HEART DISEASE & PREGNANCY

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AORTIC STENOSIS

Background

The etiology of aortic valve stenosis (AS) in women of childbearing age is most commonly related to congenitally bicuspid aortic valves. In developing countries, AS from rheumatic heart disease may be more common and is usually associated with mitral valve disease. The classic symptoms associated with AS include: heart failure symptoms, chest pain, and presyncope/syncope. Severe AS is defined as: 1) an aortic valve area < 1 cm^2, 2) a mean gradient across the aortic valve >40 mmHg, or 3) a peak gradient across the aortic valve >64 mmHg by echocardiography.

In addition to AS, women with bicuspid valves may have an associated aortopathy or aortic dilatation even in the absence of hemodynamically significant aortic stenosis or regurgitation. Aortic coarctation can occur in conjunction with bicuspid aortic valve disease. Left ventricular outflow tract obstruction from other causes such as supravalvular stenosis (with or without Williams-Beuren syndrome) or subvalvular stenosis (discrete membrane or tunnel-type) and are physiologically similar to aortic valve stenosis.

Effects of Pregnancy Related Hemodynamic Changes

Pregnancy is associated with significant hemodynamic changes including increases in blood volume and cardiac output. (*see* Cardiovascular Changes During Pregnancy) Adapting to these changes can be difficult in the setting of a fixed outflow tract obstruction. Women with significant aortic stenosis who do not adapt adequately to the increased demands of pregnancy may develop exercise intolerance, heart failure, angina, palpitations, or presyncope/syncope.

Women with left ventricular dysfunction are at a greater risk due to a greater inability to accommodate the increased hemodynamic load of pregnancy. Symptoms tend to occur at the end of second trimester or in the third trimester when cardiac output has peaked. A further transient increase in cardiac output occurs at the time of delivery, and may lead to cardiac decompensation at that time. The hypertrophied ventricle that accompanies AS is sensitive to abrupt changes in preload so vasodilation from anesthetic agents or hemorrhage around the time of labour and delivery can destabilize cardiac function more profoundly in the patient with aortic stenosis.

Maternal Cardiac Complications

Women with mild or moderate AS generally do well during pregnancy (1,2,3,4). In a recent series of 49 pregnancies in women with congenital AS the overall maternal cardiac complication rate was 6%, and increased to 10% in the presence of severe AS (2); no maternal death was reported in this study. The ZAHARA investigators reported a 9.4% rate of maternal complications in AS (3.8 % heart failure, 5.7% arrhythmia; no death) (3). Heart failure typically occurs only in those women with severe AS. In the 3 contemporary series of women with AS during pregnancy, 1 maternal death was reported, which occurred 10 days postpartum in conjunction with aortic valve replacement in a woman with severe AS and coarctation (1,2,3). Other cardiac characteristics can also have an impact on outcomes (see General Considerations).

In women with bicuspid aortic valve, risk of aortic dilatation or dissection during pregnancy has been described, however few data are available to help guide risk stratification. (5) Guidelines suggest that women with aortic root diameters greater than 45 mm should be counselled about the high-risk nature of pregnancy.

Fetal Complications

Women with AS are also at risk for fetal complications. In a series of 49 pregnancies in women with congenital AS, 12% of pregnancies were complicated by adverse fetal and neonatal outcomes: premature birth (8%), small for gestational age (2%), and respiratory distress syndrome (6%). (1) Higher rates of neonatal complications (24.5%) have been reported by other groups. (3)

Management Strategies

Preconception counseling/Contraceptive methods

Severity of aortic stenosis and function of the left ventricle are important determinants of outcomes during pregnancy. Women who with a history of heart failure or arrhythmias are at higher risk for complications during pregnancy. Other cardiac characteristics can also have an impact on outcomes (see General Considerations).

Women with severe symptomatic AS should be considered for intervention prior to pregnancy. In asymptomatic women, time of intervention needs to be tailored to the individual.

A discussion about contraceptive methods is appropriate in women with aortic stenosis. In general, most forms of contraception are safe in women with AS. (*see Contraception*)

Transmission of congenital heart disease to offspring should be discussed. The risk of transmission of congenital heart disease is approximately 10%, compared to a background risk of approximately 1% of having a baby with congenital heart disease.

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.

Given the aortopathy associated with bicuspid aortic valve, some experts suggest therapy with beta-blockers during pregnancy in women whose aortas are dilated to reduce the chance of aortic root dilation or dissection.

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 and echocardiograms should be dictated by women's functional status and the severity of her AS.

Women with significant AS require close follow up by a dedicated multidisciplinary team of experienced cardiologists, high-risk obstetricians, and anesthetists.

Women who develop cardiac symptoms, have a decrease in left ventricular systolic function, or have a fall in the peak aortic velocity, may need to be considered for intervention. Initial therapy should include bed rest, treatment of aggravating factors such as anemia, and treatment of heart failure or arrhythmias. Interventions, specifically valvuloplasty or valve surgery may be needed in women with

refractory symptoms. These procedures should only be performed by operators and in centres with expertise in the procedures. Valvuloplasty during pregnancy exposes the fetus to radiation. Special attention should be given to minimizing radiation exposure in the fetus by shielding the gravid uterus and keeping fluoroscopy time to a minimum. Cardiopulmonary bypass surgery for aortic valve replacement during pregnancy carries a high risk of fetal loss.

All women with congenital AS 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.

Generally, vaginal delivery is recommended. Good pain management for labour and delivery is important in order to minimize maternal cardiac stress. Care should be taken to avoid drops in blood pressure that can occur with anesthesia. 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 is dictated by the severity of AS and functional status of the women. Women with significant AS may require invasive blood pressure monitoring.

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

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 women with moderate or severe AS can develop symptoms de novo after pregnancy, extended postpartum follow up is important.

References:

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