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Aorta pathology and pregnancy

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In addition to the haemodynamic changes in pregnancy, hormones also induce changes in the aortic wall. Women with diseases like Marfan syndrome, Ehlers–Danlo syndrome, or other aortic abnormalities, have an increased risk of complications during pregnancy. Counselling and risk assessment before pregnancy is mandatory for all women with known aortic disease. Proper information should be provided about the risks of morbidity and mortality during pregnancy and information on the risks for the fetus, including the potential recurrence of disease in the offspring. Evaluation of past medical and family history, the aortic size before conception, and any increase in size before and during pregnancy, is essential to try and estimate the risk of aortic dissection. If the aorta is dilated, prophylactic repair before pregnancy may be indicated. In some cases, elective surgery during pregnancy may be warranted. In women with a severely dilated ascending aorta, caesarean section is, at present, the advised mode of delivery.

Introduction

In young women, the occurrence of an aortic complication is fortunately rare, with a reported incidence in the general female population under 45 years of age of 0.4 per 100,000 person years.[1] When acute aortic dissection (AOD) occurs in a person under the age of 40 years, it is frequently caused by an underlying disorder such as Marfan syndrome or a bicuspid aortic valve. In this young age group, aortic dissection has a mortality rate of 22%, which is similar to the mortality rate in people aged over 40 years.[2]

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In pregnancy, haemodynamic adaptations influence the vascular system, including the aorta. The major cause of maternal mortality in the UK between 2006 and 2008 was cardiac disease, with 2.31 deaths per 100,000 maternities. Although the incidence of aortic dissection is low, it was one of the most important causes of cardiac death.[3] A challenging situation exists for women at risk of aortic complications who contemplate pregnancy. Moreover, some women may have AOD during pregnancy as the first presentation of their aortic disease. Women with known aortopathy reaching reproductive age who are at high risk should be counselled about the risks of pregnancy. They should be referred to a tertiary centre for the purpose of surveillance during pregnancy and management of delivery.

General recommendations and guidelines

Taskforces have made great efforts to provide recommendations on this important topic.[4,5] The two most recent provide helpful advice on how to manage women with aortopathy, but prospective studies are needed to assess and provide better data to support the recommendations. The Registry of Pregnancy And Cardiac diseases is currently the largest worldwide prospective study on pregnancy in women with cardiac disease, and includes women with aortopathy. The registry is still ongoing.[6]

Effect of pregnancy on the aorta

The aorta is subject to structural and functional changes throughout pregnancy. Cardiac output, heart rate, and circulating volume increase, causing the aortic size to increase. The aortic size is also related to haemodynamic conditions, and varies in dimension with conditions such as preeclampsia.[7] The hormonal changes in pregnancy influence connective tissue, changing the microstructure of the medial layer. Analysis of histopathology of the tunica media reveals hypertrophy and hyperplasia of smooth-muscle cells and fragmentation of reticulin, which normally surrounds elastic fibres.[8] Subsequently, elastic fibres lose their organised structure, changes that are most notable in late pregnancy.

The above effects of pregnancy on the aorta explain why pregnancy itself is thought to be a risk factor for acute aortic dissection, although this topic is subject to debate, and contradictory reports exist. A population-based study in Sweden reported that pregnancy-related AOD is responsible for 60% of all AOD in young women, with a 25-fold increased risk of dissection during pregnancy[9]; however, another population-based report[1] in Austria reported no increase in risk of aortic dissection. In these population-based studies, the difference in reported outcomes and risk of AOD might be related to different observation periods (1987–2007 compared with 1994–2004), and the small absolute total numbers of dissections (29 compared with 15). It is, however, clear that women with an increased risk of aortic dissection are those with connective tissue diseases such as Marfan syndrome, Ehlers–Danlos syndrome, and other genetic conditions pre-disposed to developing thoracic aneurysms and aortopathy. Aortic abnormalities in the context of bicuspid aortic valves will be reviewed elsewhere.

Pre-conceptional counselling

All women at increased risk of aortic complications, including those with known Marfan syndrome, Ehlers–Danlos syndrome, Loeys–Dietz syndrome, or familial thoracic aorta dissection syndrome, should be counselled before pregnancy. Preferably, counselling should be carried out once a girl reaches reproductive age. Maternal and fetal risks should be discussed, as well as recurrence risk of the disease in the offspring and of the possible effect of medication use. It is also important that women are made aware of the value of computed tomography and magnetic resonance imaging (MRI) of the aorta before pregnancy, because the results, if abnormal, may mean that pregnancy is inadvisable or elective aortic surgery is warranted.[4,5] The risk of aortic dissection is most likely a result of a combination of pregnancy effects, pre-existence of an abnormal aortic wall, and an increased aortic size at baseline. Hypertension is a significant risk factor in a healthy woman affected