Pregnancy and Delivery in Women With Congenital Heart Disease

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Because of the growing population of patients with congenital heart disease (CHD), most maternal cardiac disease is now congenital in origin. For women with complex CHD, pregnancy poses an increased risk for both the mother, with complications of arrhythmias and heart failure being the most common, and the baby, with a higher chance of miscarriage, intrauterine growth retardation, and the need for early delivery. Pre-pregnancy counseling must be performed by cardiologists who have expertise in both CHD and pregnancy, with a detailed clinical assessment of the patient and the current hemodynamic situation, including echocardiography and an exercise test. In each case the approach must be individualized with consideration of the risks in each case. In some cases, such as Eisenmenger syndrome, pregnancy is contraindicated. Optimum outcomes in these complex patients are achieved when a multidisciplinary approach is used, involving maternal-fetal medicine specialists, cardiologists with expertise in CHD and obstetric anesthesia. (Circ J 2015; 79: 1416–1421)

Key Words: Congenital heart disease; Eisenmenger syndrome; Genetic counseling; Preconception counseling; Pregnancy

Approximately 2% of pregnancies involve cardiac disease, and in the current era, most maternal cardiac disease is congenital in origin, in large part because of the success of cardiac surgery over the past 50 years, leading to increased numbers of survivors with congenital heart disease (CHD); in the United States alone the population is estimated to exceed 1 million. For such women, the desire to bear children is an important component of their care, so pregnancy counseling and management are important issues for cardiac health providers. For many women with CHD (either unoperated or repaired), pregnancy can be accomplished with minimal increase in risks for mother and baby, but each case must be individualized. This review will focus on the congenital cardiac lesions that pose the most risk for mother and baby. In each case it is very important that preconception counseling is reviewed carefully in these patients before a pregnancy is undertaken.

Preconception Counseling

Issues to be discussed at the time of preconception counseling are listed in Table 1. For most women with CHD, pregnancy can be accomplished successfully, but it may be associated with an increased risk for both mother and baby. The general hemodynamic changes of pregnancy need to be reviewed: pregnancy is associated with a 50% increase in plasma volume load and a lesser rise in red cell mass, causing the physiological anemia of pregnancy. The afterload tends to fall, in part because of the hormonal changes of pregnancy, and so in general, regurgitant lesions in the mother are better tolerated than stenotic ones. Because of the volume load that pregnancy imposes, it is possible that in some circumstances, there may be a deterioration in maternal cardiac hemodynamics, which might accelerate the mother’s deterioration in the long term. With some cardiac lesions, ventricular function and/or valve regurgitation might be expected to progress with time and so the timing of pregnancy is another important consideration. Maternal risk may increase with delayed pregnancy and this needs to be reviewed. A sensitive discussion should also be had about the long-term outlook for the mother, and in some cases, the possibility of limited longevity. The risks of miscarriage and poor fetal growth also need to be reviewed with the mother and in circumstances where pregnancy is not feasible, alternative options such as adoption and surrogacy should also be reviewed. At this time, if pregnancy is not advisable, appropriate advice regarding contraception is important.

Preconception counseling should be accomplished in a regional center where there is expertise in both adult CHD and high-risk pregnancy care. The visit would generally involve a detailed history, physical examination, electrocardiogram, and chest X-ray, as well as an echocardiogram. Transthoracic echocardiography facilitates assessment of ventricular function, valvular function and the status of the aorta and pulmonary artery and the degree of shunting. It may be able to reliably assess the pulmonary artery pressure, but if there is any doubt about the possibility of important pulmonary hypertension, cardiac catheterization should be undertaken before pregnancy proceeds. Exercise testing is particularly useful at this visit.
because many patients with CHD have a relatively sedentary existence and may perceive their exercise capacity to be normal. An exercise test may be helpful therefore in reliably assessing the degree of functional aerobic capacity.

A preconception visit with the high-risk obstetric team may also be helpful for those women who are at high risk. Others issues that need to be addressed are outlined in Table 2. Any drugs that are potentially teratogenic will need to be discontinued prior to pregnancy. Particularly common are the use of angiotensin-converting enzyme (ACE) inhibitors and angiotensin receptor blockers (ARBs), which must be avoided during pregnancy. If it is determined that pregnancy is feasible and advisable, a plan needs to be established for monitoring antepartum care and where obstetric care and delivery should be undertaken. This might be at a tertiary/quaternary care center or at a step-down unit). Finally, appropriate contraceptive advice should be given when necessary.

**Genetic Counseling**

Genetic counseling, often with the help of a properly trained genetic counselor, should discuss the assessment of genetic risk to the patient and her partner with a detailed family history of CHD and the possibility of any genetic or chromosomal abnormality. A discussion of genetic screening tests should also be undertaken. For most women with CHD arising de novo, the possibility of having a baby with CHD is approximately 3–4%. If >1 sibling is affected by CHD, the recurrence risk may be as high as 10%. The risk of recurrence is higher with some lesions, such as bicuspid aortic valve and left-sided outflow tract obstructive lesions. Some lesions that are associated with autosomal dominant lesions, such as cono-truncal abnormalities (truncus arteriosus, pulmonary atresia, and tetralogy of Fallot), are associated with 22q11.2 deletion and this testing should be offered to anyone suspected of carrying this chromosomal abnormality. Other testing may be indicated for genetic abnormalities such as Marfan or Noonan syndrome should be considered. Fetal cardiac echocardiography should be offered at approximately 20 weeks’ gestation.

**Predictors of Maternal Cardiovascular Events**

Several predictors of maternal cardiovascular events have been published in an effort to try and ascribe a more accurate maternal risk at the time of pre-pregnancy counseling. The best known of these are the CARPREG study and the Zahara study. Generally, in these 2 series the risks of cardiac complications occurring in the mother averaged between 7% and 13%, the most prevalent cardiac complications usually being arrhythmias and heart failure. Supraventricular arrhythmias are particularly common in this population and if they have occurred pre-pregnancy, are likely to recur during pregnancy. The challenge with these predictors, however, is that they are highly population dependent; for example, the CARPREG study included patients with congenital and acquired heart disease and the Zahara study included patients with CHD only. Important risk factors such as severe pulmonary hypertension and severely dilated aortas are not identified in either study and are therefore, under-represented. Thus these predictors only provide a “snapshot” of what the anticipated cardiovascular risk might be. A recent study identified in a series of over 200 pregnancies that these risk predictors tend to overestimate the degree of risk and perhaps the most reliable is the modified WHO classification, which provides the most favorable risk estimation model.

This review will focus on the congenital cardiac lesions that pose the most challenge for pregnancy, labor and delivery (ie, those that fall into WHO categories III and IV; Table 3). Category III are lesions associated with a significant increase in maternal mortality or morbidity and requiring expert counseling. If pregnancy is undertaken these patients require expert care. Class IV includes patients with cardiac lesions that have extremely high maternal mortality or severe morbidity and for whom pregnancy is contraindicated. If pregnancy occurs a discussion of termination should be undertaken.

**Labor and Delivery**

With each uterine contraction, approximately 500 ml of blood is released into the maternal circulation, causing abrupt changes in hemodynamics. The cardiac output increases approximately 80% above baseline at the time of a normal vaginal delivery. Blood loss of approximately 400 ml occurs at the time of a normal vaginal delivery, but is approximately 800–1,000 ml with a cesarean section. Delivery is followed by an increase in maternal volume secondary to an increase in venous return from the uterus and inferior vena caval decompression. For women with tenuous hemodynamics, labor and delivery must be accomplished in a tertiary care setting and monitored carefully. For most women, vaginal delivery is preferable and safer.
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Saturation and pressure in preference to automated blood pressure is useful, and in some circumstances (cyanotic heart disease) dynamics by monitoring fluid shifts. Continuous pulse oximetry facilitates the stabilization of hemodynamic perturbations (e.g., cyanotic heart disease) by detecting cyanosis, and the hemodynamic perturbations during labor and delivery (including blood loss) may precipitate cardiovascular collapse. Death frequently occurs during labor and delivery, or within the days and weeks following delivery, from refractory decline in peripheral resistance and progressive hypoxia, or in situ pulmonary thrombus. Although the addition of pulmonary vasodilator therapies may improve the outcome for mothers with Eisenmenger syndrome, it should be recognized that the outcome for many of these women is still often fatal, and publication bias means that only the best outcomes are usually reported. Nonetheless, successful outcomes have been reported with both vaginal delivery and delivery via cesarean section. Intravenous prostacyclin (epoprostenol) may be helpful; sildenafil and tadalafil have also been used, as well as nebulized and intravenous iloprost. Inhaled nitric oxide may also be helpful around the time of delivery. All these therapies are aimed at reducing the pulmonary resistance and thereby stabilizing the right ventricular function. If a woman with Eisenmenger syndrome becomes pregnant, therapeutic termination is the safer option, with the caveat that, this too, is not without risk and must be managed carefully in a center where there is expertise in treating both CHD and pulmonary hypertension. If the mother elects to continue the pregnancy, she needs to be followed frequently by a multidisciplinary care team with regular clinical evaluation and monitoring in a tertiary care center. Activity will need to be minimized and early hospital admission for bed rest and monitoring should be individualized depending on maternal cardiac status. Elevation of the jugular venous pressure and signs of right heart failure need to be treated judiciously with diuretics. The use of anticoagulation is controversial because of concerns about bleeding, but low-dose heparin during bed rest is probably reasonable.

The mode and timing of delivery needs to be individualized and depends on maternal cardiac status; frequently an early delivery is necessary because of maternal cardiovascular deterioration and thus a cesarean section may be necessary. It can be accomplished with general anesthesia (often combined obstetric and cardiac anesthesia) and a multidisciplinary team should be used to optimize the outcome. If regional anesthesia is used, such as epidural anesthesia, small incremental doses must be given to avoid hypotension. Intra-arterial pressure monitoring, together with central venous pressure monitoring, will facilitate prompt detection of hemodynamic changes; pulmonary artery pressure catheters are best avoided because they may precipitate ventricular arrhythmias. Recovery should be undertaken in an intensive care unit and maternal monitoring is necessary for at least 1 week after delivery. Meticulous attention to the leg veins is necessary because deep venous thrombosis may cause a paradoxical embolus.

### Table 3. Congenital Cardiac Lesions for Which Pregnancy Risk Is WHO Class III or IV

<table>
<thead>
<tr>
<th>Class III</th>
<th>Class IV</th>
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<tr>
<td>Cyanotic heart disease</td>
<td>Pulmonary hypertension</td>
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<tr>
<td>Mechanical valve</td>
<td>Severe systemic ventricular dysfunction (ejection fraction &lt;30%, NYHA III–IV)</td>
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<tr>
<td>Systemic right ventricle</td>
<td>Severe symptomatic aortic stenosis</td>
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<tr>
<td>Fontan</td>
<td>Native severe coarctation</td>
</tr>
<tr>
<td>Marfan syndrome: aorta 40–45 mm</td>
<td>Marfan syndrome: aorta &gt;45 mm</td>
</tr>
<tr>
<td>Bicuspid aortic valve: aorta 45–50 mm</td>
<td>Bicuspid aortic valve: aorta &gt;50 mm</td>
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Modified from Thorne S, et al.4

### Table 4. Indications for Cesarean Section Delivery

- Patient on coumadin (to avoid risk of fetal intracranial hemorrhage)
- Severe obstructive lesions (e.g., aortic stenosis)
- Aortopathy with unstable aorta (Marfan syndrome, dissection, bicuspid aortic valve, coarctation)
- Severe pulmonary hypertension?

unless there is an obstetric indication. In some circumstances a cesarean section delivery may be preferable for obstetric reasons, but some cardiac situations may be safer with a cesarean section delivery to avoid any pushing and to minimize the hemodynamic perturbations (Table 4).

In order to minimize maternal risk, the timing and delivery in a tertiary care center are crucial, with a multidisciplinary team involving experts in cardiology, maternal-fetal medicine, anesthesia and neonatology. If the cervix is favorable and vaginal delivery is planned, labor may be induced with good pain control, usually with epidural anesthesia. Labor can be conducted with the mother in the left lateral position to avoid inferior vena caval compression and maintain venous return. Bearing down (Valsalva maneuver) is often best avoided in patients with complex CHD, but the approach must be individualized. The second stage can be assisted with forceps or vacuum extraction if necessary. Prolonged labor should be avoided.

In addition to fetal monitoring, maternal ECG monitoring should be performed to detect any arrhythmia during labor and the puerperium. Intravenous lines, and if necessary central venous pressure catheters, facilitate the stabilization of hemodynamics by monitoring fluid shifts. Continuous pulse oximetry is useful, and in some circumstances (cyanotic heart disease) intra-arterial monitors may detect early changes in oxygen saturation and pressure in preference to automated blood pressure monitoring. The duration and location of postpartum monitoring must be individualized, but in general for those women in WHO Class III or IV, should be continued at least 24h after delivery in an intensive care unit.

### Pulmonary Hypertension and Eisenmenger Syndrome

The outcomes of pregnancy for women with Eisenmenger syndrome are poor for both the mother and fetus. Maternal mortality is approximately 30% and women should be strongly counseled against pregnancy at the time of pre-pregnancy counseling. Fetal loss is also high, with high rates of spontaneous abortion and intrauterine growth retardation. The volume load of pregnancy poses an added burden for the already compromised right ventricle. The fall in systemic vascular resistance increases right to left shunting and exacerbates cyanosis, and the hemodynamic perturbations during labor and delivery (including blood loss) may precipitate cardiovascular collapse. Death frequently occurs during labor and delivery, or within the days and weeks following delivery, from refractory decline in peripheral resistance and progressive hypoxia, or in situ pulmonary thrombus. Although the addition of pulmonary vasodilator therapies have improved the outcome for mothers with Eisenmenger syndrome, it should be recognized that the outcome for many of these women is still often fatal, and publication bias means that only the best outcomes are usually reported. Nonetheless, successful outcomes have been reported with both vaginal delivery and delivery via cesarean section. Intravenous prostacyclin (epoprostenol) may be helpful; sildenafil and tadalafil have also been used, as well as nebulized and intravenous iloprost. Inhaled nitric oxide may also be helpful around the time of delivery. All these therapies are aimed at reducing the pulmonary resistance and thereby stabilizing the right ventricular function.

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Cyanotic Heart Disease

All cyanotic heart disease poses difficulty for both mother and fetus. Declining afterload worsens maternal cyanosis, the anemia of pregnancy exaggerates maternal hypoxia and the prothrombotic state of pregnancy increases the risk of thrombosis, not just in the pelvic and leg veins but also in the pulmonary vasculature. In addition, maternal hypoxia poses a handicap to fetal growth, so there is increased risk of spontaneous abortion and intrauterine growth retardation. In the largest series of pregnancy and cyanotic heart disease (Eisenmenger patients excluded), Presbitero et al\textsuperscript{16} noted that fetal outcome was related to the degree of maternal cyanosis. In mothers whose oxygen saturation was $\geq 90\%$, 92\% of babies were live born. In contrast, when the mother’s oxygen saturation was $\leq 85\%$, only 12\% of babies were live born. Maternal complications also occurred, including thrombosis of a pulmonary artery and a cerebral infarct and 3 cases of endocarditis; none of the patients received endocarditis prophylaxis.

Aortic Stenosis (AS)

Severe AS (aortic velocity on echo/Doppler $>4\text{m/s}$, mean gradient 40mmHg) in women of childbearing years is almost always (in Western societies) related to the bicuspid aortic valve. A pre-pregnancy evaluation is necessary and even if the patient is asymptomatic, exercise testing is recommended to confirm asymptomatic status and evaluate exercise tolerance, blood pressure and ECG response and to determine if intervention is necessary before pregnancy is undertaken.\textsuperscript{17} Any patient with symptoms, impaired left ventricular function or a pathological exercise test, should be strongly counseled against pregnancy\textsuperscript{17, 18} and valvuloplasty or valve replacement should be undertaken before pregnancy. Discussion about the type of valve replacement should be undertaken with a cardiologist with expertise in treating patients with valvular heart disease and pregnancy regarding the risks and benefits of all options and choice of valve (tissue vs. mechanical).\textsuperscript{19} Because the bicuspid aortic valve is associated with an aortopathy, detailed imaging of the ascending aorta by MRI or CT should also be performed pre-pregnancy. If the ascending aortic dimension exceeds 50mm, pre-pregnancy surgery should be considered.

If a patient with severe AS becomes pregnant, the fall in afterload exaggerates the aortic gradient and maternal complications include heart failure, arrhythmias and angina.\textsuperscript{19} The fixed cardiac output also impairs blood flow to the uterus and fetus, potentially leading to intrauterine growth retardation, preterm birth and low birth weight, as well as fetal death. Multidisciplinary follow-up is necessary and as cardiac demands increase, bed rest may need to be implemented. Clinical evaluation and echocardiography facilitate hemodynamic assessment of the valvular gradient and ventricular function. Beta-blockade may be helpful if there is hyperdynamic ventricular function and tachycardia. Diuretic therapy may be used cautiously in the event of heart failure. Early delivery may be necessary if the fetus is viable. Percutaneous aortic valvuloplasty has been accomplished successfully during pregnancy with lead shielding of the fetus, but should only be performed in experienced centers and can only be performed when the valve is pliable and non-calcified and there is minimal aortic regurgitation.\textsuperscript{20} For symptomatic patients refractory to medical therapy, aortic valve replacement can be accomplished during pregnancy, and if the fetus is viable it can be timed concomitantly with cesarean section delivery.\textsuperscript{21}

Coarctation of the Aorta

For women with coarctation of the aorta, there are risks for both mother and baby. Because of impaired flow to the uterus, fetal growth may be impaired. For the mother, there is concern because of hypertension with its attendant complications. In addition, because of the aortopathy associated with coarctation, there is an increased risk of aortic dissection not only because of the increased cardiac output, but because hormonal changes of pregnancy “soften” the aorta, making it more vulnerable to dilatation and dissection. Beta-blockers are the most frequently used antihypertensive agent in this context, but blood pressure control needs to be meticulous because aggressive control may impair blood flow to the fetus. Rarely, intervention during pregnancy to relieve coarctation is necessary, and balloon and stent placement have been accomplished with the concern for tearing the aortic wall and dissection because the aorta is more fragile during this time. Most women will be able to undergo successful delivery, albeit with hypertensive complications, but outcomes are clearly better after successful repair.\textsuperscript{22, 23}

Impaired Ventricular Function and the Systemic Right Ventricle

Women with and ejection fraction $<40\%$ are likely to encounter problems in pregnancy as the volume load progresses and they may develop heart failure. For those with an ejection fraction $<30\%$, pregnancy is absolutely contraindicated. In patients with transposition complexes, the systemic ventricle is likely to be the right ventricle. In patients with d-transposition, the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle. The earliest surgical repair (called either a Mustard or Senning operation) involved an atrial repair that redirected the blue deoxygenated blood to the left ventricle and red oxygenated blood from the pulmonary veins to the right ventricle and then to the aorta. Thus the circulation moves in the correct direction but the right ventricle functions as the systemic ventricle. As such, it is vulnerable to failure, together with progressive tricuspid regurgitation. Patients with l-transposition have both atrioventricular discordance as well as ventriculo-arterial discordance, meaning that the right atrium connects to the left ventricle, which gives rise to the pulmonary artery, and the left atrium enters the right ventricle, which gives rise to the aorta. Thus this “double disconnect” means that the circulation moves in the correct direction but the right ventricle serves as the systemic ventricle and so this lesion is also called “congenitally corrected transposition”. These patients may survive to adulthood without surgery, although some may have had surgery in childhood. In both of these circumstances, the volume load of pregnancy may precipitate heart failure if the ventricular function is depressed at the outset. If the maternal hemodynamics are borderline, minor disturbances in ventricular geometry and annular dilatation may cause more tricuspid regurgitation, which in turn begets more tricuspid regurgitation, and heart failure may ensue, particularly if the pregnancy is complicated by an atrial arrhythmia.

Successful pregnancies have been reported in patients with d- and l-transpositions,\textsuperscript{24, 25} provided the patients are carefully selected, and their systemic ventricular function is reasonably well preserved before pregnancy is undertaken. One important issue in these patients, however, is that the volume load of pregnancy may cause more systemic ventricular dilatation and cause more tricuspid regurgitation, which may not return to baseline after delivery. Guedes et al\textsuperscript{26} noted in a small series
of patients with d-transposition after the Mustard operation that right ventricular dilatation progressed in approximately one-third of patients and did not recover. Right ventricular dysfunction also progressed in approximately one-quarter of patients, with no recovery in the majority, and tricuspid regurgitation also increased in approximately half of the patients, some of whom did not subsequently return to baseline after the pregnancy, including a permanent deterioration in functional capacity. Thus for some of these patients, the preconception counseling discussion should include not only a discussion about the advisability of a pregnancy, but the possibility that pregnancy might cause a deterioration in maternal hemodynamic status that might affect her long-term outcome.

### Single-Ventricle Physiology and the Fontan Operation

The data on pregnancy in women with a single ventricle and Fontan operation are scarce but clearly such patients need to be selected carefully because they are vulnerable to heart failure and arrhythmias even before pregnancy, and some remain cyanotic and are on anticoagulants. Many are on anti-arrhythmic agents and either ACE inhibitors or ARBs; the latter are contraindicated in pregnancy. In the first multicenter study, the risk of spontaneous abortion was high, with only 45% of live births from a small series of 33 pregnancies. Pregnancy complications, particularly heart failure and arrhythmias, are common, particularly in those women who had arrhythmias before pregnancy. Patients must be managed by an experienced multidisciplinary team, and maternal decompensation with heart failure is frequent in the 3rd trimester, necessitating early delivery (often by cesarean section). Concerns for the fetus include intrauterine growth retardation and premature rupture of membranes with high prematurity rates.

### Mechanical Valve Prostheses

For the woman with a mechanical valve prosthesis, there is no perfect anticoagulant strategy during pregnancy and each anticoagulant is associated with increased risk for both the mother and baby. All anticoagulants are associated with an increased risk of maternal, placental and fetal hemorrhage. Several issues must be considered, including the fact that pregnancy is the most prothrombotic state, and cannot be managed in the same way as patients with mechanical valves are managed through other non-cardiac surgery. The tilting disc mitral valve is the most vulnerable to thrombus and a careful assessment of valve function must be made by clinical and echocardiographic evaluation before pregnancy is undertaken. Counseling of patients must be performed by a cardiologist with expertise in the management of pregnancy and valvular heart disease.

Oral vitamin K antagonists (eg, warfarin) are the most effective in terms of preventing maternal valve thrombosis, but because they cross the placenta, they may be associated with fetal abnormalities, including embryopathy, central nervous system deformities, bony abnormalities (stippled epiphyses) and blindness when used in the 1st trimester, as well as increased risk fetal loss, hemorrhage and still birth when used in the 2nd and 3rd trimesters. Heparin, either unfractionated or low-molecular weight (LMWH), does not cross the placenta, but is not such an effective anticoagulant as warfarin and its use is associated with an increased risk of maternal valve thrombosis and embolus. The relative risks and benefits of warfarin vs. heparin are shown in Table 5. The risk of fetal embryopathy appears to be dose-related, with a lower incidence of embryopathy when the maternal dose of warfarin is ≤5 mg/day, but the INR must be therapeutic at all times between 2.5–3.5.

Therapeutic approaches have included the use of unfractionated heparin in the 1st trimester (to avoid fetal exposure to warfarin), with a change to warfarin in the 2nd and 3rd trimesters and a return to heparin at approximately 35 weeks in anticipation of delivery. Intravenous unfractionated heparin can be used at that time to avoid the risks of fetal intracranial hemorrhage during a vaginal delivery, and anticoagulation interrupted for as short a time as possible in an effort to avoid maternal valve thrombosis. Monitoring of the activated partial thromboplastin time (aPTT), however, needs to be frequent and meticulous because there is considerable variability in dosing, mandating hospital admission if this is to be accomplished. The aPTT level must be maintained at more than twice control levels at all times. Historical data, however, have demonstrated that this approach is associated with an increased risk of valve thrombosis (=double vs. continued use of warfarin).

LMWH is an attractive alternative because of its greater ease of use, good bioavailability and administration by subcutaneous injection. Targeting the inhibition of factors II and Xa, its use was initially problematic when used by weight-based dosing and fatal valve thromboses were reported. Subsequent studies, as well as the ESC and ACC/AHA guidelines have made the strong recommendation that, if LMWH is used, it should be given at least twice daily and anti-Xa monitoring must be performed with a target anti-Xa level of 0.8–1.2U/ml measured 4–6h after injection. The optimum frequency of anti-Xa measurements is not entirely clear, (probably at least every 2 weeks), nor whether trough levels should be performed. Even with this approach and meticulous monitoring of the anti-Xa levels, valve thromboses have still been reported, with an incidence of 5–17%. In some cases, patient compliance may have been an issue, but in the absence of randomized control trials and with only small cases series on which to base recommendations, appropriate therapeutic strategies remain somewhat controversial.

The current ESC and ACC/AHA guidelines recommend the use of warfarin if the mother has a therapeutic INR on a dose of warfarin ≤5 mg/day and this should be continued until prior to delivery. Women who require >5 mg/day and those

<table>
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<tr>
<th>Table 5. Anticoagulants and Pregnancy</th>
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<tr>
<td>Warfarin</td>
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<tr>
<td>Better anticoagulant</td>
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<tr>
<td>Poor anticoagulant: doubles risk of valve thrombosis</td>
</tr>
<tr>
<td>Crosses placenta: fetal embryopathy</td>
</tr>
<tr>
<td>Does not cross placenta</td>
</tr>
<tr>
<td>Fetal hemorrhage</td>
</tr>
<tr>
<td>Lab control difficult; UFH: aPTT 2.5–3.5xcontrol</td>
</tr>
<tr>
<td>CNS abnormalities</td>
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<tr>
<td>LMWH: anti-Xa 0.8–1.2U/ml 4–6h post dose</td>
</tr>
<tr>
<td>Retro-placental bleeding</td>
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CNS, central nervous system abnormalities; LMWH, low-molecular weight heparin; UFH, unfractionated heparin.
unwilling to use warfarin in the 1st trimester should be treated with heparin in the manner described above. During the 2nd trimester, all women should be transitioned to warfarin and changed to unfractionated heparin prior to delivery. All women should receive low-dose aspirin (75–100 mg once daily).

Ideally, delivery is induced with interruption of heparin for as short a time as feasible – often within 24 h of delivery. Cesarean section delivery is necessary if the mother has not been off warfarin for 1 week. The risks of postpartum hemorrhage are increased in women receiving anticoagulation and postpartum monitoring must be continued for several days.

Conclusions

The population of patients with CHD continues to grow. Women with complex CHD are at higher risk of maternal complications during pregnancy, and there is a higher risk of fetal complications. Patients should receive pre-pregnancy counseling by a cardiologist with expertise in the management of such cases, and when pregnancy is deemed advisable, a multidisciplinary team approach improves the outcomes for both mother and baby.

References


Pregnancy and CHD