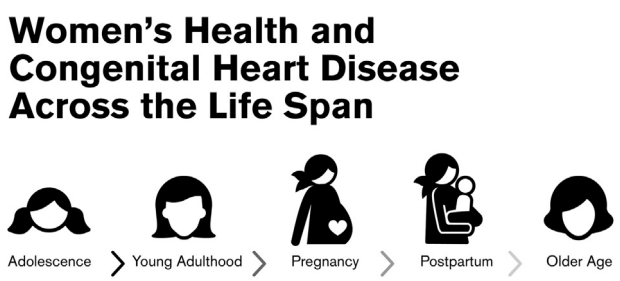


Figure 1. Women's heath and congenital heart disease across a life span. Icons from nounproject.com.

Adulthood

Maternal risks during pregnancy

Pregnancy risks are important to consider for young women with CHD because the hemodynamic and hormonal changes of pregnancy can destabilize the cardiovascular system and lead to complications.

During pregnancy, there is an increase in blood volume of up to 40%-50%, peaking in the third trimester.[24,25] Plasma volume increases disproportionately compared with red cell volume, leading to the physiologic anemia of pregnancy.[25,26] Heart rate increases as well, and overall there is a resulting increase in cardiac output of ~ 40% during pregnancy.[27-29] Peripheral vascular resistance falls by 30% until ~ 24 weeks of gestation, leading to an overall decrease in blood pressure. During labour and delivery, cardiac output increases a further 50% in response to anxiety and painful uterine contractions.[30] Once delivery is accomplished, relief of caval obstruction by the gravid uterus leads to an autotransfusion that increases maternal cardiac output a further 60%-80%.[30,31] This may be offset by bleeding at the time of delivery. Within hours of delivery, cardiac output falls dramatically, but fluid shifts are common after delivery, and the risk of developing heart failure is increased at this time. It can take up to 6 months for the maternal circulation to return to its prepregnancy state.[32]

Women who are unable to tolerate the hemodynamic changes of pregnancy develop cardiac complications.[33-36] Risk assessment is discussed elsewhere in this review. The most common maternal cardiac complications that occur during pregnancy are arrhythmias and congestive heart failure. These occur in, respectively, ~ 4.5% and 4.8% of pregnancies with CHD, but the rates vary significantly according to underlying lesion.[33] Other cardiac complications include thromboembolic events, cerebrovascular events, and endocarditis. Maternal mortality is rare, but can occur in women with high-risk cardiac lesions (Fig. 2). Women with CHD are also have a higher rate of adverse obstetrical (8.4%) and fetal and neonatal events (24%) compared with the general population.[37] Pre-eclampsia is higher in certain subgroups, such as those with coarctation of the aorta,[11,33,38] and there are higher rates of hemorrhage and placental abruption.[39]

Fetal and neonatal risks

Women with CHD have higher rates of miscarriage, premature labour (16%), and neonates that are small for gestational age (8%-25%) compared with the general population.[33,37,40] Particularly complex CHD, such as single-ventricle physiology, is a risk factor for premature birth, with rates of 22%-65%.[33,41] Neonatal mortality is increased in the CHD population at ~ 4% compared with 1% in the general population, owing in part to the high rate of premature birth. However, the mechanisms that contribute to poor fetal and neonatal outcomes are poorly understood. Placental insufficiency appears to play a role. In several studies, intrauterine markers of placental health have correlated with impaired neonatal outcomes in women with CHD.[42,43] These early studies provide support to the theory that maternal cardiac function and abnormal placental function are important mechanisms contributing to adverse outcomes in this group of women. Indeed, recent research has suggested that even in women without known heart disease, maternal cardiac function may play an important causative role in preeclamptic pregnancies.[44,45]

The impact of these outcomes is far reaching. Aside from the short-term morbidity associated with low birth weight and premature birth, there are potential long-term implications for the children of mothers with CHD. The Barker hypothesis states that children born small for gestational age or in an environment complicated by placental insufficiency may be at higher risk for cardiovascular and metabolic complications later in life.[46] There is also some thought that these children may themselves be at risk for hypertensive disorders of pregnancy as adults. Thus the impact of CHD may have transgenerational consequences that outstrip simply the risk of transmission of CHD.

The risk of transmission of heart disease to offspring ranges from 3%-5% in patients for whom a genetic cause is not known to 50% in those with conditions with an autosomal dominant pattern of inheritance, such as 22q11.2 deletion syndrome or Noonan syndrome.[47,48] All patients should have a thorough 3-generation pedigree performed.[41] Genetic counselling should be available and the risks of transmission clearly discussed. All pregnant women with CHD should be offered a fetal echocardiogram between 18 and 22 weeks of gestation. For those with known autosomal

Table 1. Congenital heart disease and women's health across the life span

Life stage	Issues requiring attention
Adolescence	Menstruation/delayed menarche. Sexuality/sexual identity/self-esteem. Contraception. Understanding diagnosis. Transition.
Young adulthood	Menstrual irregularities. Sexuality/sexual identity/self-esteem. Contraception. Pregnancy planning. Subfertility/infertility.
Childbearing	Effect of maternal heart disease on pregnancy. Effect of pregnancy on maternal heart disease. Obstetrical risk. Fetal/neonatal risk.
Late adulthood	Long-term impact of pregnancy on heart. Maintenance of health, role as parent, caregiver.
Older age	Menopause. Acquired medical conditions and assessment of cardiovascular risk.

High-Risk Cardiac Lesions mWHO Class III	Extremely High-Risk Cardiac Lesions: Pregnancy Contraindicated mWHO Class IV
Cardiomyopathies	
• Moderate left ventricular systolic dysfunction • Previous peripartum cardiomyopathy without residual left ventricular impairment • Subaortic morphologic right ventricle with good or mildly decreased ventricular function	• Severe left ventricular systolic dysfunction (ejection fraction < 30%, NYHA class III-IV) • Previous peripartum cardiomyopathy with residual ventricular impairment • Subaortic morphologic right ventricle with moderate or severely decreased ventricular function
Valve Lesions	
• Mechanical valve • Severe asymptomatic aortic stenosis • Moderate mitral stenosis	• Severe mitral stenosis • Severe symptomatic aortic stenosis
Congenital Heart Disease	
• Fontan circulation with no complications • Unrepaired cyanotic heart disease • Other complex heart disease	• Pulmonary arterial hypertension • Severe (re)coarctation • Fontan circulation with any complications
Aortopathies	
• Marfan syndrome and other heritable thoracic aortic disease with aortic dimensions of 4.0–4.5 cm • Bicuspid aortic valve associated aortopathy with aortic dimensions of 4.5–5.0 cm • Turner syndrome with an aortic size index of 2.0–2.5 cm/m2	• Marfan syndrome and other heritable thoracic aortic disease with aortic dimensions > 4.5 cm • Bicuspid aortic valve associated aortopathy with aortic dimensions > 5.0 cm • Turner syndrome with an aortic size index of > 2.5 cm/m^2 • Vascular Ehlers-Danlos
Arrhythmias	
• Ventricular tachycardia	

Figure 2. High risk cardiac lesions for maternal cardiac complications. mWHO, modified World Health Organization; NYHA, New York Heart Association. Data from Regitz-Zagrosek et al.[50]

dominant conditions, preimplantation genetic testing may be discussed.

Preconception counselling

Preconception counselling should begin in adolescence and continue into adulthood as an essential part of transition planning. It should be performed by specialists with knowledge of pregnancy in the CHD population. The focus is on the importance of pregnancy planning to optimize cardiac status and minimize risk to both mother and the baby including the risk of genetic transmission of heart disease to offspring. All women should be educated about the maternal cardiac risks, the obstetrical risks, and the fetal and neonatal risks. Because subfertility and infertility can occur in this population, discussion on the safety of fertility treatments may be required. For women at very high risk for complications in pregnancy, alternate options, such as adoption or surrogacy, may be important to address at this time. Finally, discussions should also include the long-term impact of pregnancy on the mother's cardiac status and life expectancy. Ideally, women with CHD who are actively considering pregnancy should have a preconception visit with a specialist in CHD and pregnancy. Consulting physicians review the complete cardiac, surgical, and obstetrical history, including an assessment of maternal functional status, cardiac symptoms, and risk factors for preeclampsia. Teratogenic medications, such as angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, and endothelin receptor antagonists, should be discontinued and switched to safer alternatives when possible. The details of the history, physical examination, and investigations to be covered during this visit are outlined in Figure 3.

Tools to assess maternal cardiac risk in pregnancy have been developed by several working groups. The original **Car**diac Disease in **Preg**nancy (CARPREG) study examined pregnancy outcomes in women with both acquired heart disease and CHD and derived a risk prediction score based on 4 baseline maternal predictors (Table 2).[34] The subsequent CARPREG II risk score included 10 risk predictors including 5 general cardiac predictors, 4 lesion-specific predictors, and 1 predictor related to delivery care predictor (Fig. 4).[4] The ZAHARA (**Z**wangerschap bij **A**angeboren **Hart**afijkingen [Pregnancy with Congenital Heart Disease]) score is selective for the CHD population and includes several weighted predictor variables for adverse maternal events during pregnancy.[49] Finally, the modified World Health Organization (WHO) score is a commonly used risk stratification tool that, based on expert opinion, places women into risk categories

based on cardiac lesions.[50] Figure 2 lists the high risk cardiac lesions for maternal cardiac complications according to the modified WHO classification. Risk scores serve as a basis for risk assessment, but information on lesion-specific risks and addition information specific to an individual from additional tests such as cardiopulmonary tests or cardiac computed tomography (CT) or magnetic resonance imaging (MRI) should also be integrated into the risk assessment. For a small group of women with CHD, pregnancy is contraindicated owing to very high maternal morbidity and mortality. Many of these lesions are included under WHO category IV (Table 2). They are also outlined in detail in a review by Elkayam et al.[51]

Management during pregnancy and the peripartum period

For those women with CHD who become pregnant, care from a multidisciplinary cardio-obstetrics team is recommended. The term "cardio-obstetrics" has been coined to emphasize the multifaceted care these women require.[52] At our center, the core team includes specialists in adult CHD, maternal-fetal medicine, nursing, fetal cardiology, and obstetric anaesthesia. Other specialties also may be involved, including genetics, hematology, general internal medicine, social work, etc, depending on the individual needs of the patient (Fig. 5).

The nature and frequency of follow-up during pregnancy will vary according to the patient's cardiac lesion, history, and hemodynamic status. For women with simple CHD lesions, a single visit during pregnancy may be adequate to ensure that there are no issues and to outline a delivery plan. These women may deliver at their local hospital. Women with intermediate and complex CHD require more frequent follow-up during pregnancy and usually deliver at a tertiary care center with experience in heart disease and pregnancy. Women with very high maternal mortality risk during pregnancy, such as those with Eisenmenger syndrome or Marfan syndrome with aortic dilation > 4.5 cm, should be educated about the risks of pregnancy, and termination of pregnancy should be discussed.[41]

Women with intermediate and complex CHD should have serial cardiology visits during pregnancy the postpartum period. At each visit, a careful history should be performed to elicit symptoms of heart failure or arrhythmia. Symptoms of normal pregnancy, such as shortness of breath, increasing heart rate, and peripheral edema, can mimic cardiac symptoms and it may be difficult to distinguish between them. The details regarding follow-up for women with low to medium risk and high-risk pregnancies are included in Figure 6. Investigations that involve radiation, such as x-rays, catheterization, or CT, should not be withheld if indicated, but efforts should be made to minimize radiation and pursue other forms of imaging whenever possible. When necessary, cardiac MRI can be performed during pregnancy, but gadolinium should be avoided.

A clear and detailed plan for labour and delivery should be created by the cardiac, obstetrical, and anaesthesia teams and distributed to care providers. The plan should include the expected mode of delivery, anaesthesia plans, monitoring requirements and plans for management of complications should they occur. Vaginal delivery is preferred for most women with CHD. In selected patients (eg, those with Fontan circulation) for whom Valsalva manoeuvre is not recommended, regional epidural and an assisted second stage of labour may help to prevent complications. During labour, women with significant arrhythmia should be monitored with the use of telemetry. Intravenous fluid administration should be closely monitored in women at risk for developing heart failure. Women with potential for right-to-left shunting should have air filters on intravenous lines to prevent air embolization. After delivery, there may be significant fluid shifts and close attention must be paid to volume status to

Table 2. Cardiac Disease in Pregnancy (CARPREG) risk score: predictors of adverse maternal cardiac events during pregnancy in women with heart disease

Predictor	Points
Systemic ventricular dysfunction	1
Left heart obstruction*	1
Prior cardiac event or arrhythmia	1
New York Heart Association functional class > II or cyanosis	1

Risk of adverse maternal cardiac events stratified according to the CARPREG risk score: risk score 0 = 5% adverse cardiac event rate; risk score 1 = 25% adverse cardiac event rate; risk score > 1 = 75% adverse cardiac event rate.

* Left heart obstruction = mitral valve area of < 2 cm^2, aortic valve area of < 1.5 cm^2, or peak left ventricular outflow gradient > 30 mm Hg. Systemic ventricular dysfunction = systemic ventricular ejection fraction < 40%. Data from Siu et al.[34]

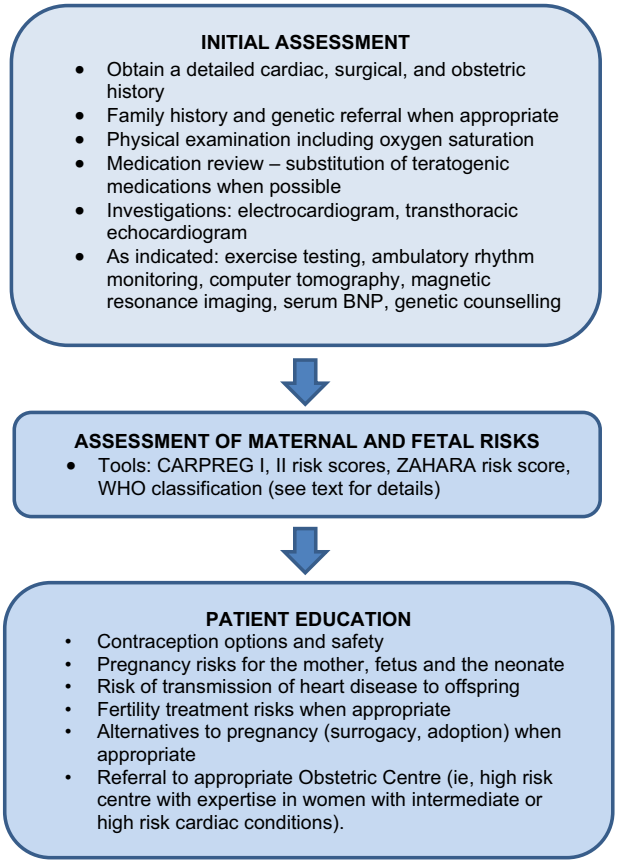
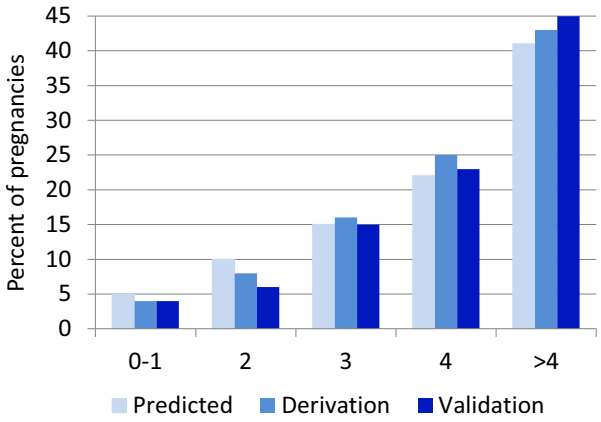


Figure 3. Preconception counselling recommendations for women with congenital heart disease.



PREDICTOR	POINTS
Prior cardiac events or arrhythmias	3
Baseline NYHA III-IV or cyanosis	3
Mechanical valve	3
Ventricular dysfunction	2
High risk left-sided valve disease/left ventricular outflow tract obstruction	2
Pulmonary hypertension	2
Coronary artery disease	2
High risk aortopathy	2
No prior cardiac intervention	1
Late pregnancy assessment	1

Figure 4. Risk predictors and frequency of adverse maternal cardiac events according to the CARPREG II risk score. Predicted (**light blue**) and observed frequencies of cardiac events in the derivation (**medium blue**) and validation group (**dark blue**). Reproduced from Silversides et al.[48] with permission from Elsevier.

prevent congestive heart failure. Although endocarditis prophylaxis is not recommended for uncomplicated vaginal deliveries, according to the American Heart Association guidelines on the prevention of infective endocarditis,[53] many experts continue to administer antibiotic prophylaxis for patients at high risk, such as those with prosthetic valve or previous endocarditis.

Standard postnatal care is suitable for most women with heart disease. However, depending on their risk profile or clinical course during pregnancy, some women may require an extended period of monitoring for hemodynamic instability after delivery. For example, women with significant pulmonary hypertension are at high risk for complications early after delivery and should be monitored in hospital for at least 7 days after delivery.

There are few contraindications to breastfeeding in women with CHD, although some experts advise against breastfeeding in women with severe systemic ventricular dysfunction due to the increased metabolic demands.[50] Many medications enter the breast milk in small amounts but are not necessarily contraindicated in breastfeeding. More lipid-soluble medications are excreted in larger amounts in the breast milk.[54] Detailed information on common medications used in the CHD population and their suitability for breastfeeding mothers is published elsewhere.[50]

Fertility treatments and alternatives to pregnancy

Some women with CHD may have difficulties conceiving. Rates of fertility likely vary by cardiac lesion, but there is little data in the literature with which to counsel women.[55] It is important to thoroughly investigate and treat standard causes of subfertility or infertility, and referral to a specialist in fertility is mandatory. However, some fertility tests, such as a hysterosalpingography, can result in a vagal reaction that may be life threatening in some patients with CHD.[55] This and other invasive procedures may need to be performed with appropriate monitoring, pain control, and clinical support. Assisted reproductive technologies (ART) include ovarian stimulation, in vitro fertilization, and intracytoplasmic sperm injection. Complications of ART include ovarian hyperstimulation syndrome and thromboembolism, and these can be particularly serious in women with CHD.[55-58] Multifetal pregnancy increases the hemodynamic stress on the heart, and the safety of implanting of more than 1 embryo is an important consideration in this population.[55-58] In addition, in the general population there is an increased risk of prematurity with ART, low-birth-weight babies, and congenital malformations, including cardiac defects.[58] These may be more significant in the CHD population who already have higher risks for these complications.

Surrogacy and adoption represent options for women in whom the risk of pregnancy is unacceptably high. There are 2 forms of surrogacy: "host" surrogacy where the eggs and sperm are obtained from the genetic parents and the resulting embryos transferred to the surrogate, and "straight" surrogacy which uses the surrogate mother's eggs. If opting for the former, patients and fertility physicians need to be made aware of the risks accompanying egg harvesting. Finally, discussion surrounding the preselection of embryos may arise in cases where there is a known genetic mutation.

Contraception

Contraceptive counselling is an essential part of the care of the adolescent and adult woman with CHD. Not only can an unanticipated pregnancy potentially lead to adverse maternal and neonatal outcomes, but certain contraceptives can themselves be harmful in selected cardiac populations. Despite this, in a study of women with CHD, only 51% could remember receiving information from a health professional about birth control.[10]

Barrier methods, such as a condom or a diaphragm, are safe for women with CHD, but their failure rates are high (13%-17% or higher) and dependent on appropriate use.[33,59] They are not recommended as the sole method of contraception if preventing pregnancy is the goal. Options for oral contraceptives include combined estrogen-progesterone and progesterone-only preparations.[60] The combined formulations, especially those with higher proportions of estrogen, increase the risk of thromboembolic complications and are therefore not recommended in patients with Eisenmenger syndrome, prosthetic valves, ventricular dysfunction, or Fontan

Figure 5. The cardio-obstetrics team. See Davis and Walsh[52] for more details on the cardio-obstetrics team.

circulation.[41,60] The oral progesterone-only formulations mitigate the risk of thromboembolism, but they medications have a higher failure rate (7%) and are therefore not advised for women in whom prevention of pregnancy is important.[59] Other forms of hormonal contraception, such as depot medroxyprogesterone acetate or progesterone-releasing intrauterine device (IUD), are often good alternatives for women with CHD. Vagal reactions can occur in women at the time of IUD insertion,[9] and this can result in serious complications in those with Fontan circulation or pulmonary vascular disease. In the high-risk CHD patient, IUD insertion should be performed by a physician with knowledge of CHD and at a site where the complication can be treated if necessary. Antibiotic prophylaxis is not required before IUD insertion, but screening for sexually transmitted infections may be done in high-risk patients.[61]

Tubal ligation may be recommended for some high-risk women; however, the risk of anaesthesia in women with high-risk cardiac lesions (eg, Eisenmenger syndrome) is not inconsequential and this procedure should only be performed at centres with expertise. Alternatively, hysteroscopic sterilization with the use of a microinsert device in the fallopian tubes has been used safely in this population.[62]

Late Adulthood

Little is still known about the long-term impact of pregnancy on the diseased heart or overall prognosis, yet this is perhaps one of the most significant areas of concern for women with CHD and their families. In general, most women with CHD can tolerate a pregnancy without complications, but the late effects of pregnancy on their overall health, quality of life, and life span remain unknown.

There are small studies showing a negative impact of pregnancy on the subaortic right ventricle (RV) and the aorta. For example, several studies have reported on adverse late outcomes in women with atrial switch repairs for transposition of the great arteries after pregnancy. This group of women have subaortic RVs that may poorly tolerate the hemodynamic stress of pregnancy.[63,64] The first study of 28 pregnancies in 16 women with atrial switch operations reported a decline in

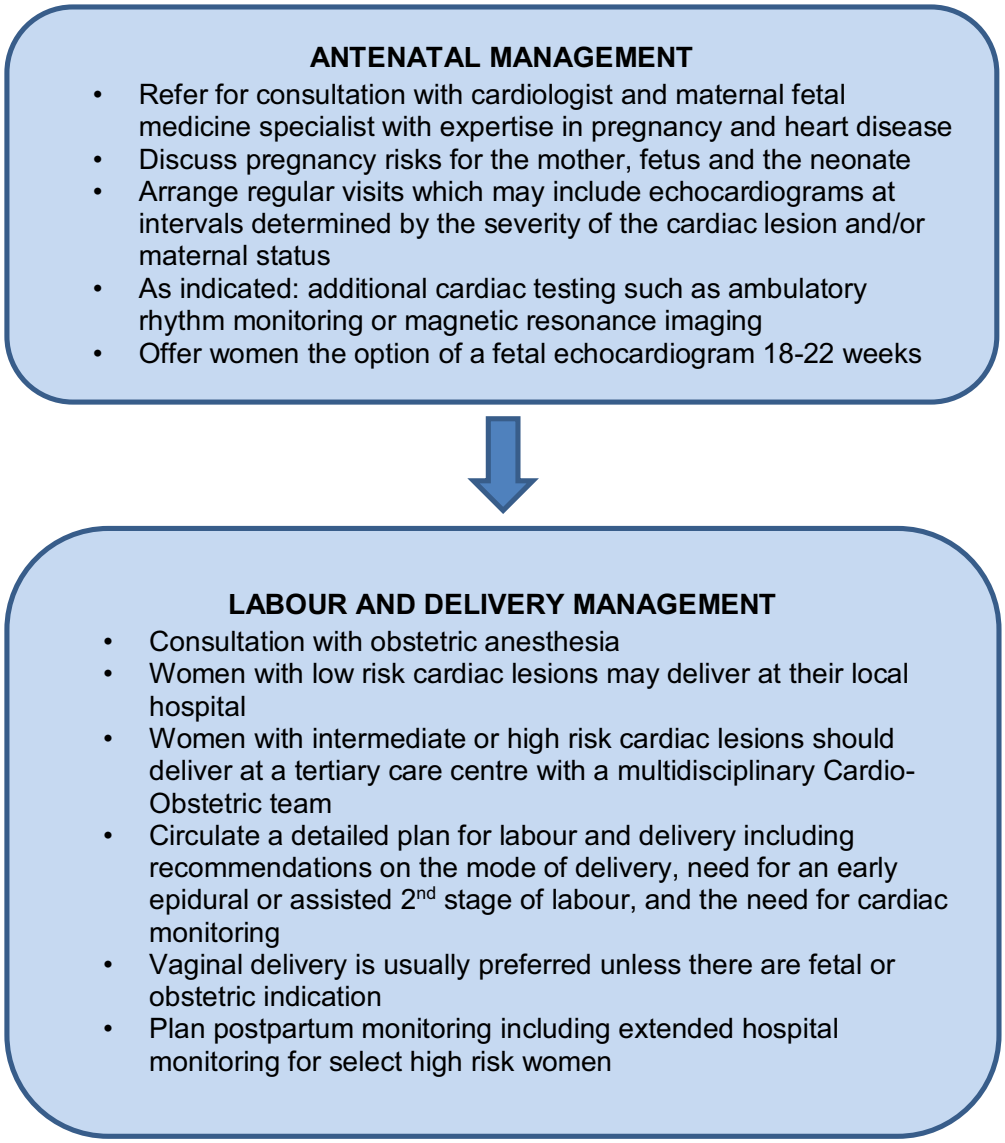


Figure 6. Pregnancy management recommendations for women with congenital heart disease

subaortic RV function in 4 women (25%), and in 3 of those 4 patients the subaortic RV function did not recover.[64] Eight women (50%) also had progression in the degree of tricuspid regurgitation, and in 3 it did not return to baseline.

Conflicting reports exist regarding the effect of pregnancy on the subpulmonary ventricle in tetralogy of Fallot. Several studies have demonstrated increases in RV dimensions in women after a pregnancy compared with control women.[65,66] A small study of 33 pregnancies demonstrated an increased late risk of death, stroke, ventricular dysfunction, and unanticipated surgery in women who had undergone pulmonary valve surgery.[67] In contrast, other groups have found no effect of pregnancy on RV volumes in women with tetralogy of Fallot.[68]

In a study of women with Marfan syndrome, aortic dissection and aortic surgery were higher in women who had undergone a pregnancy compared with nulliparous control women.[69] The number of pregnancies and the lack of beta-blocker use during pregnancy was associated with adverse long-term outcomes.[69] However, other groups have not demonstrated an impact on the rate of aortic root expansion in long-term follow-up for women who had undergone pregnancy.[70]

Long-term cardiovascular risks

Complications of pregnancy such as preeclampsia, miscarriage, and preterm delivery have recently become recognized as a risk factor for later cardiovascular complications.[3] Women with frequent miscarriages and preeclampsia have impaired vascular function and a greater risk of subsequent cardiovascular disease.[71-73] Women who give birth prematurely have a nearly 2-fold greater long-term risk of cardiovascular disease.[74,75] Many of the same risk factors, such as obesity, smoking, and hypertension, that predispose one to cardiovascular complications also predispose to obstetrical

complications. What remains unclear is whether these pregnancy complications unmask an underlying predisposition toward cardiovascular disease or whether the endothelial dysfunction that occurs as a result of the placental derangement increases later risk. Because women with CHD have a higher risk of obstetrical and fetal complications, close attention needs to be paid to their pregnancy history in evaluating cardiovascular risk.

Older Age

Menopause

Very little data are available regarding menopause in the CHD population. As increasing numbers of women with CHD age, physicians will need to be aware of the physiologic and psychologic implications of menopause and options for treatment.

There has been controversy over the past 2 decades regarding hormone replacement therapy (HRT) in menopausal or postmenopausal women. The Women's Health Initiative, published in 2002, raised alarm in the medical community when it demonstrated an increased risk of coronary disease and breast cancer in women in the combined estrogen and progestin treatment arm.[76] Since then, trials have demonstrated a protective effect of HRT in younger women (aged 50-59 years) or those less than 10 years after menopause onset. Guidelines now suggest that HRT is effective and safe for preventing the symptoms of menopause when prescribed at or around the onset of menopause.[77] There are no data regarding safety of HRT in the CHD population, but as with contraceptives, estrogen remains a risk factor for thromboembolism. Transdermal estrogen therapy has been thought to have a lower risk of thromboembolism, but the data in this population are still lacking.[9]

Geriatric adult CHD

The number of adults over the age of 65 years with CHD is on the rise worldwide with admissions in this group becoming more frequent and cost intensive.[78-80] The geriatric adult CHD (ACHD) population is one whose medical needs have yet to be clearly defined. On one hand, they present with complications from ACHD requiring specialized care. As surgical techniques are modified (eg, the increasing trend toward valve-sparing surgery in tetralogy of Fallot or the use of transcatheter interventions) the presentation and needs concerning those complications will change. On the other hand, acquired medical conditions such as dementia, chronic kidney disease, and cardiovascular disease are also important variables effecting morbidity and mortality.[79] The geriatric ACHD population will increase with time.[79] Early attention should be paid toward assessing and preventing cardiovascular risk in the ACHD population. Adult women with acquired cardiovascular disease have been found to have poorer outcomes after coronary bypass surgery and recently a similar disparity has been found among women undergoing aortic, tricuspid, and mitral valve surgery.[81-86] The reasons behind this disparity are not entirely clear but may involve atypical presentations of cardiac symptoms in this population.

Conclusion

For women with CHD, there are many facets of sexual and reproductive health that require attention. Research in this area is hampered by several factors. First, both surgical and medical therapies for CHD patients are constantly evolving and long-term outcomes continue to change. Second, studies in this area are often limited by small numbers of both patients and appropriate control subjects. And third, given the heterogeneity of CHD and individual presentation, it can be difficult to elicit which outcomes are attributable to the hemodynamic effects of pregnancy. Overall, given the limited nature of the literature in this area, there remains a significant need for long-term, prospective, multicentre case-control studies in this field.

Starting from birth, the life experience of women with CHD is shaped by their disease and the host of unique issues carried with it. Awareness of the challenges faced by women with CHD is essential for both pediatric and adult practitioners caring for these patients. Pregnancy can no longer be seen as an event that occurs in isolation, disconnected from adolescence and older age, but as a stage in the continuum of a life span that has potential implications for both long-term health and future generations.

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