HEART DISEASE & PREGNANCY

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AORTIC COARCTATION

Background

Coarctation of the aorta is defined as a stenosis located usually at the junction of distal aortic arch and proximal descending aorta (almost invariably at the insertion of the ligamentum arteriosum). A significant aortic coarctation usually presents with right arm hypertension and a peak pull back gradient of more than 20 mmHg across the coarctation site at catheterization. However, if there is extensive collateral circulation, there may be minimal or no pressure gradient in spite of a significant aortic coarctation

Associated cardiovascular abnormalities include: bicuspid aortic valve disease, intracranial aneurysms of the circle of Willis (Berry aneurysms) and aortic medial disease in the para-coarctation aorta or in the ascending aorta associated with a bicuspid aortic valve.

Adults with unrepaired coarctation usually present with systemic arterial hypertension or a systolic murmur. Adult patients with coarctation are often asymptomatic. A significant proportion of adults with repaired coarctation have persistent hypertension. Typical symptoms attributed to upper body hypertension include headache, nose bleeding, dizziness, tinnitus, and exertional leg fatigue.

Most women of childbearing age have had coarctation repair, either surgically or by balloon angioplasty with or without stent. Late complications after coarctation repair include residual hypertension, re-coarctation, aneurysm at the site of the repair, premature coronary artery disease, ventricular dysfunction or complications related to bicuspid aortic valve disease. (see Aortic Stenosis)

Effects of pregnancy-related hemodynamic changes

The normal increase in blood volume and cardiac output during pregnancy can contribute to hypertension and increased shear stress on the aortic wall. (see Cardiovascular Changes During Pregnancy) Weakening of the aortic wall may also be mediated by hormonal changes. Placental blood flow is reduced in cases of significant coarctation.

Maternal complications

The most common pregnancy associated risk relates to systemic hypertension. (1,2,3,4) Hypertension can have health consequences for the mother, especially one with aortic involvement, and the fetus. Hypertension is more common in women with unrepaired coarctation or re-coarctation.

A less common, but more serious risk relates to the diseased aorta, which can dilate and dissect during pregnancy. Congestive heart failure and angina pectoris have been reported. (1,2,3,4) There are other cardiac characteristics, which can have an impact on outcomes. (see General Considerations) Maternal mortality has been reported and therefore careful risk stratification prior to pregnancy is important.

The most serious non-cardiac complication is cerebral hemorrhage from rupture of an aneurysm of the circle of Willis.

Women with coarctation and Turner syndrome have a much higher pregnancy risk and pregnancy is generally not advised in this group of women.

Fetal complications

Women with coarctation have a high risk for adverse fetal events. They are at risk for miscarriages. Prematurity is reported in 9% of pregnancies. Fetal deaths, although rare, have been reported. (4)

Management strategies

Preconception counseling/Contraceptive methods

Successful pregnancies are reported in women with coarctation; however, preconception risk stratification is important. The severity of the coarctation gradient, the size of the aorta, the presence of an aneurysm at the repair site, the presence of hypertension and the severity of bicuspid aortic valve disease, if present, are important issues to consider when determining an individual's risk. There are also other cardiac characteristics, which can have an impact on outcomes (see General Considerations).

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. All women should have cardiac magnetic resonance imaging to assess the entire aorta including the repair site. Catheterization may be indicated for women requiring surgery prior to pregnancy or if there are other unaddressed hemodynamic issues.

Repair of significant coarctation should occur prior to pregnancy. Pregnancy should be postponed in women in whom blood pressure is not well controlled.

Transmission of congenital heart disease to offspring should be discussed. The risk of transmission of congenital heart disease is approximately 5-10%, compared to a background risk of approximately 1%.

A discussion about contraceptive methods is appropriate in all women with coactation. Combined oral contraceptives containing estrogen/progestin should be used with caution in women with repaired coarctation with dilated aortas, patch aneurysms or hypertension. (see Contraception)

Women treated with angiotensin converting enzyme inhibitors or angiotensin receptor blockers need to have these medications stopped prior to pregnancy. 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 clinical and echocardiographic assessments during pregnancy should be determined on an individual basis.

Women with minimal coarctation gradients, normal aortic root sizes and no significant associated bicuspid aortic valve disease have the best chance for an uncomplicated pregnancy.

Treatment of hypertension is important during pregnancy. However, antihypertensive medications can exacerbate hypotension distal to the coarctation site and result in diminished placental perfusion. Optimal blood pressure targets are not defined in this population. Because of their potential role in protecting the diseased aorta, beta blockers should be considered first line agents in women with aortic dilatation.

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

Labour and delivery

Labour and delivery should be planned 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 and help to control blood pressure. To decrease maternal expulsive efforts during the second stage of labour, forceps or vacuum delivery is often utilized. Uterine contractions are utilized to facilitate the initial descent of the presenting part in order to decrease potential harmful complications from difficult mid cavity-assisted delivery,

The need for maternal monitoring at the time of labour and delivery is dictated the severity of the coarctation, the degree of ventricular dysfunction, and the existence of associated lesions. Invasive blood pressure monitoring may be required for women with poorly controlled blood pressure.

In general, endocarditis prophylaxis at the time of labour and delivery is not recommended in women with coarctation. 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.

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.

References:

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