Pregnancy in Women with Heart Disease in Vietnam:

With the progression of corrective surgery technologies and procedures, 85% of babies born with congenital heart disease will survive until adult hood. This means that more and more women with congenital and acquired heart disease will reach child-bearing age in Vietnam. Pregnancy makes a significant demand on the cardiovascular and respiratory system. Healthy women and those with mild cardiac abnormalities can compensate for these increased demands, and have an uncomplicated pregnancy. In some conditions, however, the body is unable to compensate for the increased demands and this can result in very serious complications for the mother and the baby.

In general most women with congenital heart defects, especially those who have had corrective surgery, can safely become pregnant. However, it depends on the type of heart defect, the severity of symptoms and the presence of pulmonary hypertension. It is also important to remember that there may be a percentage of women who will not have had corrective surgery for congenital heart defects such as VSD's or ASD's, which could have resulted in more serious complications and progression of congenital heart disease. Congenital heart disease is the most common birth defect and just less than 1% of newborns have CHD. There are several risks involved with pregnancy in women with CHD which can cause adverse maternal and fetal outcomes. Acquired heart disease is seen very rarely amongst women of child-bearing age in many western countries. However, Vietnam has a high incidence of Rheumatic Fever and it is estimated that 4-10/1000 people in Vietnam have acquired Rheumatic Heart Disease.

It is recognized that discussions on future pregnancies and prevention should begin in adolescence, both to prevent accidental and possibly dangerous pregnancies, and to allow patients to come to terms with their future child-bearing potential. It is for this reason that the CHIA medical team has developed this information resource, along with a protocol and associated patient education materials.

Physiological Changes during Pregnancy:

There are many physiological changes that take place in a woman's body when she becomes pregnant. The impact of these physiological changes to pregnant women with cardiac disease will vary according to several factors and can result in increasing symptoms, morbidity or mortality. One of the first changes is that peripheral vasodilation that lowers blood pressure and provides less resistance for the heart to beat against. This causes cardiac output (the amount of blood being pumped out the heart) to increase. By the 20^{th} week of gestation, cardiac output increases by 40%, due to increased stroke volume and heart rate. Blood volume increases by up to 50% while systemic vascular resistance (the resistance to flow that must be overcome to push blood through the circulatory system) falls. As systemic vascular resistance falls, there will be increased right-to-left shunting which may reduce pulmonary perfusion. Coupled with the increased oxygen demand of pregnancy, this can lead to increasing hypoxia. This places additional strain on a failing heart and can significantly affect women who have a significant shunt. Pregnant women are also particularly susceptible pulmonary oedema due to physiological changes that affect pressures in the lungs.

Labor and delivery can further compromise cardiac function. Pain may lead to a tachycardia which reduces diastolic coronary filling as well as increasing circulatory catecholamines which increase

systemic vascular resistance. Labor leads to further increases in cardiac output by 15% in the first stage and 50% in the second stage due to the combination of auto-transfusion of 300-500ml of blood back into circulation with each uterine contraction. This can increase the risk of pulmonary edema in women with heart disease, and further compromise the cardiovascular system.

Table 1: Physiological changes during pregnancy.

Pregnancy Parameter:	Alteration:
Heart rate	Increased by 12bpm
Stroke Volume	Increased by 27%
Cardiac Output	Increased by 4.6-6.0 L/min
Blood volume	Increased by 25-50%
Plasma volume	Increased by 45-50%
Red Cell Mass	Increased by 20%
Glomerular Filtration Rate	Increased by 50%
Oxygen consumption	Increased by 21%
Minute ventilation	Increased by 48%
Tidal volume	Increased by 40%
Respiratory rate	Unchanged
SVR	Decreased to 1200-1250
Hematocrit	Decreased to 33-38%
Hemoglobin	Decreased to 11-12g/dL
FRC	Decreased by 18%
pCO2	Decreased to <30mmHg

Complications for women with Heart Disease:

As discussed above, there are many cardiorespiratory changes that occur during pregnancy. This can cause significant complications for women with congenital or acquired heart disease. Some of the more common complications are listed below:

1. Myocardial infarction (heart attack) – is a very real risk for women with cardiac conditions during pregnancy
2. Aortic dissection – common in women with Marfan's syndrome. It is a tear in the inner wall of the aorta that causes blood to flow between the layers of the wall of the aorta, forcing the layers apart.
3. Maternal and/or fetal death
4. Hemorrhage – impaired clotting factors and platelet function
5. Paradoxical embolism in those with Cyanotic Heart Disease as they shunt right to left
6. Heart failure – precipitated by the additional volume load of pregnancy on an already volume loaded ventricle.
7. Increasing cyanosis caused by the vasodilation of pregnancy
8. Potential problems at delivery:

a. Blood loss may be poorly tolerated since many women are unable to compensate by the usual response of increasing heart rate and stroke volume and may rely on venous return to maintain cardiac output
b. Bleeding may be increased due to concurrent anticoagulation
c. Pulmonary edema is more likely to occur peri-partum due to fluid shifts and as a consequence of additional IV fluids
d. Women with a right to left shunt may be at risk of air embolism
e. Acute pulmonary hypertension at the time of delivery can cause right ventricular failure and cardiac ischaemia. The risk of death is high in those with severe pulmonary hypertension, and severe morbidity can occur in those with relatively mild pulmonary hypertension.

9. Complications for the fetus:
    a. Spontaneous abortion
    b. Prematurity
    c. Low-birth weight - Babies may be smaller if the mothers heart does not pump as efficiently as it should and delivers less oxygen and nutrients to the placenta and developing baby
    d. Inheritance of congenital heart disease
    e. Stillbirth

Risk of Pregnancy:

There are many different congenital heart defects, and presentations of congenital heart disease and acquired heart disease. Furthermore, each of these different conditions can differ significantly in severity. For example women with conditions such as repaired Tetrology of Fallot may be low, medium or high risk depending on the underlying physiology, ventricular function, valvular function and history of corrective surgery. It is therefore necessary to distinguish risk of pregnancy in women with heart disease further, so that an informed decision can be made by CHIA and the girl as to her specific risks during pregnancy.

Table 2: Risk of complications according to maternal heart disease

Risk of complications	Maternal Cardiac Lesions
Low Risk	ASD, VSD, PDA, asymptomatic AS, AR with normal LV function, MVP, MR with normal LV function, mild or moderate MS without severe PH, mild or moderate PS
Intermediate Risk	Large left to right shunt, coarctation of the aorta, Marfan syndrome with a normal aortic root, moderate or severe MS, mild or moderate AS, severe PS, repaired Tetrology of Fallot without pulmonary insufficiency or stenosis, complex heart disease, disease with the anatomic right ventricle serving as systemic ventricle and mild left sided valve stenosis
High Risk	Eisenmenger's syndrome, unrepaired Tetrology of Fallot, severe PH, complex cyanotic heart disease, Marfan syndrome with aortic root or valve involvement, severe AS with or without symptoms, aortic or mitral

	valve disease (stenosis or regurgitation) with moderate or severe LV dysfunction. severe aortic stenosis or left ventricular outflow tract obstruction symptomatic ventricular dysfunction with an ejection fraction <40%

*High Risk conditions are associated with increased maternal an fetal mortality. Pregnancy is not advised.

*Cyanotic Heart Disease includes: unrepaired Tetrology of Fallot, total anomalous pulmonary venous connection, hypoplastic left heart syndrome, transposition of the great arteries, truncus arteriosus, tricuspid atresia, interrupted aortic arch, pulmonary atresia, pulmonary stenosis, Eisenmenger syndrome, late stage PDA.

CONGENITAL HEART DISEASE:

Atrial Septal Defects:

Atrial septal defects (ASD) are the most common cardiac lesions found in adults, making up between 10-17% of congenital heart defects. ASDs are often left uncorrected unless there is either a large hole or significant left to right shunt causing right ventricular overload and dilatation. If small or corrected, they cause no specific or significant problems during pregnancy. In an uncorrected defect, the effect of an increase in cardiac output on a volume-loaded right side is counterbalanced by the decrease in peripheral vascular resistance. The two main areas of concern are paradoxical embolus or arrhythmia. The former is rare but if the pregnant woman is immobilized or has other risk factors for thrombosis, the risk increases. The risk of atrial arrhythmias is well established with ASDs and remains even with correction. It is also suggested that pre-pregnancy correction of ASD can reduce the risk of miscarriage, pre-term labor and development/worsening of cardiac symptoms. Although most patients with ASD tolerate pregnancy well, secondary pulmonary hypertension may develop in a small subset of these patients by the time they reach adulthood. This in turn could lead to reversal of the shunt, cyanosis and increased morbidity and mortality.

These complications are rare and pregnancy is generally well tolerated in this group of patients. It is recommended that CHIA staff counsel girls with ASD that:

1. While pregnancy is well tolerated, there are risks, particularly with an uncorrected ASD, and this needs to be taken into consideration
2. Ensuring they inform their doctor that they have a corrected/uncorrected ASD
3. That they should have their ASD corrected before considering pregnancy to avoid maternal or fetal complications.

Ventricular Septal Defect:

Ventricular septal defects (VSD) are the commonest form of congenital cardiac lesion in children (1.5-3.5 per 1000 live births). Many of these defects do not require surgery and close spontaneously. Pregnancy is well tolerated in women with a corrected VSD. The only risk from a small haemodynamically insignificant VSD is of endocarditis, which is often prevented with prophylactic antibiotics. Some patients with a larger, uncorrected defect may develop congestive heart failure, arrhythmias, or pulmonary hypertension. These conditions have a higher risk in pregnancy and will be discussed later in this document.

It is recommended that CHIA staff counsel girls with VSD that:

1. While pregnancy is well tolerated, there are risks, particularly with an uncorrected VSD, and this needs to be taken into consideration
2. Ensuring they inform their doctor that they have a corrected/uncorrected VSD

3. That they should have their PDA corrected before considering pregnancy to avoid maternal or fetal complications.

Patent Ductus Arteriosis:
Patients who have corrected PDA generally have an uncomplicated course in pregnancy. Those with uncorrected PDA with a small or moderate sized ductus and normal pulmonary arterial pressure can also expect an uncomplicated pregnancy. In patients with a significant left to right shunt, secondary pulmonary hypertension might occur and result in the increased morbidity and mortality of Eisenmenger syndrome (discussed later).

It is recommended that CHIA staff counsel girls with PDA that:

1. While pregnancy is well tolerated, there are risks, particularly with an uncorrected PDA, and this needs to be taken into consideration
2. Ensuring they inform their doctor that they have a corrected/uncorrected PDA
3. That they should have their PDA corrected before considering pregnancy to avoid maternal or fetal complications.

Aortic Stenosis:
While all valves can be congenitally affected, the commonest lesion is that of a bicuspid aortic valve leading to aortic stenosis. This results in a fixed cardiac output. The majority of young women are asymptomatic and pregnancy is likely to be tolerated well. Most are able to increase cardiac output appropriately in order to compensate for the physiologic changes of pregnancy. With severe disease however, cardiac output can remain relatively fixed and even limited exercise may put these patients at risk for sudden cardiac or cerebral hypoxia, resulting in angina, myocardial infarction, syncope, or even sudden death. Patients reported to be at most risk are those whose aortic valve peak-to-peak systolic gradient is more than 100mmHg. Complications of Aortic Stenosis in pregnancy include can maternal mortality, stillbirth, premature birth, pulmonary oedema and persistant arrhythmia. Patients with severe AS are at greatest risk during termination of pregnancy of at delivery as these are times of significant potential for critical falls in cardiac output.

It is important to assess these women prior to pregnancy to determine whether it would be safer undergoing a pregnancy with the native valve or whether surgery should be contemplated prior to conception.

Pregnancy is likely to be well tolerated if:

1. Resting ECG has no ST segment depression.
2. On exercise testing, there is an appropriate increase in BP and HR and no ST change.
3. The pressure across the aortic valve measures less than 80 mmHg at peak and mean pressure gradient is less than 50 mmHg.
4. There is good left ventricle function.

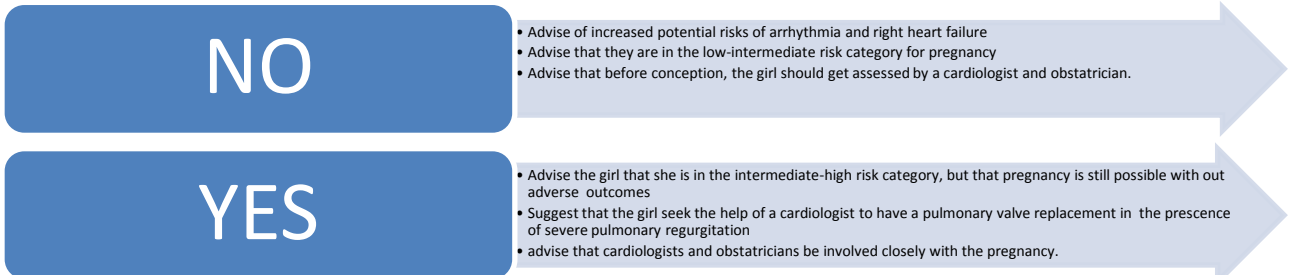
Complications for women with a Bicuspid Aortic Valve defect are exaggerated tachycardia, dyspneoa (painful breathing), angina (chest pain), pulmonary edema or ECG changes. Some women with bicuspid aortic valves have post stenotic dilatation of the aorta and as such are at risk of aortic dissection.

Repaired Tetralogy of Fallot

Women who have a repaired Tetralogy of Fallot have a moderate risk of complications during pregnancy. Despite the complexity of the original lesion, a successful correction is likely to lead to an uncomplicated, successful pregnancy. However, residual cardiac defects in combination with the circulatory changes observed in pregnancy may affect pregnancy in women with a corrected Tetrology of Fallot. The most common complications for a woman with a repaired Tetrology of Fallot in pregnancy are arrhythmias and right heart failure. The strongest predictor of developing right heart failure is the presence of severe pulmonary regurgitation after the correction has been performed. It is recommended that women who have severe pulmonary regurgitation and wish to conceive should have a pulmonary valve replacement. Also, if there is still the presence of a significant right ventricular outflow obstruction, or reduction in right ventricular function, the inherent risk of pregnancy is higher and women should be counseled appropriately.

It is recommended that CHIA staff:

1. Determine whether the girl with repaired Tetrology of Fallot has severe pulmonary regurgitation, a significant right ventricular outflow obstruction, or reduction in right ventricular function.



Repaired transposition of the great arteries

There are several surgical techniques that can be used to repair a transposition of the great arteries (TGA), including, the Mustard, Senning and Arterial Switch Operation. These operations have different risks in pregnancy, so it is therefore necessary to discuss them separately. If patients are well and have no deterioration in right (systemic) heart function, they will probably tolerate pregnancy well. However, right heart function as well as systemic tricuspid atrioventricular valve regurgitation may deteriorate in pregnancy.

The Mustard operation uses a conduit to direct pulmonary venous return into the right ventricle and transposed aorta, and the systemic venous blood via the mitral valve and left ventricle. Essentially, this allows blood to go in the right direction but through the wrong ventricle. Therefore the right ventricle and tricuspid valve support the systemic circulation with the same problems as in congenitally corrected transposition. The Senning operation is physiologically the same as the mustard. The difference is that with the Senning, the conduit is created from right atrial wall and atrial septal tissue whilst the Mustard creates the conduit using pericardium or synthetic material. The main complication from repaired transposition of the great arteries is narrowing of the pulmonary venous pathway because of the conduit structure. This fixed obstruction acts as a physiological equivalent of mitral stenosis. Several studies have shown that there are frequent cardiac, obstetric and fetal complications in pregnancy in women with Mustard or Senning repaired TGA. The most common cardiac complication is arrhythmias

(~20%), particularly supra ventricular tachycardias. In addition to the atrial scar tissue from the procedure to repair at TGA, the circulatory burden of pregnancy might trigger the development of an arrhythmia. Other complications include new onset heart failure, spontaneous miscarriages (24%), hypertension related disorders including pre-eclampsia, thrombo-embolic complications, premature rupture of membranes and low birth weight. The increased risk for adverse maternal and fetal outcomes places women who have had Mustard or Senning repaired TGA in the intermediate-to-high risk category for pregnancy. This condition does not have the prohibitively high complication, particularly in terms of mortality, of other congenital heart conditions. However, it is recommended that CHIA staff counsel these women very carefully, fully informing them of the inherent risks to the mother and baby of an attempted pregnancy. If a woman decides that she wishes to become pregnant, she will need very involved care from an experienced cardiologist and obstetrician.

Women who have had an Arterial Switch Operation (ASO), where the aorta and pulmonary artery are surgically 'switched' to their anatomically correct location, have significantly less risks than above. The ASO became the surgical approach of choice in the 1980's. However, surgical techniques in Vietnam can be significantly behind, and there may be many women of child bearing age who have had their TGA corrected using Mustard or Senning procedures. There are fewer risks with this group of women because there is less intra-atrial manipulation, so therefore less scarring and potential for damage, and the left ventricle is working as it is anatomically intended to. However, increase of myocardial oxygen demand and the increased potential for anaemia during pregnancy theoretically may place women who have undergone the ASO at increased risk of complications during pregnancy. These women require complete assessment before or early in pregnancy. However, providing the right ventricular function is good, pregnancy is usually well tolerated. These women have a Low Intermediate risk in pregnancy.

Repaired Coarctation of the Aorta:

Coarctation of the aorta has a prevalence of 0.3-1 in 1000 in the female population. It is usually women with repaired coarctation who present for pregnancy. There is no contraindication for pregnancy with repaired coarctation. More recently collected series in patients reveal a low maternal mortality (0%-3.5%) with a good fetal outcome.

In patients with uncorrected coarctation, pregnancy was once thought to carry such a severe risk to life that termination of the pregnancy and sterilization were recommended. However, current medical opinion is that these women can safely carry a pregnancy to full term with adequate medical care. In a Vietnamese context, it is important to assess the severity of the woman's condition, and the availability of adequate care when advising on how to proceed during a pregnancy.

Blood pressure during pregnancy in patients with uncorrected coarctation of the aorta usually follows a similar pattern to that of patients with essential hypertension in pregnancy; that is, it is unchanged or falls in the second trimester, with a return to baseline levels near term. Aortic rupture or dissection is a serious concern in patients with coarctation of the aorta. Correction of coarctation of the aorta in pregnancy with successful maternal and fetal outcomes has been reported. Risks in pregnancy are increased in patients with associated cardiac lesions such as septal defects or bicuspid aortic valves as well as in patients with aortic, intervertebral or Circle of Willis aneurysms. Recoarctation can also occur.

Cyanotic Heart Disease:

Even though these women do not have pulmonary hypertension, they are often still very unwell and are therefore are at high risk of mortality and morbidity in pregnancy. Women in this category have diseases such as unrepaired Tetrology of Fallot, unrepaired Transposition of the Great Arteries; hypoplastic left heart and Eisenmenger's syndrome. Many people in this category will develop pulmonary hypertension at some time.

In complex cyanotic heart disease without PH pregnancy can cause the following complications:

1. Decreasing ventricular function
2. Hemorrhage due to impaired clotting factors and platelet function
3. Paradoxical embolus as all cyanotic patients shunt from right to left
4. Heart failure
5. Increasing cyanosis secondary to the vasodilatation of pregnancy.

Cyanosis is associated with adverse effects on the fetus with an increased incidence of low birth weight, prematurity and fetal loss (see Table 2.4). These patients need careful monitoring in a unit that understands the complex physiology of the maternal circulation. Women often require bed rest and oxygen which may reduce cyanosis and, as such, improve fetal well-being. While spontaneous vaginal delivery is best for the mother, fetal growth restriction or fetal distress often precipitates a Cesarean section. It is important in these situations to maintain the circulatory volume and avoid significant vasodilatation. It's been shown that maternal oxygen saturation has a direct link to the livebirth rate of offspring.

Maternal Oxygen Saturation	Live Birth Rate
>90% SpO2	92%
85-90% SpO2	63%
<85% Sp02	12%

It is important to consider some of these Cyanotic Heart Diseases separately. There is no evidence of pregnancy in women with an unrepaired transposition of the great arteries, so if CHIA comes across adolescents with this condition, follow the recommendations for an unrepaired Tetrology of Fallot. However, all women who have a cyanotic heart disease carry an inherently high risk of pregnancy, and should therefore be advised to consider contraception, and avoidance of pregnancy.

Unrepaired Tetrology of Fallot:
There is very little evidence on pregnancy in this group of women, because it is rare for Tetrology's to be left unrepaired in many countries, and those that are unrepaired rarely reach child bearing age. Prognosis for people who do not undergo surgery for this condition is poor. Most commonly, the experience pulmonary haemorrhage, brain abscesses and thrombo-embolic complications. Before the introduction of surgical repair, few patients with T4F reach childbearing age and successful pregnancy was uncommon. Pregnancy in an unrepaired Tetrology of Fallot carries a major risk of maternal complications including heart failure, arrhythimia, and endocarditis, which can give rise to fetal problems including miscarriage and preterm labour.

It is recommended that CHIA staff advise young women with an unrepaired Tetrology of Fallot:

1. to avoid pregnancy due to high risks to the mother and baby
2. to begin appropriate contraception

3. Advise that if they do become pregnant they consider medical termination as a life saving measure

Eisenmenger's Syndrome:
Eisenmenger's syndrome is a broad term applied to any cardiac anomaly in which the pathological process of increased pulmonary flow leads to obliterative pulmonary vascular disease. The result is pulmonary hypertension (PH), which is severe enough to reverse the normal left-to-right shunt in the heart to right-to-left. The PH is usually irreversible even after corrective surgery. 5-10% of patients with congenital heart disease will develop some degree of pulmonary hypertension. Women with Eisenmenger's Syndrome have a very high risk of maternal/fetal morbidity and mortality during pregnancy. Maternal mortality has been reported to be 30-50% and fetal mortality at up to 40%. At least 30% of infants show evidence of intrauterine growth restriction.

It is recommended that CHIA staff advise young women with an Eisenmenger's Syndrome:
1. to avoid pregnancy due to high risks to the mother and baby
2. to begin appropriate contraception
3. Advise that if they do become pregnant they should consider medical termination as a life saving measure

Single Ventricle Congenital Heart Defect:
A single ventricle usually occurs when there is a large ASD which allows free mixing of oxygenated blood from the lungs with deoxygenated blood from the Vena Cava. It can also occur when there is tricuspid atresia, which means no direct connection between the right atrium and the right ventricle. Or there is a VSD which allows free mixing of blood in the left and right ventricles.

The combination of chronic hypoxaemia (low blood oxygen) before palliation and chronic venous congestion afterwards may influence ovarian function resulting in menstrual cycle disorders or infertility. For those who do become pregnant, the most common obstetric complications are preterm rupture of membranes and premature labour. There is a significant risk of neonatal death and postpartum haemorrhage.

It is recommended that CHIA staff advise girls with this condition:
1. The possibility that they might be infertile as part of their condition. Encourage the girls to get tested either at Marie Stopes or Danang Hospital
2. If they are not infertile, explain the high risk of pregnancy for them and their baby
3. Advise the use of contraception
4. Advise that if they do become pregnant they should consider medical termination as a life saving measure

Post-Fontan procedure
The Fontan circulation was developed for patients with a functional single ventricle. The single ventricle supports the systemic circulation while the systemic venous return is directed to the pulmonary artery directly through a conduit. This means there is no pump directing the blood through this shunt. While this repairs any shunts and thus abolishes cyanosis, the circulation is prothrombotic (increasing likelihood of blood clotting), has a limited ability to increase cardiac output and poorly tolerates atrial arrythmias and hypovolemia. However, in those women with good ventricular function and few symptoms, pregnancy may be well tolerated.

Pulmonary Hypertension:

Pulmonary Arterial Hypertension (PAH) of any cause complicated by pregnancy carries a 30-60% risk of maternal death and it is therefore considered too dangerous for these women to become pregnant. Women with PH should be strongly advised against pregnancy and adequate contraception is imperative (discussed later). The physiologic changes of pregnancy are poorly tolerated in women with PAH. Women with PAH are unable to make the physiologic adjustments that are needed to compensate for increased blood volume, increased stroke volume and a decrease in pulmonary vascular resistance. This can result in progressive right ventricular failure and sudden death from arrhythmias. During delivery, the pain of contractions is associated with increases in heart rate, blood pressure and oxygen consumption. In women with PAH this can result in vagal responses, leading to hypotension and sudden death. Pregnancy is also associated with a hypercoagulable state (blood clotting) which can increase risks of thromboembolism. It has been reported that Eisenmenger Syndrome and primary PAH have a similar mortality rate in pregnancy women of 30-36%. Secondary PAH can have a mortality rate of 50%.

Women with PH who do become pregnant should be advised of the high risk of mortality and termination of pregnancy should be advised. Termination of pregnancy can be performed medically or surgically depending on facilities available. A medical termination can be performed at any gestation. There are risks to termination, however, in women with pulmonary hypertension, and these women should be monitored closely. The procedure should be performed in a high dependency area with full monitoring. The risks of a vagal response, uterine perforation and bleeding are reduced with skilled operators. If women decline termination of pregnancy, they require careful management with a multidisciplinary team with expertise in PAH. Most women with Eisenmenger's Syndrome who die as a result of pregnancy, do so soon after delivery. There is no evidence that neither Cesarean versus vaginal delivery, nor regional versus general analgesia/anesthesia reduces this risk. The dangers relate to increasing the right-to-left shunt and escalating pulmonary hypertension often despite intensive and appropriate care. Postpartum management should be in an intensive care environment by intensivist anesthetists, cardiologists and obstetricians.

ACQUIRED HEART DISEASE:

Valvular Diseases:
In young women most cases of acquired valvular heart disease are due to rheumatic heart disease (RHD), or previous endocarditis. RHD accounts for 90% of cardiac disease in pregnancy worldwide. Approximately 3% of people with untreated group A streptococcal pharyngitis will subsequently develop rheumatic fever. In RHD, mitral stenosis is the most common lesion and potentially the most likely to cause maternal and fetal compromise. In patients with RHD, the mitral valve is most commonly affected. Mitral stenosis occurs in approximately 90% of patients and mitral regurgitation in 7%. The aortic valve is the second most commonly affected valve, although to a lesser degree. Aortic regurgitation is present in 2.5% of patients and aortic stenosis in only 1%. Although the tricuspid and pulmonary valves may also be affected, such involvement is almost always combined with mitral or aortic disease.

Mitral stenosis.
In mitral stenosis, the restriction in outflow from the left atrium results in higher left atrial pressures, left atrial enlargement and eventually right sided heart failure. The reduced blood to the left ventricle causes stroke volume to be reduced. Heart rate then increases to maintain cardiac output. This will reduce the time for filling of the left ventricle in diastole, and results in further increases in left atrial

pressure. This will ultimately cause pulmonary edema and Pulmonary Hypertension. For patients with moderate uncomplicated mitral stenosis, symptoms may peak at around 20-24 weeks gestation as cardiac output and intravascular volume peaks and then stabilizes.

Many patients with mild mitral stenosis are asymptomatic until they become pregnant. At that time, prenatal physiologic changes contribute to an increase in left atrial pressure and subsequent symptoms of dyspnea (painful breathing) and eventually tachpnea (fast breathing), orthopnea (shortness of breath) and paroxysmal nocturnal dyspnea (painful breathing at night). Management of mitral stenosis in pregnancy depends on the level of symptoms. Patients who are asymptomatic may continue, within reason, their normal activities or living. Patients who develop dyspnea with exercise must restrict their activities and depending on the severity, may require continuous bed rest. In patients with severe mitral stenosis, pulmonary edema usually occurs at 30 weeks gestation. A blood clot in the left atria can occur due to stagnation of blood flow. If the clot dislodges, a stroke or other serious complications can occur. The patient may also develop secondary pulmonary hypertension

Mechanical valve replacement might be necessary to improve long term outcomes in pregnancy for the mother and fetus. However, it is important to discuss this issue before women become pregnant. The success of open valve replacements in pregnancy for severe mitral stenosis is similar to the non-pregnant state, but the stillbirth rate is 10-30%.

It is recommended that CHIA staff:

1. Advise the woman that pregnancy will likely be well tolerated
2. Advise that the woman should seek the advice of a cardiologist before become pregnant to discuss further treatment options including valve replacement, medication or any complications.

Mitral Regurgitation:

Mitral Regurgitation (MR) is a condition in which the incompetent mitral valve allows a portion of the left ventricular stroke volume to enter the left atrium. It occurs in 7% of people who have RHD. Many patients also have an associated mitral stenosis. Pregnancy is generally well tolerated by patients with mitral regurgitation and it has been well theorized that mitral regurgitation may actually improve in pregnancy because of the physigiologic reduction in systemic vascular resistance which promotes better cardiac output and less regurgitation. There are relatively rare complications in pregnancy except for in patients who have severe MR. Those who have severe MR have a 7% risk of mortality during pregnancy. Previously asymptomatic women may worsen immediately after delivery because of sudden increases in systemic vascular resistance. It has been suggested that these patients are at increased risk of left atrial enlargement and subsequent atrial fibrillation.

It is recommended that CHIA staff:

1. Advise the woman that pregnancy will likely be well tolerated
2. Advise that the woman should seek the advice of a cardiologist before become pregnant to discuss further treatment options including valve replacement, medication or any complications.

Aortic Regurgitation:

Aortic Regurgiation (AR) involves a faulty valve that allows a portion of the stoke volume to travel backwards into the left ventricle during contractions. As with mitral regurgitation, this lesion is generally well tolerated in pregnancy.

As with mitral regurgitation, it is thought that the physiologic changes in pregnancy can improve regurgitation. However, in severe cases, long standing AR can lead to left ventricular dilation and dysfunction.

It is recommended that CHIA staff:

1. Advise the woman that pregnancy will likely be well tolerated
2. Advise that the woman should seek the advice of a cardiologist before become pregnant to discuss further treatment options including valve replacement, medication or any complications.

Artificial heart valves

Cardiac valve replacement is indicated when a patient with valvular disease has symptoms of heart failure and a lower quality of life. With many women, having a cardiac valve replacement can reduce their risk of serious complications during pregnancy. However, there are still risks involved when a woman has a mechanical cardiac valve and becomes pregnant.

There is a 3-5% mortality associated with pregnancy if a woman conceives with a mechanical valve in place. Thrombosis is a major complication of artificial valves, and the main issue concerning management of pregnant women with mechanical prosthesis is anticoagulation. This needs to be handled by an experienced doctor.

It is important for women who have mechanical cardiac valves to be informed of their risks before they decide to have children.

Marfan Syndrome:

Marfan syndrome is rare, with an incidence of 5/100 000. Mortality rates as high as 50% have been reported for pregnant patients. However, those women with a documented aortic root diameter of less than 40mm without an abnormal aortic valve have a mortality rate of less than 5%. Aortic or mitral regurgitation is also seen in 60% of patients with Marfan syndrome and may complicate pregnancy.

Aortic dissection (a tear in the aorta causes blood to flow through the layers of the aorta, forcing them apart). The risk of aortic dissection during pregnancy or shortly thereafter was 17% in one series of 36 women without cardiac symptoms prior to pregnancy. Pregnancies complicated by Marfan syndrome are not associated with poor perinatal outcomes, though some suggest an increased risk of incompetent cervix. Patients should be counseled regarding the risk of autosomal dominant inheritance and the need for follow-up for their offspring.

CONTRACEPTION IN HEART DISEASE:

Certain contraceptive methods are associated with unacceptable increases in risk for specific cardiac conditions, and it is important for women with congenital or acquired heart disease to choose the most appropriate and safe contraception for them. It is important that all contraception be discussed individually with a specialized doctor, preferably from the Marie Stopes Clinic. Different women with cardiac conditions will need different forms of contraception for maximum safety and efficacy.

Combined Oral Contraceptives (COC):

COC is a safe effective and popular method of contraception. It combines both estrogen and progestogen. However, the estrogen component is associated with increased risk of arterial and venous thromboembolism, which could limit the use of COC in some women with cardiovascular disease. Women whose cardiac status in prothrombotic may be at particular risk. Women with right to left shunts due to cyanotic heart disease or pulmonary arteriovenous malformations are at risk of paradoxical embolism and stroke if they develop thrombosis whilst on COC.

Women with the following conditions should not use COC

1. Arterial fibrillation or flutter if not anticoagulated
2. Pulmonary hypertension or pulmonary vascular disease e.g. Eisenmenger's syndrome
3. Dilated left atrium >4cm
4. The Fontan heart
5. Cyanotic heart disease, even taking warfarin
6. Pulmonary arteriovenous malformation
7. Poor LV function of any cause
8. Coronary artery disease

Progestogen only contraceptive (POC):

Contraceptive doses of progesterones used alone are not associated with an increased risk or arterial or venous thrombosis. Therefore all progestogen only methods are usable when there is an arterial or venous thrombotic risk, and broadly speaking are safe for all form of heart disease. Progestogen only preparations may produce irregular menstrual bleeding

Progestogen Only Pills (POP)

POP must be taken at the same time each day to achieve good contraceptive effect, since unlike COC they do not inhibit ovulation.Therefore, although safe in cardiac disease, their use is generally not recommended for those with major heart disease where maximum efficacy is needed.

Implanon:

This progesterone implant has no cardiac contraindications. It's as effective as sterilsation and produces steadier blood levels (and generally less side effects). It needs replacing every 3 years. 20% of women using implanon became amenorrhoeic. The efficacy of implanon is also affected by bosentan so a supplementary method of contraception should be used.

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