Pregnancy heart team: A lesion-specific approach

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With 90% of children with congenital heart disease (CHD) surviving to adulthood,1 more females with CHD are living to childbearing age. Many of these women have varying degrees of residual lesions and/or altered physiology and require maintenance medications, with potential implications for pregnancy and lactation, as well as the possibility of further surgical interventions. Confronted with this burgeoning population approaching their reproductive years, cardiac surgeons must consider operative choices in the context of potential future pregnancies and also must be versed in situations warranting surgical intervention prior to conception and during pregnancy. The surgeon is a critical component of the pregnancy heart team.

Women with cardiac diseases including CHD who become pregnant or are contemplating pregnancy should be managed by an experienced pregnancy heart team (Figure 1). This team involves specialists from adult congenital cardiology, cardiovascular imaging, interventional cardiology, high-risk obstetrics, genetics, and adult and/or congenital heart surgery, and in the setting of intervention and potential delivery, additional team members include neonatology, cardiac anesthesia, obstetric anesthesia, and perfusion specialists. Generally, this team is led by the cardiologist, with the patient having a critical role on the team and in decision-making.

Given that these cases, and particularly those that require intervention, are less frequent within an institution, maintaining a consistent team to manage these patients is advantageous. Although many women with repaired CHD can safely carry a pregnancy with low risk, careful planning and management by an experienced multidisciplinary team both preconception (if possible) and during pregnancy are essential. This article focuses on the surgical considerations for women with CHD contemplating pregnancy as well as during pregnancy, and also briefly reviews the physiologic changes of pregnancy, describes risk stratification in the context of pregnancy, and highlights key issues pertinent to surgeons.

CENTRAL MESSAGE

Although many women with repaired congenital heart disease can carry a pregnancy with low risk, management by an experienced pregnancy heart team is essential, and the surgeon has a critical role.

Hemodynamic Changes During Pregnancy

During pregnancy, blood volume increases by ~50%, comprising a 30% to 50% increase in plasma volume and a 20% to 30% increase in erythrocyte mass,2 resulting in a relative (dilutional) anemia. Other changes include decreases in systemic and pulmonary vascular resistance.3 The decrease in afterload during pregnancy has important sequelae that includes regurgitant lesions generally are better tolerated than obstructive lesions.4 There is also an increase in heart rate with an overall increase in cardiac output; this is a gradual process from the late first trimester until approximately 32 weeks of gestation, reaching a plateau that is 30% to 50% above preconception values. However, cardiac output increases still further during labor and delivery, to as much as 80% above the prepregnancy level.1 Inferior vena cava

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congestive heart failure from systemic ventricular systolic dysfunction, severe obstructive lesions, vascular Ehlers–Danlos syndrome, single-ventricle (Fontan) circulations with concurrent organ dysfunction, an aortic root diameter hypercoagulable state. Warfarin, the standard therapy for anticoagulation for mechanical valves, has teratogenic effects during the sixth to ninth weeks of pregnancy (usually dose-related), rarely has been associated with fetal intracranial hemorrhage, and there are also reports of high rates of fetal loss.

Unfractionated heparin does not cross the placenta and thus has not been associated with deleterious effects on fetal development. Valve guidelines support the use of warfarin during the second and third trimesters, with transitioning to intravenous unfractionated heparin near term and cessation of heparin immediately prior to delivery. In the first trimester, continuation of warfarin is reasonable if the dose to attain therapeutic anticoagulation is ≤5 mg/day, given the teratogenic effect of <3% at those doses. For patients receiving higher warfarin doses, LMWH or intravenous unfractionated heparin is recommended given the >8% risk of warfarin affecting fetal development.

Data on the risks and optimal management of anticoagulation for mechanical valves are limited. A meta-analysis comparing outcomes of warfarin throughout pregnancy, LMWH throughout pregnancy, LMWH in the first trimester and then warfarin, and heparin in the first trimester and then warfarin found the lowest composite maternal risk of death, thromboembolism, and valve failure of 5% with warfarin throughout pregnancy, compared to 16% for each of the other groups. The composite fetal risk of spontaneous abortion, death, and congenital defects was lowest with LMWH throughout pregnancy at 13%, with risk for the other regimens ranging from 23% to 39%; however, no significant difference in fetal risk was observed for those receiving ≤5 mg/day of warfarin and LMWH.

Data from the Registry of Pregnancy and Cardiac Disease (ROPAC) reported a 1.4% maternal mortality for those with mechanical valves, matching the rate in those with bioprosthetic valves. According to the ROPAC, valve thrombosis occurred in 5% (10 patients), 5 in the first trimester, all of whom had been switched to heparin. Hemorrhagic events occurred in 23%, compared with 5% in patients with bioprosthetic heart valves. Warfarin was associated with higher rates of miscarriage (29% vs 9% for heparin) and late fetal death (7% vs 1%). Overall, only 58% had a pregnancy free of serious adverse events, compared with 79% of those with a bioprosthetic valve.

Contraindications to Pregnancy

There are certain conditions for which pregnancy should be avoided for the safety of the mother, and if pregnancy occurs, termination should be considered. These conditions include severe pulmonary hypertension, congenital heart failure from systemic ventricular systolic dysfunction, severe obstructive lesions, vascular Ehlers–Danlos syndrome, single-ventricle (Fontan) circulations with concurrent organ dysfunction, an aortic root diameter...
of $\geq 45$ mm in the setting of Marfan syndrome, and an aortic dimension of 50 mm for bicuspid aortic valve aortopathy without high-risk features and 40 mm for Loeys–Dietz syndrome. Historically, maternal mortality has been reported to be 25% to 30% for those with severe pulmonary hypertension, while a more recent study reported a rate of 12%. In the current era, with careful management, maternal mortality in patients with severe aortic stenosis should be close to 0%, although hospitalization during pregnancy is common. Severe cyanosis is considered a relative contraindication to pregnancy, primarily because of the risks related to poor fetal outcome.

The risk profile of pregnancy for each woman with CHD should be individually assessed by experts in this area, because there will be many women who do not fit into the above categories but whose complex CHD may make pregnancy ill-advised. Ideally, these discussions are carried out prior to conception.

**Risk Stratification**

Surgeons should be aware of the algorithms that have been developed to help risk-stratify women with cardiac disease who become pregnant. Although these algorithms highlight major factors and provide a broad level of risk, each has limitations. Individual assessment by a specialty care team in experienced centers remains key in assessing the risk for a given patient. The most well-known risk stratification systems include CARPREG (Cardiac Disease in Pregnancy Study) I and II, ZAHARA (Zwangerschap bij Aangeboren HARtAfwijkingen; in English, Pregnancy in Women with Congenital Heart Disease), and maternal World Health Organization (mWHO). First published in 2001, the CARPREG score was derived from a study of 562 pregnant women with both acquired heart disease and CHD, and predicted cardiovascular events based on factors such as prior cardiac events, New York Heart Association (NYHA) functional status, left heart obstruction, and ventricular systolic dysfunction. A revised second version of this score (CARPREG II) includes more specific parameters with individual weights. The ZAHARA risk stratification was developed based on the CARPREG data specifically for women with CHD. The mWHO classification assigns risk for women with CHD and acquired cardiac disease and is the recommended tool of the American Heart Association and American College of Cardiology (Figure 3). A study of 213 pregnancies found that these risk stratification systems tended to overestimate actual risk, with mWHO the most accurate, although still not very discriminatory. Analyses from the ROPAC did find that mWHO was strongly associated with adverse events.

**Surgical Decisions for Women Prior to Childbearing**

In general, regurgitant lesions are better tolerated during pregnancy and might not necessitate intervention (Figure 4). Decision making is individualized and depends on the additional factors, such as symptomatology, results of exercise testing, ventricular dilation, systolic ventricular function, and other cardiac lesions. In addition, the feasibility of valve repair versus replacement also helps guide the decision of whether to repair prior to pregnancy or delay until after delivery.

In contrast to regurgitant lesions, obstructive lesions (of any valve or vessel) are more problematic and usually require intervention prior to pregnancy. Anatomic factors determine whether a percutaneous approach is feasible (either native valve or valve-in-valve) or stenting in the case of aortic coarctation or pulmonary artery/conduit abnormalities. In the event that surgical valve replacement is needed, prosthetic selection is individualized. In the aortic position, the Ross procedure would be preferred, with bioprosthetic replacement the second-best option. In the pulmonary position, bioprostheses have demonstrated good durability. The best option in the mitral position is also a stented bioprosthesis, but durability is more limited, and eventual reoperation is inevitable despite the use of percutaneous valve-in-valve therapy as a bridge. The tricuspid valve lends itself to repair in most circumstances, but if replacement is necessary, porcine bioprostheses function well and have good durability in this age group. Mechanical prostheses in any position are usually avoided if pregnancy is planned, given the anticoagulation management is challenging, with potential complications for both the mother and fetus.

**Considerations by Lesion**

**Left ventricular outflow tract obstruction.** Left ventricular outflow tract obstructive lesions require particular attention by surgeons and may require intervention prior to conception and rarely during pregnancy. Severe obstructive lesions are a contraindication for pregnancy.
Obstructions to left ventricular outflow include aortic valvular stenosis, coarctation, subaortic stenosis, supravalvular aortic stenosis, and hypertrophic obstructive cardiomyopathy. Severe aortic stenosis is usually secondary to bicuspid or unicuspid valves in women of childbearing age. Aortic stenosis requires comprehensive evaluation preconception even if the patient is asymptomatic. An exercise test is recommended to assess exercise tolerance, symptoms, blood pressure response, heart rate recovery, and electrocardiographic changes. Patients with decreased resting left ventricular systolic function or an abnormal exercise test should not pursue pregnancy until valve repair or replacement has been performed.\(^\text{14,26}\) These patients also require cross-sectional imaging to evaluate for bicuspid valve–related aortopathy or associated coarctation. If the ascending aorta is \(>50\) mm, aortic replacement should be considered prior to pregnancy.\(^\text{4}\) If aortic valve replacement is planned, then the threshold is \(45\) mm. Of note, those with aortic stenosis and a moderate degree of obstruction prior to pregnancy also can experience symptomatic deterioration during pregnancy.

For women with aortic stenosis who do become pregnant, the physiologic changes of pregnancy (eg, reduced systemic vascular resistance, higher cardiac output) increase the gradient and can result in angina, heart failure, syncope, and arrhythmias.\(^\text{27}\) Additionally, cardiac output reserve in these patients is relatively impaired, which limits perfusion to the uterus and can lead to intrauterine growth retardation, preterm birth, and low birth weight.\(^\text{4}\) Depending on the situation, medical treatment with a beta-blocker to decrease the heart rate, especially if the left ventricle is hyperdynamic or hypertrophied, can be helpful.\(^\text{4}\) If heart failure develops, cautious use of a diuretic can reduce circulatory overload. When feasible, percutaneous balloon valvuloplasty has been successfully performed with lead shielding the fetus and should be applied in the setting of a pliable, noncalcified aortic valve with minimal regurgitation and surgical backup.\(^\text{28}\) For those who remain symptomatic despite

**FIGURE 2.** Safety profiles of commonly used medications for the treatment of arrhythmias in pregnancy. (Diagram used with permission.\(^\text{6}\))
medical therapy, antepartum valve replacement should be considered. Depending on gestational age, this can be performed concomitantly with cesarean section delivery.29 There are also limited reports regarding the success of transcatheter aortic valve replacement during pregnancy.30 Subvalvar (such as subaortic membrane) and supravalvular aortic stenosis are handled similarly to aortic stenosis in terms of indications for surgery, with the exception that catheter-based options are not effective.

Coarctation carries a risk of impaired blood supply to the fetus with subsequent risk of compromised fetal growth and is also associated with risks to the mother. Many of these patients have already undergone interventions, and their prognosis depends on the degree of residual or recurrent coarctation. These patients commonly also have a bicuspid aortic valve and/or Shone syndrome and require evaluation for those associated lesions. There is also a slightly increased risk of aortic dissection secondary to both the increased cardiac output and the hormonal changes with pregnancy.4 Interventions during pregnancy are generally limited to cases of aortic dissection, uncontrollable hypertension, or heart failure. Although percutaneous intervention may be considered, there may be an increased risk of dissection during pregnancy.4,31

In general, pregnancy is well tolerated in patients with obstructive hypertrophic cardiomyopathy (HCM), with only 3 maternal deaths reported in the literature.7 There have not been any reported differences in outcomes for women with obstructive HCM and those with nonobstructive HCM. One-quarter of women report symptoms during pregnancy, with the majority having been symptomatic prior to pregnancy.7 Concomitant arrhythmias, such as atrial fibrillation, are managed as in other pregnant women. Beta-blockers are used as needed and should be continued when initiated prior to pregnancy.7,32 For patients who are significantly symptomatic, consideration should be given to interventions prior to pregnancy, such as septal myectomy for patients with symptomatic obstruction, advanced heart failure therapies for those in heart failure, and implantable cardiac defibrillator implantation for those with high-risk ventricular arrhythmias.7 Genetic counseling is indicated for all women with HCM. Counseling against pregnancy is generally reserved for only a small subset of patients, including those with an ejection fraction <30%, NYHA class III to IV with restrictive physiology, or severe obstruction not amenable to resection.32 A recent study of pregnancy in patients with HCM found that nearly one-quarter had adverse events, including atrial arrhythmias (12%) and heart failure (15%), with most occurring during the third trimester or postpartum.33 Impaired systemic ventricular function and systemic right ventricle. Preserved systolic function is important when considering pregnancy. For patients with a systemic ventricular ejection fraction <40%, the volume load of pregnancy is more likely to cause or exacerbate heart failure, and those with a systemic ventricular ejection fraction <30% are advised to avoid pregnancy. Additional considerations should be given for patients with a systemic

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**FIGURE 3.** Modified World Health Organization classification of maternal cardiovascular risk adapted from ESC guidelines on the management of cardiovascular diseases during pregnancy.22 \*WHO, World Health Organization; PS, pulmonary stenosis; PDA, patient ductus arteriosus; ASD, atrial septal defect; TOF, tetralogy of Fallot; BAV, bicuspid aortic valve; Ao, aorta; CHD, congenital heart disease; PH, pulmonary hypertension; LVEF, left ventricular ejection fraction; CV, cardiovascular.
(subaortic) morphologic right ventricle, who are at risk for progressive systolic dysfunction and atrioventricular valve (tricuspid) regurgitation, particularly with the increased volume load during pregnancy. Furthermore, changes in ventricular geometry can lead to significant regurgitation and clinical heart failure. Concomitant atrial arrhythmias or heart block predispose to overt heart failure. Although successful pregnancies have been reported, especially when ventricular function is preserved at conception, postpartum ventricular dilation does not return to preconception baseline in all patients. Longitudinal studies also have shown that ventricular dysfunction progresses in 25% of patients and systemic tricuspid regurgitation increases in 25%, with some patients demonstrating no recovery after delivery. Given these data, preconception counseling should include discussion of the potential long-term deleterious consequences of pregnancy.

Right ventricular outflow tract pathology. Right ventricular outflow tract pathology is most common following repair of tetralogy of Fallot or other conotruncal anomalies with right ventricular–pulmonary artery reconstruction and generally have a favorable prognosis with pregnancy, assuming the absence of symptoms and arrhythmias. In tetralogy of Fallot, the most common residual hemodynamic lesion is severe pulmonary regurgitation. This is generally well tolerated if right ventricular function is no more than mildly reduced and sinus rhythm is present. For patients with repaired pulmonary atresia, prognosis depends on right ventricular–pulmonary artery conduit function, right ventricular and pulmonary artery pressures, as well as right ventricular function. For all patients with CHD and right-sided pathology, evaluation for concomitant lesions, such as tricuspid regurgitation, residual/recurrent intracardiac shunts, and aortic valve pathology, also must be performed, because the presence of multiple “moderate” lesions may mandate intervention. For patients with repaired conotruncal abnormalities, genetic assessment for 22q11 deletion should be included for counseling regarding the risk of CHD recurrence.

Mitral valve disease. Congenital mitral valve disease includes atrioventricular septal defects (most previously repaired), myxomatous disease, and congenital (or rheumatic) mitral stenosis (also commonly previously repaired). The prognosis and management of these patients depend on the degree of regurgitation and/or stenosis, as well as on the size and function of the left ventricle and status of the pulmonary vasculature. Moderate to severe mitral regurgitation generally is well tolerated during pregnancy, presuming that systolic left ventricular function and pulmonary pressures are normal.

Tricuspid valve disease and Ebstein anomaly. Congenital tricuspid valve disease may include tricuspid valve dysplasia, Ebstein anomaly, and, less frequently, atrioventricular septal defects. These lesions are usually regurgitant and well tolerated during pregnancy if right ventricular systolic function is preserved. However, Ebstein anomaly is also accompanied by right ventricular and, less commonly, left ventricular myopathy that can be quite profound; generally, these women tolerate pregnancy, as long as right ventricular function is not severely reduced, regurgitation is moderate or less, and functional capacity is maintained. They are at risk for atrial arrhythmias, and one-half of those who go unrepaired have an atrial shunt that can result in hypoxia. Depending on severity, this hypoxia can impact...
fetal outcomes, and the shunt increases the risk of paradoxical embolism.4

Intracardiac Shunts

Most intracardiac shunts do not require intervention prior to pregnancy; however, there are specific circumstances when this may be necessary, such as large left-to-right shunts with evidence of volume overload. A history of endocarditis related to a restrictive ventricular septal defect may prompt closure prior to pregnancy. When there is right-to-left shunting across an atrial shunt, there is a risk of paradoxical embolism and hypoxemia, but a thorough evaluation is essential, because the more significant issue is why there is right-to-left shunting—for example, pulmonary hypertension, severe tricuspid regurgitation, etc. Percutaneous closure may or may not be indicated depending on the results of a comprehensive evaluation, which emphasizes the importance of an experienced multidisciplinary team.

Palliated single-ventricle physiology. There are limited data on the outcomes of pregnancy in patients with palliated single-ventricle physiology (ie, Fontan circulation). Even the Fontan patients with the most ideal hemodynamic circulation characteristics (ie, low pulmonary vascular resistance, left ventricle morphology with normal function, competent atrioventricular valve[s], and no conduit obstruction) are prone to heart failure and arrhythmias prior to the reproductive years. These patients all experience some degree of hypoxemia, whether from a fenestration or from collateralization. Other issues include potentially teratogenic medications that many patients require, which should be avoided during pregnancy if possible.

Successful pregnancy occurs infrequently and reflects a highly selected population among Fontan patients. In a multicenter study including 33 pregnancies, only 45% of the pregnancies resulted in a successful live birth.38 A more recent study from our institution assessed the outcomes of 70 pregnancies post-Fontan, there were 35 miscarriages, 29 live births, and 6 therapeutic abortions, and there were no maternal deaths.36 In another, more recent multicenter study of 37 Fontan patients who had 59 pregnancies, there were 16 miscarriages (27%) and 36 live births.40 Cardiac events occurred in 6 (10%) of the pregnancies, most commonly atrial arrhythmias (in 3 pregnancies). There were 3 thromboembolic events and 7 hemorrhagic events occurring antepartum and postpartum. Prematurity was common (69%), and anticoagulation was associated with adverse neonatal outcomes. At a median 24-month follow-up after pregnancy, there was no significant worsening of clinical status. Heart failure and arrhythmias are common, especially in those who had similar issues prior to pregnancy. Given the high incidence of heart failure, especially in the third trimester, preterm delivery is common, along with intrauterine growth retardation and premature rupture of membranes.4 Although pregnancy in certain settings has been accomplished successfully, it comes with significant risks, and pregnancy in this group in particular requires close observation and care by a highly experienced multidisciplinary team.

Pulmonary hypertension and Eisenmenger syndrome. Maternal and neonatal outcomes in Eisenmenger syndrome are generally poor, and pregnancy is not recommended.16,41 Not only is maternal mortality high, but also there are high rates of spontaneous abortion and fetal growth restriction. The physiology of Eisenmenger syndrome often cannot accommodate the hemodynamic challenges of pregnancy, particularly the volume load on the right ventricle coupled with inability to augment cardiac output. Furthermore, the drop in systemic vascular resistance increases right-to-left shunting, thereby increasing hypoxemia.4 The acute changes occurring during labor and delivery are particularly dangerous, and complications, including death, most commonly occur during labor and delivery or soon thereafter.4 Certain pulmonary vasodilators that are safe to use during pregnancy have demonstrated some improvement in outcomes,42 but this may be affected by publication bias. Support during labor and delivery focuses on decreasing pulmonary vascular resistance and supporting right ventricular function.

Heart Transplantation

Key considerations regarding pregnancy after heart transplantation include allograft function, management of immunosuppressants, genetic counseling (particularly for patients with inherited cardiomyopathy), and management of such comorbidities as hypertension and diabetes, which are increased in the setting of immunosuppressant use.43 Conception is not recommended within the first year after transplantation.43

Turner Syndrome and Pregnancy

Turner syndrome is associated with a high risk for primary ovarian insufficiency and infertility. Approximately 75% of affected individuals have no spontaneous pubertal development, 90% have primary amenorrhea, and a small percentage have residual ovarian follicles in early childhood.44 Only 2% to 5% are able to conceive spontaneously, although this subgroup usually are mosaic karyotypes.44 Fertility counseling in this population is challenging, because ovarian reserve may be present only relatively early in life. There are notable differences in ovarian function depending on the specific karyotype. Oocyte cryopreservation is possible and has been performed with ovarian stimulation in adults and more recently in the 13- to 15-year age bracket.44 For patients with Turner syndrome interested in childbearing, pregnancy with donor oocytes offers the...
The highest likelihood of success, although the rate of miscarriage is slightly higher compared with the general population. Of particular note, however, is the risk of aortic complications during pregnancy in patients with Turner syndrome who have cardiac disease. Overall, the maternal mortality is 1% to 2% (100 to 200 times) greater than that of non-Turner pregnancies. Preconception counseling suggests that those with an indexed aortic diameter of >2/cm² should not conceive, and those with a high index also appear to be at risk for dissection in the postpartum period. The cardiac risk is in addition to other concurrent risks present in these patients, including renal and thyroid abnormalities, among others.

Peripartum Management and Postpartum Surgery

Peripartum management of patients with CHD is not standardized and depends on institutional and provider preferences. A meta-analysis by Asfour and colleagues showed that vaginal delivery is relatively safe in patients with CHD of all degrees of severity and that a higher rate of cesarean delivery did not improve outcomes but was associated with an increased risk of adverse maternal events. Similarly, anesthetic management of labor and delivery is not standardized and based on maternal and fetal condition; however, neuroaxial anesthesia is preferred, and general anesthesia is usually reserved for emergent situations. Pregnancy, labor, and delivery in women with high-risk of peripartum complications should be managed in tertiary referral centers. Ideally, planning for labor and delivery should be done far in advance and include input from cardiovascular, obstetric, anesthesia, and, as-needed, neonatology experts.

For those women who successfully complete the pregnancy and delivery with CHD and without decompensation, the timing for cardiac surgery will depend on the lesion and clinical status and should be a shared decision with the patient. Patients with regurgitant lesions, especially with preserved ventricular function, might not require surgery for a number of years, given that they tolerated the volume load and other hemodynamic changes with pregnancy that then resolved. For obstructive lesions, postpartum intervention may be required more frequently; this may be as soon as the day of delivery, within weeks of delivery, or in the upcoming months. The importance of the mother bonding with her newborn baby, and how that would be impacted with surgery, should be taken into consideration.

Emergent and Salvage Situations

Extracorporeal life support can be instrumental in saving both the mother and baby’s life in certain settings. Although the use of veno-veno extracorporeal membrane oxygenation (ECMO) has been reported in various case series and by the Extracorporeal Life Support Organization (ELSO) for supporting pregnant women with respiratory failure, veno-arterial ECMO has been reported less frequently. A meta-analysis of 45 patients supported with ECMO at a mean gestational age of 26.5 weeks and for a median duration of 12 days found a maternal survival rate of 78% and a fetal survival rate of 65%. Forty-one of those patients were supported with veno-veno ECMO, the majority for H1N1 influenza acute respiratory distress syndrome, and 4 patients were supported with veno-arterial ECMO, 1 of whom died. In the aforementioned review, bleeding was a common complication. The effect of ECMO on uterine circulation is unknown, and patients should remain in the left lateral position, especially after 20 weeks, to avoid compression of the inferior vena cava and aorta. An important factor regarding the use of ECMO is to clarify its role either for reversible causes or as bridge to a more definitive therapy, and not typically as a destination strategy. Therefore, in pregnant women with cardiovascular collapse, the question remains the endpoint. For some patients, this may be considered as a bridge to the ability to deliver a viable fetus followed by transplantation.

In a recent multi-institutional report of 49 pregnant women with pulmonary hypertension, 6 women were placed on ECMO, including 3 placed on ECMO postpartum at the time of cardiac arrest, and all died: only 1 woman was successfully weaned from ECMO and discharged home, but she died at 89 days postpartum.

At times, the pregnancy heart team may conclude that cardiac surgery during pregnancy, either while maintaining the pregnancy or immediately after delivery under the same anesthetic, is warranted. These cases require careful coordination with multiple teams preoperatively, including timing of the surgery relative to gestational age, as well as intraoperatively in terms of the conduct of the procedures. The specifics of managing these cases, from surgical considerations to monitoring, have been summarized recently.

Mechanical valve thrombosis is a rare and potentially life-threatening complication in patients with a prothrombotic state such as pregnancy, and its management during pregnancy warrants specific discussion. A multidisciplinary pregnancy heart valve team assessment is needed to guide the choice between fibrinolysis and surgery. Urgent management of left-sided mechanical (or bioprosthetic) valve thrombosis is needed for those with high-risk features, such as symptomatic NYHA functional class III/IV or a mobile thrombus >0.3 cm in diameter or any thrombus with area ≥1.0 cm². When the patient is critically ill and is a surgical candidate, surgery is preferred.

In selected pregnant hemodynamically stable patients with obstructive left-sided mechanical valve thrombosis, the American College of Cardiology/American Heart Association valve guidelines indicate that it is reasonable to treat with slow-infusion, low-dose fibrinolytic therapy rather than valve surgery. Limited data suggest that fetal
and maternal risk may be lower with this strategy as opposed to surgery, although direct comparative data are lacking. There is no documented transplacental passage of tissue plasminogen activator, but there is a risk of placental hemorrhage.

**Genetic Counseling and Recurrence Risk**

As noted above, all women with CHD considering pregnancy should seek genetic counseling. The specific testing recommended will depend on the patient and the CHD. The risk of CHD recurrence depends on the disorder and whether a genetic etiology is found. For autosomal recessive disorders, such as Ellis–van Creveld syndrome, the risk would be 25%. When a de novo autosomal dominant variant is found, there is a possibility of gonadal mosaicism, with recurrence risk of ~1%. For sporadic CHD, the risk of recurrence if 1% to 5%. A number of technologies are used in prenatal diagnostic testing including karyotyping, fluorescence in situ hybridization, chromosome microarray analysis (CMA), and DNA testing. The DNA for these tests can be extracted from either a chorionic villus sample or an amniocentesis sample. Karyotyping, fluorescence in situ hybridization, and CMA can detect aneuploids, and CMA also is able to detect duplications and deletions of genetic material, as in 22q11.2 syndrome. For families with a known pathogenic variant, DNA can be analyzed to identify these specific variants. The risk of fetal loss after chorionic villus sampling or an amniocentesis is approximately 1%. Recently, cell-free fetal whole-genome sequencing has been developed as a noninvasive screen for chromosomal aneuploidy. Although this is not currently used as a noninvasive diagnostic for single-gene conditions, it is sure to expand as the technology improves.

**CONCLUSIONS**

Although many women with CHD can safely carry a pregnancy with low risk, careful planning and management by an experienced pregnancy heart team is essential. Indicated surgical interventions are optimally completed prior to conception. Preconception counseling should focus on anticipated clinical consequences of physiologic adaptations during pregnancy, the possibility of antepartum maternal cardiac events, and the potential for preterm delivery with attendant neonatal sequelae.

**Conflict of Interest Statement**

The authors reported no conflicts of interest.

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**Key Words:** adult congenital heart disease, fetal, heart team, mechanical valve, pregnancy