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Adult Congenital Heart Disease (A Bhatt and K Niwa, Section Editors)

Hemodynamically Significant Congenital Cardiac Lesions in Pregnancy

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Opinion statement

The incidence of congenital cardiac disease among reproductive-aged women is increasing. Understanding the unique physiology of pregnancy and the postpartum period is critical to helping women achieve successful pregnancy outcomes. Risk assessment models estimate the cardiac, obstetric, and neonatal risks a woman may face and influence the conversations regarding pregnancy and contraception management. This review focuses on some of the most common congenital cardiac lesion encountered during pregnancy, as well as key aspects of antepartum, intrapartum, and postpartum care for these women. A multidisciplinary team, with Maternal-Fetal Medicine, Cardiology and Obstetric Anesthesiology specialists, is critical to the care of these patients.

Introduction

The incidence of congenital heart disease at birth is approximately 1 % [1]. With improved treatment in infancy and childhood, more patients are living into adulthood, thus knowledge of the impact of pregnancy on congenital heart disease and vice versa is critical to obstetricians, cardiologists, anesthesiologists, and women themselves. The most common congenital heart disease lesions in pregnancy are aortic stenosis (20 %), ventricular septal defect (13 %), tetralogy of Fallot (12 %), atrial septal defect (9 %), pulmonary stenosis (8 %), coarctation of the aorta (8 %), and transposition of the great vessels (5 %) [2]. This review focuses on risk stratification and management of the most prevalent and hemodynamically significant congenital lesions in the antepartum, intrapartum, and postpartum periods.

Hemodynamics of pregnancy

Increased cardiac output is the final common pathway of the multiple hemodynamic changes that occur in pregnancy. Total blood volume increases 50 % to a maximum of 4700–5200 cc in the early third trimester, resulting in increased preload and stroke volume. This increase is attributed to induction of the renin-angiotensin-aldosterone system, which stimulates sodium and water retention. The addition of the parallel uteroplacental circulation and the vasodilatory effect of progesterone cause systemic vascular resistance (SVR) to fall. SVR begins to decrease as early as 5 weeks gestation, nadirs between 14 to 24 weeks gestation, then slowly rises as pregnancy progresses to term. The trend in mean arterial pressure across pregnancy parallels SVR, with an initial decrease by 7 weeks gestation, a nadir from 24 to 32 weeks gestation, then a gradual rise to approach non-pregnant levels near term. Heart rate increases by 20 % in pregnancy, with most of the increase occurring in the first 16 weeks of pregnancy, and the remainder in the late third trimester, to a maximum of 15–20 beats per minute above baseline. This increase may be mediated by the decrease in SVR. Cardiac output therefore increases by 50 % and reaches its peak by 16 weeks gestation, with a small fall at term. The early increase in stroke volume maintains the increased cardiac output early in pregnancy; near term, the increase in heart rate maintains cardiac output [2, 3, 4•]. These changes are illustrated in Fig. 1.

The impact of labor on maternal hemodynamics is significant. Pain associated with contractions results in increased heart rate and sympathetic tone, which increases cardiac contractility and results in peripheral vasoconstriction and increased afterload. Uterine contractions autotransfuse 300–500 cc of

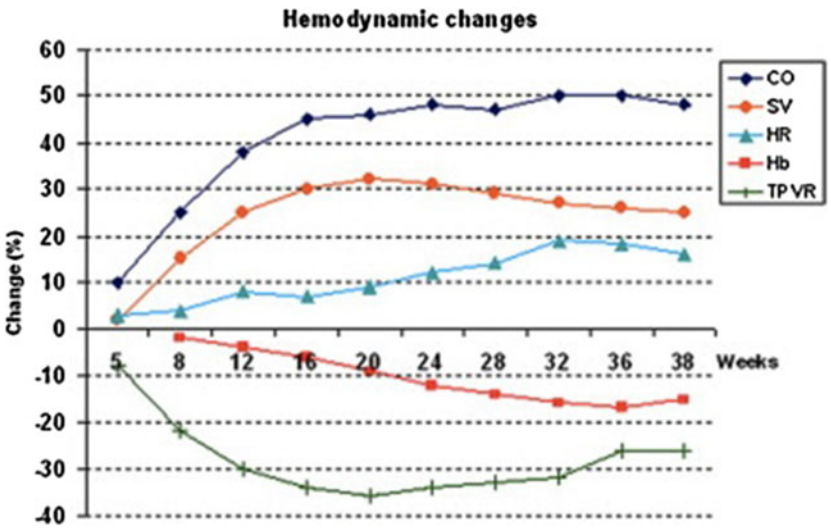

Fig. 1. Hemodynamic changes in normal pregnancy. Reprinted from the Journal of Cardiology, Vol 61, Ruys TPE, Cornette J, Roos-Hesselink JW, Pregnancy and delivery in cardiac disease, 107–112. Copyright 2013, with permission from Elsevier [37].

blood per contraction, thus increasing preload. The net effect is an increase in cardiac output of 30 %. As labor progresses, cardiac work increases [2, 3, 4•]. The second stage of labor requires repeated Valsalva maneuvers to assist in fetal descent and expulsion, which decrease venous return, resulting in decreased stroke volume and compensatory tachycardia.

In the immediate postpartum period, several events occur which influence maternal hemodynamics and increase the risk of cardiac decompensation: (1) the rapid involution of uterus and associated autotransfusion of 500 cc of uteroplacental blood; (2) the release of caval compression, with immediate increase in venous return; (3) bleeding associated with placental delivery; and (4) immediate increase in SVR due to loss of uteroplacental circulation. Cardiac output increases 60–80 % immediately after delivery and normalizes by 2 weeks postpartum. Heart rate remains elevated for 24 h postdelivery, and then declines to prepregnancy ranges within 10 days. Blood pressure and blood volume also return to prepregnancy levels by 10 days postpartum [2, 3, 4•]. Of note, extravascular fluid is mobilized between 2 and 4 days postpartum, posing a higher risk for pulmonary edema after the immediate postdelivery hours.

Hemodynamic changes in pregnancies complicated by structural heart disease are not as well studied. One study of 29 patients with primarily congenital heart disease revealed less drastic increases in stroke volume and cardiac output. Diastolic dysfunction becomes more apparent, as the increased volume load of pregnancy cannot be compensated for with myocardial hypertrophy; therefore, increased filling pressures are noted. At 6 months postpartum, decreased systolic function and diastolic dysfunction persist [5••].

Stratification of pregnancy-related risk

Three risk assessment models have been developed to stratify the maternal, and in some cases, neonatal risks associated with pregnancy among women with cardiac disease: CARPREG, the modified World Health Organization (WHO) model, and ZAHARA (Table 1).

Maternal outcomes

The CARPREG model was developed to identify predictors of new-onset heart failure, symptomatic arrhythmia, stroke, or cardiac death. The model has been prospectively validated among 546 pregnant women with cardiac disease, 74 % of whom had congenital heart disease, and the following independent predictors were identified: prior cardiac event or arrhythmia, New York Heart Association (NYHA) greater than class II or baseline cyanosis, left heart obstruction, and myocardial dysfunction defined as ejection fraction (EF) <40 %. Each predictor in the model receives 1 point. The risk of a cardiac event associated with a score of 0 is 4 %; score of 1, 27 %; score >1, 62 %. Maternal stroke or cardiac death occurred in 1 % of pregnancies [6].

Table 1. Risk-based stratification models: CARPREG, WHO, and ZAHARA

	CARPREG	WHO	ZAHARA
Inputs			
	(1) Myocardial dysfunction, as defined by EF < 40 %, hypertrophic cardiomyopathy, or restrictive cardiomyopathy	*WHO 1* Uncomplicated, small or mild Pulmonary stenosis, Ventricular septal defect, Patent ductus arteriosus, or Mitral valve prolapse with no more than trivial mitral regurgitation Successfully repaired simple lesions, e.g., Ostium secundum atrial septal defect, Ventricular septal defect, Patent ductus arteriosus, or Total anomalous pulmonary venous drainage	Moderate-to-severe pulmonary or systemic atrioventricular valve regurgitation (0.75 points per valve affected)
	(2) Left heart obstruction, as define by aortic valve area <1.5 cm^2, mitral valve area <2.0 cm^2, or left ventricular outflow tract peak gradient of >30 mmHg	*WHO 2* Unrepaired ASD Repaired TOF Most arrhythmias	Cyanotic heart disease (1 point)
	(3) Prior cardiac event or arrhythmia	*WHO 2–3*[a] Heart transplant Marfan syndrome without aortic dilation Hypertrophic cardiomyopathy Mild LV impairment Native or tissue valvular Heart disease, not WHO 4 *WHO 3* Mechanical valve Systemic right ventricle Post Fontan Cyanotic heart disease Other complex congenital cardiac disease	Mechanical valve (4.25 points)

Table 1. (Continued)

	CARPREG	WHO	ZAHARA
	(4) NYHA > Class II or baseline cyanosis	*WHO 4* Marfan's syndrome with aorta dilated > 40 mm Severe left heart obstruction Previous PPCMP with any residual left heart dysfunction Pulmonary hypertension NYHA Class III–IV or EF < 30 %	History of arrhythmia (1.5 points) NYHA > Class II (0.75 points) Left heart obstruction, including either peak aortic gradient > 30 mmHg or aortic valve area <1.5 cm^2, or mitral valve area <2.0 cm^2 (2.5 points) Use of cardiac drugs in pregnancy (1.5 points)
Scoring criteria	Each factor received 1 point	Not applicable	See scoring criteria above
Risks of adverse maternal outcomes			
	0—risk 4 % 1—risk 27 % >1—risk 62 %	WHO 1—risks comparable to general population WHO 2—small increased risk of maternal morbidity/mortality WHO 3—significant increased risk in maternal morbidity/mortality & expert care required WHO 4—pregnancy contraindicated; very high risk of maternal mortality or severe morbidity. Termination should be discussed.	0–0.5 points—risk 2.9 % 0.51–1.5 points—risk 7.5 % 1.51–2.5 points—risk 17.5 % 2.51–3.5 points—risk 43.1 % >3.5 points—risk 70 %

[a]Classification as WHO 2 or WHO 3 depends on comorbidities

In 2006, the WHO modified its classification of risk associated with contraceptive options among women with cardiac disease to reflect risks of pregnancy among these women. Cardiac lesions of varying hemodynamic and structural significance have been classified into four broad categories (WHO 1–4) and are shown in Table 1 [7•].

The ZAHARA model retrospectively investigated predictors of adverse cardiac outcomes among 1302 pregnant women with congenital heart disease specifically. Adverse outcomes occurred in 7.6 % and were defined as significant arrhythmia or heart failure, cardiovascular complication such as venous thromboembolic disease, myocardial infarctions, and/or stroke, and endocarditis up to 6 months postpartum. Independent predictors included moderate-to-severe pulmonary or systemic atrioventricular valve regurgitations, repaired or unrepaired cyanotic heart disease, mechanical valve, history of arrhythmia, NYHA class >II, left heart obstruction, and use of cardiac drugs. Each of these was variably weighted, and the additive score predicts the adverse outcome (see table) [8].

Subsequent to their development, several studies have compared the relative performance of the models in predicting adverse cardiac events. The ZAHARA investigators validated both the CARPREG and ZAHARA scores in an independent prospective cohort of 213 women with congenital heart disease. Both CARPREG and ZAHARA models overestimated adverse outcomes, and the modified WHO model had the best predictive power [9]. Additional studies have confirmed the superiority of the modified WHO model [10, 11].

The European Society of Cardiology (ESC) recommends the use of the modified WHO criteria to risk stratifying women with congenital heart disease, primarily because only this model incorporates disease states in which pregnancy is directly contraindicated [12]. The American College of Cardiology, American Heart Association, American Congress of Obstetricians and Gynecologists, and the Society for Maternal-Fetal Medicine have no specific statements regarding risk stratification and pregnancy management in this population.

Neonatal outcomes

Among women in the CARPREG cohort, adverse neonatal outcomes occurred in 20 % of pregnancies, the majority of which were attributable to premature birth. Predictors of adverse neonatal outcomes included NYHA class >II or baseline cyanosis, left heart obstruction, smoking during pregnancy, multiple gestation, and use of anticoagulants. Presence of any single risk factor increased the likelihood of perinatal death from 2 to 4 %. Incidence of neonatal congenital heart disease among women with congenital heart disease was 7 % in this cohort [6].

In the ZAHARA cohort, adverse neonatal outcomes occurred in 25 %, again predominantly attributable to prematurity. The incidence of perinatal death was 4 % [8]. Predictors of adverse outcomes were largely similar to those defined by the CARPREG study. The WHO model does not address neonatal outcomes.

Review of Specific Lesions (by WHO category)

WHO class 4 congenital heart disease

Aortic stenosis

Congenital aortic stenosis (AS) most commonly occurs in the context of a bicuspid aortic valve. As heart rate and stroke volume increase in pregnancy, patients with the fixed cardiac output state of AS are at increased risk of pulmonary edema. Adequate stroke volume is necessary to achieve adequate cardiac output. AS is, therefore, preload dependent. A small loss in filling volume results in a large loss in filling pressure and cardiac output, given a steep volume-pressure curve. Patients with AS may have left ventricular hypertrophy, a compensatory mechanism to generate adequate force across the obstruction. As such, wall tension and myocardial oxygen demand are increased [13].

The WHO considers pregnancy to be contraindicated in severe AS, defined as aortic valve area of <1 cm^2 or mean gradient of >40 mmHg. Bicuspid aortic valve has also been associated with aortic dissection, particularly with an enlarged aortic root [14]. AHA/ACC guidelines recommend caution regarding pregnancy with aortic root diameter >45 mm [15].

In one of the largest series of 58 pregnancies among 35 women with congenital AS, 45 % of whom had undergone prepregnancy repair, cardiac and obstetric outcomes were favorable overall. Cardiac events occurred in 9 %; among those with severe AS, heart failure was the predominant complication. Those with mild-to-moderate AS experienced declines in NYHA class and arrhythmias. All women with declines in NYHA class required aortic valve replacement within 6 to 48 months postpartum. Regarding obstetric outcomes, the incidence of hospitalization for hypertensive disorders of pregnancy was 11 %, which was higher than in the general US population [16, 17]. Spontaneous preterm labor was more than twice as common in women with severe AS as the general population [16, 18]. Among women who underwent a trial of labor, 15 % had an intrapartum cesarean delivery, and 28 % had an operative vaginal delivery. Perinatal outcomes were overall favorable, with 13 % of neonates born prematurely, 13 % born small-for-gestational age (the majority of whom were in the mild-to-moderate AS group), and 4 % with recurrent congenital heart disease. There were no perinatal deaths [16].

The antepartum management of AS includes heart rate control with beta-blockers to improve oxygen delivery to potentially hypertrophied myocardium and careful management of fluid status to maintain preload. Vasodilators are generally contraindicated except in the context of decompensated heart failure or severe systemic hypertension.
Intrapartum, for women with severe AS, preload can be affected by intravenous fluid administration, hypotension associated with neuraxial anesthesia, and hemorrhage. An arterial line and a pulmonary artery catheter is

beneficial in women with peak gradients >60 mmHg [2]. Delivery should be effected by 39 weeks for a well-tolerated pregnancy, and sooner as needed for maternal indications. It is preferable to avoid prolonged inductions of labor, but cesarean delivery is reserved for obstetric indications, or, per expert opinion, for aortic dissection, aneurysm, or critical AS [15]. Data regarding the management of the second stage of labor are limited, with operative vaginal delivery recommended by expert opinion. Given the rapid changes in hemodynamics up to 48 h postpartum, close monitoring should continue through this period. Women generally autodiurese, but diuresis can be assisted with furosemide as needed for symptomatic pulmonary edema, being mindful to maintain adequate preload to allow for coronary and cerebral perfusion in particular [2].

Mechanical aortic valve

Pregnancy after mechanical aortic valve replacement is considered WHO class 3. The primary management considerations include management of anticoagulation antepartum and intrapartum, which is beyond the scope of this review. Endocarditis prophylaxis is not recommended with mechanical valves [15].

WHO class 3 congenital heart disease

Transposition of the great vessels

Congenitally corrected transposition of the great vessels

In congenitally corrected transposition of the great vessels (also known as L-transposition), the morphologic right ventricle acts as the systemic ventricle. Long-term complications include hypertrophy, dilation, and heart failure of the right ventricle, with tricuspid regurgitation. In pregnancy, the decreased afterload results in fewer loads on the right ventricle, and consequently less tricuspid regurgitation. However, the increased volume load of pregnancy may result in right ventricular dilation and decreased function. Arrhythmias may occur due to atrial dilation as well as an inherently abnormal conduction system, including complete heart block in a small subset of patients [19]. In a large review of patients with congenitally corrected transposition, the incidence of arrhythmia and heart failure in pregnancy were 3.6 and 7.1 %, respectively, and in general, pregnancy is well-tolerated in this population [19, 20]. Obstetric outcomes studied include preterm labor, hypertensive disorders, and thromboembolic complications, the incidences of which were comparable to the general population. Rates of neonatal complications, including preterm delivery, small-for-gestational age, and perinatal mortality, were also comparable to the general population [20].

Repaired D-transposition of the great vessels

In D-transposition, the parallel circulation associated with the transposition was classically repaired with an atrial switch procedure (the Mustard or Senning procedure), which maintains the morphologic right ventricle as the systemic ventricle. The physiology and associated complications are similar to that described for congenitally corrected transposition, with the exception of

increased arrhythmogenicity due to surgical scarring.

In a systematic review of pregnant women with D-transposition, incidence of arrhythmia during pregnancy was 15.6 %, and that of heart failure, 10.8 %. The incidence of preeclampsia was elevated at 15 %, and preterm labor occurred in almost 30 % of patients with D-transposition, resulting in increased rates of preterm birth and small-for-gestational age neonates, as well as higher perinatal mortality, at least in part attributable to prematurity [20].

Since the 1980s, the arterial switch (the Jatene procedure) has become the procedure of choice for D-transposition, resulting in the morphologic left ventricle functioning as the systemic ventricle. In a series of 9 patients with D-transposition after arterial switch, all 13 pregnancies studied went to full-term. Two adverse cardiac events occurred: non-sustained ventricular tachycardia and mechanical mitral valve thrombosis in the context of suspension of anticoagulation peripartum. One patient also had progression of supravalvular pulmonic stenosis over the course of three pregnancies and required balloon valvuloplasty 1 year after her last pregnancy. While this is a small case series, these outcomes are favorable and improved as compared to the adverse outcomes seen after the atrial switch procedure. The authors hypothesize that the reduction in intraatrial manipulation and the use of the morphologic left ventricle as the systemic ventricle have resulted in fewer adverse events [21•].

As arrhythmia and heart failure are the primary adverse cardiovascular events, judicious management of fluid status and monitoring of symptomatic palpitations are paramount. Afterload reduction helps to decrease the load on the systemic right ventricle (for patients without an arterial switch) and is likely to attenuate the decline in right heart function, which may decrease the incidence of arrhythmia. In general, experience has demonstrated that arrhythmia is common and is frequently the cause of cardiac decompensation. Overall, D-transposition and congenitally corrected transposition are both well tolerated in pregnancy [2].

Fontan palliation

Fontan palliation overcomes congenital anomalies, such as tricuspid atresia, hypoplastic left heart, and other conditions characterized by a single systemic ventricle, by creating passive connections from the right heart to the pulmonary arterial circulation. In other words, there is no right heart pump, and the central venous pressure becomes the driving force for right-sided forward flow, with assistance from the negative intrathoracic pressure generated by inspiration [2, 22]. For those patients surviving to adulthood, Fontan repair predisposes to arrhythmias, systemic ventricular dysfunction, thromboembolic disease, and protein-losing enteropathy [22]. The increased volume load of pregnancy increases atrial stretch and thus arrhythmogenicity, resulting both in atrial arrhythmias and supraventricular tachycardia, as well as increases venous congestion.

Supraventricular arrhythmias are the most common adverse cardiovascular outcomes, followed by functional decline in NYHA Class, and no cardiac complications occurred among women who underwent a trial of labor [23, 24]. Adverse obstetric outcomes were far more common than adverse cardiac outcomes, driven primarily by the high incidence of prematurity. Seventy-eight percent of women had preterm births, and 48 % had small-for-gestational age

fetuses [23, 24]. In addition, the incidence of spontaneous abortion is high, with estimates of approximately 30 % [25, 26].

Given a fixed cardiac output, diuretics may be necessary to manage the increased volume load of pregnancy. Care should be taken to avoid interventions that may increase pulmonary vascular resistance. The procoagulant state of pregnancy combined with the low-flow state of the right heart increases the risk of venous thromembolism, and some experts suggest prophylactic anticoagulation in pregnancy [23].

WHO class 2 congenital heart disease

Unrepaired atrial septal defect

Atrial septal defects (ASDs) are the third most common cause of congenital heart disease and are the most common among reproductive-aged women [27, 28]. ASDs result in a left-to-right shunt and are often undetected prepregnancy, given the subtle physical exam findings and asymptomatic nature, despite high shunt fractions at times [3]. Long-standing large ASDs can lead to prepregnancy remodeling of the heart, with right atrial dilation, right ventricular hypertrophy, and possibly pulmonary hypertension, and can progress ultimately to Eisenmenger's syndrome. Closure of ASDs is straightforward and may be recommended before pregnancy, to decrease the risk of pulmonary hypertension developing or worsening over time [3].

Outcomes for women with unrepaired ASDs are overall favorable. In a series of 133 pregnancies among women with unrepaired ASDs, the incidence of arrhythmia was approximately 5 % and postpartum persistent decline in functional status was 3 %. The incidence of hypertensive disorders was 10 % and that of postpartum hemorrhage was 8 %, both of which are higher than the incidence in the general obstetric population; rates of prematurity were comparable to the general population, however. The incidence of small-for-gestational age was 21 %, and that of perinatal mortality was 3 % [29].

The management of ASDs in pregnancy largely depends on the presence or absence of heart remodeling. In the absence of heart remodeling, women with an unrepaired ASD tolerate pregnancy, labor, and the immediate postpartum period well without specific management of their hemodynamics. With the increased stroke volume of pregnancy, increased left-to-right shunt pulmonary flow may unmask symptoms of dyspnea and functional decline. Conditions that increase SVR, such as hypertensive diseases of pregnancy, will augment the left-to-right shunt and may also precipitate symptomatic dyspnea. Atrial arrhythmias are more likely to arise in women with a long history of a large, uncorrected ASD [3]. Filtered IVs are recommended for all patients to prevent paradoxical emboli.

Repaired tetralogy of Fallot

Tetralogy of Fallot is the most common cyanotic heart disease, with an incidence of 1 in 3600 live births. With refined surgical technique and repair in infancy or early childhood, survival to adulthood is almost universal. Residual

postsurgical deficits include pulmonic valve regurgitation, which is associated with progressive exercise intolerance, and right heart failure, as well as arrhythmias (due to the hemodynamic changes and surgical scars) and sudden cardiac death [30]. Late complications occur in 10–15 % of women with repaired tetralogy of Fallot, usually 20 years after repair [31]. Risks in pregnancy are higher in the presence of persistent right ventricular outflow tract (RVOT) obstruction, severe pulmonary regurgitation, tricuspid regurgitation, and right and left ventricular dysfunction postrepair, all of which predispose to right heart failure and arrhythmia [30]. Recent studies have demonstrated unfavorable right ventricular remodeling after pregnancy [32].

In a series of 157 pregnancies among 74 women in the Netherlands with repaired tetralogy of Fallot, the miscarriage rate was 19 %. Among those with completed pregnancies greater than 20 weeks gestation, 56 % had moderate or severe pulmonary regurgitations, 44 % had RVOT obstruction, and 20 % had tricuspid regurgitation; however, only 7 % of women were NYHA Class III or IV prepregnancy. Notably, 24 % of women smoked prepregnancy, and 16 % continued to smoke during pregnancy. Atrial and ventricular arrhythmias occurred in 6.5 % of patients; heart failure occurred in 1.6 % of patients in the third trimester, all among patients with severe pulmonic regurgitation. Prior pulmonic valve regurgitation, prior arrhythmia, and need for cardiac medications prepregnancy were most predictive of adverse cardiovascular events in pregnancy. Preeclampsia/gestational hypertension occurred in 8 % of the cohort, and postpartum hemorrhage occurred in 10 % of the cohort, both of which were more common than among a healthy population. The cesarean delivery rate was 20 %, which is much greater than the Dutch national average; maternal cardiac status was the indication for 20 % of the cesarean sections. The assisted vaginal delivery rate was 13 %, comparable to the Dutch national average. Finally, the rates of both prematurity and growth restriction were 18 %, perinatal mortality was 6 % (attributable to prematurity, congenital heart disease, or growth restriction), and the incidence of congenital heart disease in the newborn was 2.4 %. Adverse perinatal outcomes were more likely among women requiring cardiac medications prepregnancy. The impact of smoking was not discussed [33•].

In general, pregnancy with repaired tetralogy of Fallot without residual deficits is well tolerated, and minimal specific hemodynamic adjustments need to be made in the antepartum, intrapartum, or postpartum periods [2]. If pulmonary regurgitation is present, monitoring for arrhythmia and right heart failure is paramount, but no specific anticipatory management is recommended.

Notably, although tetralogy of Fallot can be sporadic, it is also associated with 22q11 microdeletion in approximately 25 % of patients, which poses different risks to offspring [30].

Hemodynamically insignificant lesions (WHO I)

Mitral valve prolapse, pulmonary stenosis, repaired or unrepaired ventricular septal defects, repaired ASDs (with no associated pulmonary hypertension), repaired or unrepaired patent ductus arteriosus, and repaired total anomalous

pulmonary venous return are all very well-tolerated lesions in pregnancy. Management of these lesions in the antepartum, intrapartum, and postpartum period mirrors standard obstetric management for low-risk patients.

General issues in antepartum, intrapartum, and postpartum management

Antepartum management

In addition to routine prenatal care, multidisciplinary care is the cornerstone of the management of women with congenital heart disease. In general, patients should see an adult congenital cardiologist at least every trimester with a maternal echocardiogram, with more or less frequent assessment as clinically indicated. A consultation with an obstetric anesthesiologist is recommended in the third trimester, or sooner if the patient is at high risk for preterm birth. A fetal echocardiogram is recommended to screen for fetal congenital heart disease in the middle of the second trimester. Serial antenatal ultrasounds to monitor adequate fetal growth in the third trimester are also recommended. Prevention of maternal anemia, with iron and folate supplementation, is recommended to decrease cardiac work.

Neuraxial anesthesia

Neuraxial anesthesia is an important consideration in the intrapartum management of parturients with congenital heart disease. As noted above, pain increases sympathetic tone and results in increased cardiac output. Sympathetic blockade occurs due to local anesthestic administration, resulting in arteriolar dilation and decreased afterload, as well as increased venous capacitance and decreased preload, with reflex tachycardia. To counteract this response, patients may require prehydration as well as slow titration of sympathectomy if their lesion is preload dependent. Spinal anesthesia produces a more profound sympathetic blockade than epidural anesthesia, and epidural or combined spinal-epidural anesthesia is recommended.

Administration of commonly used drugs in labor

Commonly used medications in labor may precipitate adverse events in patients with congenital heart disease. Terbutaline, a beta-agonist occasionally used to halt uterine contractions, results in significant tachycardia. Methylergonovine, an effective and rapid-acting ergot alkaloid for the management of postpartum hemorrhage, acutely increases systemic vascular resistance, and its use is strongly discouraged. Rapid infusions of oxytocin, used postpartum to prevent and treat postpartum hemorrhage, can precipitate acute hypotension. Hemabate, a prostaglandin used to treat postpartum hemorrhage, may result in tachycardia.

Endocarditis prophylaxis in labor

Although there is a transient bacteremia associated with delivery, this alone does not warrant routine endocarditis prophylaxis. According to the AHA/ACC guidelines, endocarditis prophylaxis is recommended only for patients with bioprosthetic valves or patients with unrepaired or palliated cyanotic heart disease [15].

Anticoagulation in labor

For patients who receive prophylactic anticoagulation during pregnancy, neuraxial anesthesia can be placed anytime after a dose of prophylactic unfractionated heparin, and 12 h after the last dose of prophylactic low-molecular weight heparin. For patients on therapeutic unfractionated heparin, neuraxial anesthesia can be placed once the PTT normalizes; for patients on therapeutic low molecular weight heparin, a delay of 24 h after the last dose is advised [34].

Mode of delivery

For the vast majority of congenital heart disease, cesarean delivery is reserved for obstetric indications. Expert opinion recommends cesarean delivery in certain limited circumstances, as highlighted above, such as avoiding the risk of aortic dissection, aneurysm, or critical AS [15]. In other cases, assisted vaginal delivery with forceps or vacuum is often recommended to minimize Valsalva; however, there is little evidence to guide these recommendations. A recent study demonstrated no value in a planned cesarean delivery (for either cardiac or obstetric indications) over a planned vaginal delivery for patients with congenital heart disease, despite the possibility of needing a cesarean delivery for intrapartum obstetric indications [35•].

Contraception counseling

Reproductive life planning, including pregnancy prevention if desired, enabling appropriate birth spacing and counseling about pregnancy risks in the preconception period, is important. In patients with congenital heart disease, this is particularly relevant. With respect to contraception, some methods pose risks of thromboembolic disease, cerebrovascular accident, and myocardial infarction and should be avoided in some patients at risk. It should be kept in mind, however, that these risks would be even higher if unintended pregnancy was to occur and thus, reliable contraception is paramount.

In general, the most effective forms of contraception include permanent sterilization and long-acting reversible contraception (progesterone-containing intrauterine device, progesterone-containing subcutaneous implant, and copper-containing intrauterine device), all of which result in annual pregnancy rates of less than 1 in 100. Progesterone depot injections every 3 months, daily combination oral contraceptives, weekly combined contraceptive patch, and monthly combined contraceptive vaginal ring result in annual pregnancy rates of 6–9 per 100 with typical use [36].

The aforementioned WHO model also classifies congenital cardiac lesions by the safety of combined hormonal contraceptives as well as progesterone-only contraceptives [7].

Conclusion

As reproductive-aged women with congenital heart disease contemplate pregnancy, knowledge of physiology and evidence-based counseling regarding

cardiac, obstetric, and neonatal outcomes is of paramount importance. Multidisciplinary care between maternal-fetal medicine specialists, cardiologists, and obstetric anesthesiologists, is of paramount importance to ensure optimal pregnancy outcomes. In general, most lesions are well-tolerated, particularly when women have optimal hemodynamics and functional status preconception.

Compliance with Ethical Standards

Conflict of Interest
Malavika Prabhu and Allison Bryant each declare no potential conflicts of interest.

Human and Animal Rights and Informed Consent
This article does not contain any studies with human or animal subjects performed by any of the authors.

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