HEART DISEASE & PREGNANCY

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CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

Background

Congenitally corrected transposition of the great arteries (ccTGA) is characterized by atrioventricular and ventriculoarterial discordance. This results in atria that are connected to the opposite ventricle and ventricles are connected to the" wrong" great artery. As a result of these anatomic abnormalities, patients have a morphologic right ventricle and a tricuspid valve in the subaortic (systemic) position.

Frequently, ccTGA is associated with additional heart defects, which are important predictors of outcome. The most common associated lesions are ventricular septal defects (60-80%), pulmonary stenosis (30-50%), and tricuspid valve abnormalities (14-56%). Often women with ventricular septal defects and pulmonary stenosis will have had intracardiac repair such as ventricular septal defect closure and valve or non-valved conduit repair of the outflow obstruction. Less frequently, women will have had a double switch operation.

There are associated conduction system abnormalities such as complete heart block, which have an impact on prognosis.

Survival into adulthood is common, either after operation or with isolated ccTGA. Late complications include progressive tricuspid regurgitation, systemic right ventricular dysfunction, and heart failure. The incidence of complete heart block is 2%/year. Sudden cardiac death is relatively uncommon.

Effects of pregnancy-related hemodynamic changes

In ccTGA the morphologic right ventricle supports the systemic circulation. In some instances, this is associated with ventricular dysfunction. During pregnancy, cardiac output increases by 50% reaching its peak at around 28-32 weeks of gestation (see Cardiovascular Changes During Pregnancy). The increased cardiac output can result in heart failure in women with impaired subaortic (systemic) right ventricular function. Dilation of the systemic right ventricle can result in progressive tricuspid regurgitation.

The fall in peripheral vascular resistance that occurs during pregnancy can augment right-to-left shunting in women with a VSD and pulmonary stenosis, resulting in worsening maternal cyanosis and hypoxemia.

Maternal complications

Pregnancy in women with ccTGA may be well tolerated. The overall risk depends largely on associated lesions, systemic ventricular function, function of the systemic atrioventricular valve (tricuspid valve), and the women's functional class at the time of conception. Heart failure (7.1%) and arrhythmias (3.6%) are the most common maternal cardiac complications reported during pregnancy in women with ccTGA. (1-5). Other complications have been described, although they occur less commonly, including worsening of

cyanosis, cerebrovascular events, and endocarditis. Other cardiac characteristics can have an impact on outcomes. (see General Considerations)

Worsening tricuspid regurgitation, right ventricular dilation, and systolic dysfunction have been described in women with systemic right ventricles after Mustard or Senning operations. (6) The late effect of pregnancy on systemic right ventricular function in women with ccTGA has not been well studied, but potentially the subaortic right ventricular function could deteriorate after pregnancy.

Some women with high-risk cardiac lesions may seek alternatives to pregnancy such as adoption.

Fetal complications

Women with no cyanosis are at risk for premature delivery (9% of pregnancies). Cyanotic women with unrepaired ccTGA/ventricular septal defects/pulmonary stenosis are at substantial risk for miscarriage as well as prematurity (1). The incidence of spontaneous abortion increases proportionally with the degree of maternal hypoxemia. Fetal deaths have been reported.

Management strategies

Preconception counseling/ Contraceptive methods

Successful pregnancies are reported in women with ccTGA; however, preconception risk stratification is important. Systemic ventricular function, tricuspid valve function, and associated lesions are important variables to consider when determining an individual's risk. There are other cardiac characteristics, which can also have an impact on outcomes (see General Considerations). The long-term maternal cardiac impact of completed pregnancy in women with a subaortic right ventricle remains unclear.

Ideally, a comprehensive cardiovascular examination should be undertaken before embarking on pregnancy. This includes a careful history and physical examination, an electrocardiogram and an echocardiogram. The additional prognostic benefit of cardiopulmonary exercise testing has not been defined, but it can useful to assess functional status and ability to increase heart rate during exercise. Other imaging modalities such as cardiac magnetic resonance imaging may be useful in select cases. Catheterization may be indicated for women requiring surgery prior to pregnancy or if there are other unaddressed hemodynamic issues.

Transmission of congenital heart disease to offspring should be discussed. The risk of transmission of congenital heart disease is approximately 5%, compared to a background risk of approximately 1% of having a baby with congenital heart disease.

A discussion about contraceptive methods is appropriate in all women with ccTGA. The combined oral contraceptive pill (containing estrogen) is associated with an increased risk of thromboembolism and is contraindicated in women with significant systemic ventricular dysfunction, arrhythmias, right-to-left shunts, cyanosis or older style mechanical valves. Progesterone-only forms of contraception are not associated with thromboembolic risk and can be suitable alternatives. (see Contraception)

Women treated with angiotensin converting enzyme inhibitors or angiotensin receptor blockers will need to have these medications stopped prior to pregnancy. Assessment of ventricular function after discontinuation of therapy is useful. Medication use should be reviewed if a woman is contemplating pregnancy or is pregnant. The MOTHERISK website (http://www.motherisk.org) is an excellent resource.

Ante-partum care

Coordinated care with a congenital heart disease specialist and a high-risk obstetrician should be implemented. The frequency of assessments (clinical and echocardiographic) during pregnancy should be determined on the basis of the presence of associated cardiac lesions, the degree of systemic ventricular dysfunction, and the women's functional status at the time of conception.

For women with isolated ccTGA and good functional capacity, the overall risk of adverse maternal pregnancy-related events is low. Systolic ventricular function and atrioventricular valve function may deteriorate because of the hemodynamic changes of pregnancy and therefore, serial echocardiographic follow up should be performed during pregnancy. The frequency of clinical follow up and echocardiograms will be based on the clinical status of the women. In women with symptoms of heart failure, cyanosis or arrhythmia, and in women with a prosthetic valve, close cardiovascular monitoring is essential throughout pregnancy and the peripartum period.

Treatment for symptomatic heart failure may be necessary in some women. The role of beta blockers in women with impaired subaortic (systemic) right ventricular systolic function is not known, but they may be helpful.

Supraventricular arrhythmia can be treated medically or with DC cardioversion when women are unstable or unresponsive to medical therapy (see Arrhythmias).

Hypovolemia can lead to shunt reversal, reduced cardiac output, and refractory hypoxemia in women with an associated ventricular septal defect/pulmonary stenosis. Volume overload should be avoided in women with impaired subaortic (systemic) right ventricular function, as it can result in heart failure.

Women should be offered fetal echocardiography at approximately 20 weeks gestation.

Labour and delivery

Labour and delivery should be planned carefully with a multidisciplinary team well in advance. It is important to communicate the delivery plan to the woman and to other physicians involved in her care. The best delivery plan is not useful if information is not readily available when needed.

Generally, vaginal deliveries are recommended unless there are obstetric indications for a cesarean delivery. Good pain management for labour and delivery is very important in order to minimize maternal cardiac stress. To decrease maternal expulsive efforts during the second stage of labour, forceps or vacuum delivery is often utilized. To decrease potential harmful complications from difficult mid cavity-assisted delivery, uterine contractions are often utilized to facilitate the initial descent of the presenting part.

The need for maternal monitoring at the time of labour and delivery is dictated by the women's functional status, the presence of cyanosis, and the associated lesions. While cyanotic women may require invasive monitoring, most women with ccTGA do not. To detect potential arrhythmias early, continuous monitoring with electrocardiography may be helpful in some instances.

In general, endocarditis prophylaxis at the time of labour and delivery is not recommended in women ccTGA. However, some experts continue to administer antibiotics because they feel that the risks of adverse reactions to antibiotics are small and the risk of developing endocarditis has major health consequences.

In women with residual interatrial or interventricular shunts, air-particulate filters (bubble trap filters) are recommended for all intravenous lines, particularly in the setting of pulmonary stenosis/VSD with right-to-left shunts.

Post-partum care

The hemodynamic changes of pregnancy may take up to six months to normalize. Women should be seen early after pregnancy (usually within 6-8 weeks). The frequency of additional follow up visits should be dictated by the clinical status of the women.
Because ventricular function may deteriorate after pregnancy, postpartum echocardiographic follow up is important.

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