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THE PRESENT AND FUTURE

STATE-OF-THE-ART REVIEW

High-Risk Cardiac Disease in Pregnancy

Part II

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ABSTRACT

Heart disease continues to be the leading cause of nonobstetric maternal morbidity and mortality. Early diagnosis and appropriate care can lead to prevention of complications and improvement of pregnancy outcome. This paper continues the review and provides recommendations for the approach to high-risk cardiovascular conditions during gestation. (J Am Coll Cardiol 2016;68:502–16) © 2016 by the American College of Cardiology Foundation.

COMPLEX CONGENITAL HEART DISEASE

Due to the successes of congenital heart surgery, congenital heart disease (CHD) now comprises up to 80% of all pregnancies in women with heart conditions in the Western world (1,2). The maternal risk of mortality (0.5%) and morbidity is, however, relatively low and is 4 to 5 times lower than that reported in valvular heart disease or cardiomyopathy (3). Maternal morbidity (mainly arrhythmias and heart failure [HF]) is reported in 11% (4.5% to 20%) (4–8). These data are from heterogeneous populations, varying from the very simple to the most complex CHD. There are subcategories of CHD in which the risk for both mother and fetus is markedly increased. Women with CHD are often not aware that their residual lesions are associated with increased pregnancy risk. Therefore, timely counseling is important in girls and women with CHD (9). To counsel each individual woman appropriately, the risk of pregnancy needs to be assessed (**Central Illustration**). As outlined before, the most reliable system of risk estimation in CHD is the modified World Health Organization classification of maternal risk (10), and this risk estimation system is recommended by the European Society of Cardiology (ESC) (11) (see Table 2 in Part 1 of the review [12]). According to this classification, the Fontan circulation, systemic right ventricle (RV), and uncorrected cyanotic disease are high-risk congenital conditions. This part of the review will focus on these conditions.

FONTAN CIRCULATION

Patients born with a functionally univentricular heart are often palliated by creation of a modification of the Fontan circulation. In all modifications of the Fontan circulation, the single ventricle is used as a systemic ventricle and pumps highly saturated blood in the aorta, whereas deoxygenated blood flows passively from the systemic veins to the lungs. This circulation is characterized by elevated systemic venous pressures, increased venous thrombotic risk, susceptibility for atrial arrhythmias that are often poorly tolerated, and impaired ability to meet demands for increased cardiac output related to decreased preload of the ventricle. Additionally, dysfunction of the single ventricle, valvular dysfunction, and protein-losing enteropathy are not uncommon complications. Only a limited number of pregnancies in

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Fontan women have been reported. A review of 71 pregnancies and a study of 59 pregnancies both describe an increased prevalence of infertility (57% and 21%) and a high miscarriage rate (34% and 27%) (13,14). Maternal complications occur in about 10% (14). Atrial arrhythmias are the most frequent complications (13,14). Thromboembolic complications have been reported in patients both with and without anticoagulation therapy (14). Other cardiac complications are HF and deterioration of functional class. HF occurs when the often anatomically and functionally abnormal ventricle cannot accommodate the requirement for increased cardiac output, and may be aggravated by atrioventricular valve regurgitation. Additionally, the passive transpulmonary circulation depends on adequate diastolic ventricular properties and may be insufficient to transport the increased plasma volume. There is a strikingly high incidence of premature delivery (39% and 69%), often in the setting of spontaneous pre-term labor, with about one-third of premature neonates born before 33 weeks of gestation (13,14). Experts agree that Fontan patients with depressed ventricular function, cyanosis, significant atrioventricular valve regurgitation, or protein-losing enteropathy should be advised against pregnancy (11). In others, careful pre-pregnancy counseling and planning of management in an experienced tertiary center is essential. Even though there are insufficient data to prove their effects, the prothrombotic state of both the Fontan circulation and pregnancy, the potentially disastrous effect of pulmonary embolism in a passive pulmonary circulation, and reports of thromboembolic complications in several patients are reasons to offer anticoagulation therapy to all pregnant Fontan patients, in line with the recommendations in the ESC guidelines (11). Sustained atrial arrhythmias constitute an emergency and usually require prompt electrical cardioversion (15). Vaginal delivery is usually preferred. Careful fluid management is essential around delivery to avoid reduction of pre-load, as well as worsening of HF. Neuraxial anesthesia is advisable to decrease the stress of delivery. Sudden decreases in systemic arterial resistance should be avoided, and coagulation abnormalities should be ruled out. An increase in pulmonary vascular resistance should also be avoided: prostaglandin F analogs should not be used for the management of postpartum hemorrhage, and when general anesthesia is necessary, ventilation should be with low airway pressures (11,16,17). Oxytocin should only be given as a continuous infusion. When blood loss is accompanied by hypotension, judicious fluid replacement is indicated. In women with abnormal ventricular function or atrioventricular valve regurgitation, post-partum administration of furosemide should be considered.

SYSTEMIC RV

Women with a systemic RV are those who have undergone an atrial repair (Mustard or Senning correction) of complete transposition of the great arteries and those with congenitally corrected transposition of the great arteries (CCTGA). More than 200 pregnancies in women with a Mustard or Senning repair have been described. Salient outcomes are a high miscarriage rate (up to 30%) and high maternal cardiac complication rates (10% to 30%), including arrhythmias and HF. Cardiac death appears to be rare. Other complications are New York Heart Association (NYHA) functional class deterioration, RV dysfunction and worsening of tricuspid regurgitation (TR) that may be persistent after pregnancy, high rates of prematurity (25% to 50%) and small for gestational age (up to 50%), as well as fetal and neonatal death (18–24). Similar complications are described in women with CCTGA, but the complication rates are reported to be lower (25,26). Women with both a Mustard or Senning operation and CCTGA need to be counseled before pregnancy. Severe RV dysfunction or TR is a reason to advise against pregnancy (11). Management of pregnancy should be in specialized centers. Vaginal delivery is usually appropriate. Arrhythmias are primarily treated with beta-blockers, but caution is necessary because of the tendency for bradycardia due to sinus node dysfunction (Mustard/Senning) or atrioventricular block (CCTGA). Frequent surveillance of RV function, heart rhythm, and clinical symptoms is recommended during pregnancy. When deterioration of RV function is noted, early delivery is advised.

UNCORRECTED CYANOTIC HEART DISEASE WITHOUT PULMONARY HYPERTENSION

Cyanotic heart disease is usually treated surgically in childhood. Limited data are available regarding pregnancy in women with inoperable or palliated cyanotic heart disease with no pulmonary hypertension. Cardiac complications have been described in 32% (26). More than 50% of all complications are due to HF; other complications include thromboembolic events, arrhythmias, and endocarditis (27,28). Fetal outcome is associated with maternal oxygen saturation at rest; with saturation ≥90%, the live birth rate is 92%, whereas with a saturation ≤85%, the live birth rate is only 12% (26). To maintain the highest possible

ABBREVIATIONS AND ACRONYMS

BAV = bicuspid aortic valve
CHD = congenital heart disease
DCM = dilated cardiomyopathy
HF = heart failure
LV = left ventricular
LVEF = left ventricular ejection fraction
MFS = Marfan syndrome
PPCM = peripartum cardiomyopathy
RV = right ventricle/ventricular