Aortic valve diseases in pregnancy

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It is not uncommon to encounter pregnant women with aortic valve diseases (AVD). Rheumatic AVD are prevalent in the emerging countries, while congenital aetiology is most common in the developed countries. Pregnant women with significant AVD should be assessed and followed up by the Pregnancy Heart Team. Surgical correction of symptomatic severe AVD is mandatory before conception. Some AVD are associated with aortopathy (e.g., Marfan, bicuspid aortic valve). Those patients carry a higher risk during pregnancy/delivery and their follow-up echocardiography should focus on the progression of the diameters of the ascending aorta.

Topic(s):
Valvular Heart Disease

Introduction

Aortic valve diseases (AVD) are often associated with a higher maternal and foetal risk. That is why patients with AVD need pre-conception counselling to risk stratify their condition to ensure a safe and eventless pregnancy.

The physiological haemodynamic changes during pregnancy may not be well tolerated in patients with significant AVD. Pre-conception surgical correction is indicated in those patients before contemplating pregnancy.

Given the maternal and foetal risks of AVD and the complex management, a collaborative work between cardiologists, obstetricians, cardiothoracic surgeons and anaesthesiologists (the Pregnancy Heart Team) is required before, as well as, throughout pregnancy and during delivery [1,2].

Aetiology of AVD

Rheumatic

This is the main aetiology of AVD in the emerging countries, where rheumatic fever is endemic. In developed countries, the incidence of rheumatic heart diseases is declining but the disease remains prevalent in the immigrant population [3].

Infective endocarditis

AVD may develop as a result of complicated infective endocarditis. Infected vegetations may cause perforation of the aortic cusps and acute severe aortic regurgitation (AR) which is poorly tolerated during pregnancy.

Congenital

With advances in surgery, an increasing number of women with congenital AVD reach childbearing age and desire to become pregnant [4].

Bicuspid aortic valve is considered the most prevalent aetiology of AVD in the developed countries. In bicuspid AVD, the aortic valve has only two cusps instead of three. Because of the turbulent flow across the two cusps, the valve may be functionally abnormal, and this may manifest itself as aortic stenosis and/or aortic regurgitation [5]. The bicuspid AV is usually associated with an abnormal aortic wall structure (aortopathy) and dilation of the ascending aorta is likely to coexist. Sometimes, the bicuspid AV is associated with heritable thoracic aortic diseases (HTAD) which is a group of thoracic aortic diseases caused by mutation of certain genes (e.g., Marfan syndrome) [6].

Degenerative

Degenerative AVD is a disease of the elderly, thus it is unlikely to be encountered during pregnancy.

Pathophysiology

Pre-conception

Apparently, only those with a known heart disease can have access to pre-conception care [7].

Usually mild and moderate AVD are well tolerated during pregnancy.

A pre-conception exercise test is needed for asymptomatic patients with severe aortic stenosis (AS) to assess the exercise tolerance, the blood pressure response and the occurrence of arrhythmias. If the test is normal, pregnancy is usually well tolerated [6]. An abnormal exercise test is defined as the following exercise-induced changes: development of symptoms, abnormal ECG, failure of improvement of left ventricular ejection fraction, failure of blood pressure rise or blood pressure drop during exercise, exercise-induced rise in the mean aortic gradient >20 mmHg or exercise-induced systolic pulmonary hypertension >60 mmHg [7].

Patients with aortopathy will need pre-conception complete aortic imaging by computed tomography (CT) scanning or magnetic resonance imaging (MRI) for appropriate pre-conception counselling. Sometimes, genetic counselling may be requested, especially in patients with Marfan syndrome and other HTAD [8]. These diseases are rare during pregnancy but associated with very high mortality [9].

Patients who have AVD who, according to guideline recommendations, are indicated for surgical correction should undergo surgery before becoming pregnant [8].

Contraindications to pregnancy

The following conditions are contraindications to pregnancy (mWHO-IV) because of the very high risk of maternal morbidity and mortality. If pregnancy is encountered, it should be terminated, and further pregnancies are discouraged until surgical correction of the lesions [8].

1. AS: severe symptomatic AS or severe asymptomatic AS when associated with either impaired left ventricular systolic function (EF <50%) or a pathological exercise stress test.
2. Isolated bicuspid AV when the ascending aorta is dilated >50 mm.
3. Bicuspid aortic valve with Marfan and other HTAD when the ascending aorta is dilated >45 mm.

During pregnancy

The plasma volume and the heart rate increase, while the systemic vascular resistance decreases throughout the first trimester and plateau about halfway through pregnancy [10]. The increased stroke volume and cardiac output may cause decompensation of a previously stable significant aortic stenosis. Mild and moderate AS lesions are usually well tolerated during pregnancy unless they are associated with left ventricular dysfunction or other comorbidities. Pregnant women with severe AS are more likely to develop heart failure and atrial arrhythmias [11].

On the other hand, AR is better tolerated during pregnancy than AS because the reduction in the systemic vascular resistance leads to a reduction of the regurgitant volume.

There are no studies to compare pregnancy outcomes in women with isolated mitral versus isolated aortic valve diseases. However, based on the WHO risk stratification, isolated severe asymptomatic AS is considered mWHO-III, while severe MS, symptomatic or not, is classified as mWHO-IV [8]. This means that, unlike MS, severe AS in a previously asymptomatic patient is often well tolerated during pregnancy [7].

It is worth mentioning that the risks of valvular lesions during pregnancy are additive, so impaired ventricular function or the presence of additional valve lesions will increase the risks [7].

Delivery

With uterine contractions, an extra amount of blood returns to the circulation (autotransfusion) and causes an extra burden on the heart. After delivery, the pressure of the gravid uterus over the inferior vena cava is released, leading to an increase in the venous return and further increase in the preload and cardiac output [10]. These changes are poorly tolerated in patients with severe AS.

Management

Diagnosis

Echocardiography is recommended once per trimester. Because pregnancy may be associated with increased aortic diameter [12], echocardiography is recommended in patients with aortopathy, where more frequent serial measurement of the ascending aortic dimensions is of utmost importance. Measurement of the aortic dimensions should be indexed to the body surface area [8].

Treatment

Medical

If heart failure develops during pregnancy, restriction of physical activity and a diuretic therapy should be prescribed, but care should be taken to avoid over-diuresis, especially in patients with severe aortic stenosis because it is a preload-dependent lesion [8].

In case of rapid arrhythmias, nodal blockade by beta-blockers may be needed.

Percutaneous

In severely symptomatic AS patients who are at an advanced stage of pregnancy, a trial of balloon dilatation of the aortic valve may be considered [11]. Transcatheter aortic valve implantation is a promising alternative, but evidence during pregnancy is lacking.

Surgical

Pre-conception surgical correction of severe AV lesions is indicated, according to the guidelines' recommendations. Surgery may be considered during pregnancy or immediately after delivery if the patient has severe symptoms that are resistant to medical treatment [13]. The best period for surgery is between the 13th and 28th weeks of gestation [8].

Mode of delivery

Caesarean section is indicated in patients with severe aortic stenosis, dilated ascending aorta (>45 mm), (history of) aortic dissection or intractable heart failure. Otherwise, spontaneous vaginal delivery is preferred [8].

Prognosis

Maternal risk depends on the severity of AS, the presence of symptoms and aortic root diameter [11]. Maternal risks include heart failure (<10% in moderate AS and 25% in severe AS), aortic dissection (<1% in bicuspid AV with aortic root diameter <50 mm, 1-10% in Marfan syndrome and other HTAD), and rarely arrhythmias [14]. Foetal risks include low birth weight, intrauterine growth retardation and preterm delivery. The tighter the valve, the higher the foetal risks [11].

Heart failure is common (20-25%) in patients with severe AR, especially when the LV systolic function is impaired [15]. The prognosis here depends on the regurgitation severity and its effects on symptoms, left ventricular size, and function [8].

Conclusion

Aortic valve diseases carry a considerable risk during pregnancy and delivery. Pre-conception counselling is required to assess the functionality of the aortic valve and the probable need for pre-conception surgical correction of the AV lesions. Mild and moderate AS and aortic regurgitation are generally better tolerated during pregnancy than severe AS. Special care should be given to patients with bicuspid aortic valve, specifically when associated with aortopathy.

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The author has no conflicts of interest to declare.