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Aortopathy in pregnancy

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ABSTRACT

Up to half of all aortic dissections and ruptures in women younger than 40 years are associated with pregnancy. In pregnancy, women with aortic disease such as arteritis and aortitis are at significant risk of aneurysmal formation and dissection with potential for catastrophic outcomes. Pregnancy places predisposed women at an increased risk of dissection due to physiological and hormonal changes that occur, particularly those with connective tissue disorders, genetic syndromes, congenital heart disease, and other heritable and acquired conditions involving the aorta. Thus, preconception counseling and preparation are advised to determine which patients may cautiously pursue pregnancy, to optimize medical management prior to conception (antihypertensive medications and anticoagulants in the setting of mechanical valves), to identify women in whom aortic root repair should occur prior to pregnancy, and lastly, those in whom pregnancy is contraindicated. Additionally, discussion of the heritable nature of many aortic conditions and associated syndromes is indicated. Preconception and genetic counseling, management by a multidisciplinary team, along with close echocardiographic surveillance and medical management, are recommended if precursors of dissection are identified.

Introduction

An obstetrician may encounter aortic pathology prior to conception, incidentally during pregnancy, when aortic pathology is identified at echocardiography performed for other indications, such as chronic hypertension, or emergently, during evaluation of acute chest pain in pregnancy.

The aorta should be assessed in patients with a personal or family history of aortic aneurysms, aortic dissection, connective tissue disorders, congenital heart disease, past cardiac surgery, and trauma. When an abnormality of the aorta is detected, an underlying genetic syndrome or congenital heart disease will often be present. The added demands of pregnancy predispose such patients to aortic dissection or rupture. The etiology of thoracic aortic dissection is believed to be related to increased wall stress and/or intrinsic abnormalities of the aortic media (Table 1).[1]

Natural history of aortic pathology

As most thoracic aortic aneurysms are asymptomatic, the patient may not be aware of its presence until a complication arises, such as rupture or dissection. An aneurysm is a pathologic dilation of the three vessel wall layers resulting in a 50% increase of the aortic diameter compared to unaffected individuals of the same age and gender.[1,2] One-fourth of aortic aneurysms are thoracic, most commonly involving the aortic root and/or ascending aorta (60%), followed by the descending aorta (40%), arch (10%), and thoracoabdominal portion (10%). Thoracic aortic aneurysms may

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Table 1 – Conditions associated with aortic dissection.[1]

Increased aortic wall stress
Hypertension
Pheochromocytoma
Stimulant drugs (cocaine)
Weight lifting (increased Valsalva)
Trauma
Torsional and deceleration injury
Coarctation
Abnormal aortic media
Genetic conditions
Marfan syndrome
Vascular Ehlers–Danlos syndrome
Turner syndrome
Loeys–Dietz syndrome
Bicuspid aortic valve
Nonsyndromic familial thoracic aortic aneurysm and dissection
Annuloaortic ectasia
Inflammatory vasculitides
Takayasu arteritis
Giant cell arteritis
Behcet arteritis
Pregnancy
Polycystic kidney disease
Chronic corticosteroid use
Immunosuppressive agent use
Infections
Syphilis
Tuberculosis
Pneumonia
Pericarditis
Osteomyelitis
Sepsis

dissect, compress, or erode into adjacent structures, leading to thromboembolism, leak, or rupture. In pregnancy, most dissections occur in the ascending aorta, though dissection or rupture of any affected artery in the body can occur.[3]

Profound physiological hemodynamic changes occur in pregnancy, which continue to increase throughout gestation. These include marked increases in blood volume, heart rate, stroke volume, cardiac output, left ventricular wall mass, and end-diastolic dimensions.[4–6] In addition, estrogen and progesterone induce histological structural changes of the aortic walls early in pregnancy that remodels the tunica media and intima. This results in risk of dissection due to fragmentation of the reticulum fibers, diminished amount of acid mucopolysaccharides, and loss of the corrugation of elastic fibers in the vessel.[7,8] These changes begin early in gestation and are most pronounced in the third trimester and peripartum period.

Women at high risk of aortic dissection will typically have an aortic root diameter $\geq$4 cm, rapid dilation of aortic dimension, or previous dissection of the ascending aorta (particularly those with connective tissue disorders and hereditary aortopathy). However, aortic dissection may occur in women with no underlying known risk factors. Thus, aortic dissection should be in the differential of chest pain in every pregnant woman.[9,10]

Ascending thoracic aorta aneurysms most often result from cystic medial degeneration. This is a process of smooth muscle cell necrosis and degeneration of elastic layers within the media that leads to circumferential weakness and dilatation. Cystic medial necrosis may normally occur with aging, being accelerated by hypertension, or with congenital connective tissue disorders. A condition commonly associated with connective tissue disorders is annuloaortic ectasia, a widening of the proximal aorta and aortic valve. Notably, Marfan syndrome accounts for 50% of annuloaortic ectasia cases. However, cystic medial necrosis and its complications can occur in young patients even in the absence of congenital connective tissue disorders.[11,12]

In pregnancy, treatment plans will depend on the etiology of aortopathy, aortic dimension, and comorbid conditions. The goal is to prevent life-threatening aortic dissection, which is associated with high maternal and fetal mortality. Up to 50% of dissections occur in the last trimester of pregnancy or in the early postpartum period (33%).[12] The average growth rate of thoracic aneurysms is 1–4 mm per year, with larger aneurysms growing more rapidly. Median diameter at aneurysm rupture is 6 cm for ascending aneurysms and 7 cm for descending aneurysms. Rupture of smaller aneurysms may occur in patients with Marfan syndrome.[3] Most thoracic aortic aneurysms are asymptomatic; however, symptoms of aortic enlargement may arise due to pressure effects on neighboring structures and hemodynamic changes. Chest pain, shortness of breath, cough, hoarseness, or dysphagia may be caused by compression or erosion of adjacent tissue by aneurysmal dilation. Aneurysms of the ascending aorta may cause congestive heart failure (consequence of aortic regurgitation); compression of the superior vena cava may produce congestion of the head, neck, and upper extremities. Survival rate of patients with untreated large thoracic aortic aneurysms is 65% at 1 year and 20% at 5 years.[3] In the event of dissection, surgical repair is required along with immediate control of hypertension.

General considerations

Prior to conception

The importance of preconception counseling in all women with aortic disease cannot be overemphasized. A woman planning her pregnancy should be informed of the impact pregnancy will have on the cardiovascular system and how these changes may affect their underlying condition. The increased risk of dissection in pregnancy should be explained as well as the intense multidisciplinary medical surveillance required to monitor aneurysmal dilation (Table 2).

Heritable disorders associated with aortic disease and dissection include Marfan syndrome, Ehlers–Danlos syndrome, bicuspid aortic valve, familial forms of aortic dissection, aneurysm, or annuloaortic ectasia. Proper family planning is recommended for patients with an underlying aortic condition or genetic syndrome. The adolescent female should be informed of her diagnosis when maturity is considered appropriate, and effective contraception should be prescribed.[13–16]

Knowledge of pregnancy associated maternal risks and the heritable nature of aortic genetic condition or syndrome may