HEART DISEASE & PREGNANCY

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TETRALOGY OF FALLOT

Background

Tetralogy of Fallot is the most common cyanotic congenital heart defect and accounts for about 3.5% of all congenital cardiac lesions. It is characterized by four features: 1) a large, non-restrictive ventricular septal defect; 2) the aorta 'overrides' the interventricular septum; 3) right ventricular outflow tract obstruction; and 4) right ventricular hypertrophy. Rather than being a single entity, tetralogy of Fallot represents a spectrum of disease, ranging from well balanced circulation with normal systemic saturations ('pink Fallot') to patients with pulmonary atresia in which pulmonary perfusion is via aorto-pulmonary collaterals at the other end of the spectrum.

Most women with tetralogy of Fallot embarking on pregnancy have undergone intracardiac repairs in childhood. After intracardiac repair patients become acyanotic; however, many have residual hemodynamic lesions, the most common of which are right ventricular outflow obstruction and pulmonary regurgitation. Over time these lesions can lead to right ventricular dilatation and/or right ventricular dysfunction.

Effects of Pregnancy-Related Hemodynamic Changes

Pregnancy is associated with significant hemodynamic changes including an increase in blood volume and cardiac output. (see Cardiovascular Changes During Pregnancy) The ability of a woman with repaired tetralogy of Fallot to tolerate these changes depends on the severity of residual lesions (i.e. valve stenosis/regurgitation and size and function of the right ventricle) at the time of conception.

In contrast, women with unrepaired tetralogy of Fallot are cyanotic. Their lung perfusion and consequently their arterial oxygen saturation depends on the balance between pulmonary and systemic vascular resistance. The hemodynamic changes of pregnancy can disrupt this balance, which results in worsening cyanosis.

Maternal Complications

Pregnancy in women with repaired tetralogy of Fallot is usually well tolerated. The risk for maternal cardiac complications depends on the severity of residual lesions at the time of conception. Women with a history of arrhythmias or heart failure prior to pregnancy are at increased risk for complications during pregnancy. Other cardiac characteristics can have an impact on outcomes (see General Considerations). The most common complications are arrhythmias (6.4%) and heart failure (2.4%). (1,2,3,4,5,6,7,8, 9) Severe pulmonary regurgitation and subpulmonic right ventricular dysfunction have also been shown to be risk factors for adverse pregnancy outcomes. (2)

The late effects of pregnancy on the heart are not known. One study demonstrated that pregnancy was associated with an increase in the subpulmonary ventricular size late after pregnancy in women with repaired TOF. (10)

In contrast, women with unrepaired tetralogy of Fallot are cyanotic and at high risk for adverse outcomes during pregnancy. (5) Although successful pregnancies in women with unrepaired tetralogy of Fallot are reported, when possible women should undergo corrective surgery prior to embarking on pregnancy.

Fetal Complications

Women with repaired tetralogy of Fallot have a risk of prematurity and small-for-gestational-age babies of 6.3% and 9% respectively.

Women with unrepaired tetralogy of Fallot have cyanosis. The risks of prematurity (44.6%) and small for gestational age babies (66.7%) are increased in women with cyanotic heart disease. (9) This risk increases with lower resting oxygen saturations.

Management Strategies

Preconception Counseling/Contraceptive Methods

Most women with repaired tetralogy of Fallot can have successful pregnancies. The risk of maternal cardiac complications depends on the severity of residual lesions at the time of conception. The most common complications are symptomatic right heart failure and arrhythmias. Maternal death is rare. (1,2,3,4,5,6,7,8,9)

A comprehensive cardiovascular examination should be undertaken before embarking on pregnancy. This includes a careful history and physical examination, an echocardiogram and an electrocardiogram, and may also include a cardiac magnetic resonance imaging study. The additional prognostic benefit of cardiopulmonary exercise testing has not been defined, but may be helpful in some cases.

Women with unrepaired tetralogy of Fallot suitable for surgical repair should be offered corrective surgery prior to embarking on pregnancy.

Transmission of congenital heart disease to offspring should be discussed. The risk of transmission of congenital heart disease is approximately 5-50%, compared to a background risk of 1% of having a baby with congenital heart disease. The risk of transmission is dependent on associated genetic syndrome. For instance, in women with microdeletions of chromosome 22q11, the risk of transmission to offspring is 50%. Genetic counseling is recommended for women with 22q11 deletion syndromes and for women with a family history of congenital heart disease or with other congenital defects.

A discussion about contraceptive methods is appropriate in all women with repaired or unrepaired tetralogy of Fallot. Combined hormonal contraceptives are contraindicated in women with unrepaired tetralogy of Fallot. (see Contraception)

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 between a congenital heart disease specialist and a high-risk obstetrician at a high-risk pregnancy center should be implemented. The frequency of follow-up visits is dictated by women's functional status and the residual lesions at the time of conception.

Women with repaired tetralogy of Fallot and few or no residual lesions are at low to intermediate risk for complications. At the other end of the spectrum, women with significant residual lesions, ventricular dysfunction, or unrepaired tetralogy of Fallot require close follow by a dedicated multidisciplinary team of experienced cardiologists, high-risk obstetricians, and anesthetists.

All women should be offered fetal echocardiography at 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.

Vaginal delivery is recommended in most instances. 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. In patients with unrepaired tetralogy of Fallot, oxytocic drugs such as oxytocin, which induces vasodilation and arterial hypotension, should be avoided.

The need for maternal monitoring is dictated by residual lesions and functional status of women. While cyanotic women with unrepaired tetralogy of Fallot may require invasive blood pressure monitoring, most women with repaired tetralogy of Fallot do not require special monitoring.

In general, endocarditis prophylaxis at the time of labour and delivery is not recommended in women with repaired tetralogy of Fallot. 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 patients with residual interatrial or interventricular shunts, air-particulate filters (bubble trap filters) are recommended for all intravenous lines.

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.

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