

Aortic dissection during pregnancy — obstetric perspective

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ABSTRACT

The risk of vascular events during pregnancy is substantially increased. Beyond comparatively frequent vascular diseases, pregnancy may lead also to the development of exceptionally rare vascular events such as the aortic dissection and aortic rupture which are conceivably endangering life conditions. Women with the connective tissue disorders and with a family history of the aorta diseases are especially prone to the aortic complications which may also develop in the absence of these risk factors due to the pregnancy-induced structural changes of the aortic wall. The preconception counselling is vital for patients with aortopathies to assess the risk of the aortic dissection and to establish the most appropriate care plan including the surgical intervention. This review presents the management guidelines in patients with the aortic dissection risk during pregnancy.

Key words: aortopathy; aortic dissection; Marfan syndrome; pregnancy

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INTRODUCTION

The aortic dissection (AoD) during pregnancy is an extremely rare condition with the incidence of 14.5/1 000 000 in pregnant women vs 1.24/1 000 000 in non-pregnant ones [1]. The maternal mortality due to AoD is assessed for up to 30% and the fetal mortality — up to 50% [2]. The reported mortality is estimated at 1% per hour during the first 48 h and exceeds 80% in the first month following the event [3].

RISK FACTORS FOR AORTIC DISSECTION

The aortic dissection should be expected in patients with diagnosed connective tissue disorders: mainly Marfan syndrome and the vascular type of Ehlers-Danlos syndrome, Loeys-Dietz, and Turner syndrome, and congenital malformations of the aorta like a bicuspid aortic valve. However, this catastrophic complication may appear in previously healthy pregnant patients [2]. In the report published by Yuan describing 122 pregnant patients with AoD, Marfan syndrome (49.25%) and pregnancy itself (25.4%) were the leading reasons responsible for AoD. The Marfan patients developed AoD earlier during pregnancy: they were diagnosed with AoD during all trimesters, mainly in the last trimester, whereas AoD occurred in healthy pregnant women only in the 3rd trimester [4]. This data confirms the

earliest observations that almost 50% of the cases of AoD arise in the last trimester or in the early post-partum period (33%) [5]. Kamel et al. have revealed that most of AoD events are observed between 6 months before and 3 months after delivery. The patients with the family history of aortic aneurysms, AoD, congenital heart disease, past cardiac surgery, and trauma are at higher risk of AoD during pregnancy [2]. AoD or the rapid dilation of the aortic dimensions has been proven to increase the likelihood of aortic disruption in pregnant women as well. It has been assessed that women with the aortic root diameter below 40 mm have the risk of either the aortic dissection or other significant cardiac events to be 1 percent [6]. Table 1 presents the risk factors for AoD [7]. According to the Stanford classification, there are two types of AoD: type A which contributes to 79–89% of AoD cases in pregnant women and involves the ascending aorta with or without its descending part, and type B dissection (the descending part of the aorta only) which is associated with 11–21% of events [8].

PREGNANCY-INDUCED CHANGES WITHIN THE VASCULAR WALL

Pregnancy is the cause of multiple changes within the circulatory system such as the increase in blood volume,

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Table 1. The risk factors for the aortic dissection [7]

Increased aortic wall stress	Hypertension, preeclampsia, pheochromocytoma, stimulant agents (cocaine), trauma, torsional and deceleration injury, coarctation
Genetic conditions	Marfan syndrome, vascular Ehlers-Danlos, Turner, and Loeys-Dietz syndrome, bicuspid aortic valve, nonsyndromic familial thoracic aortic aneurysm and dissection, annuloaortic ectasia
Inflammatory vasculitis	Takayasu arteritis, Giant cell arteritis, Behcet arteritis
Others	Pregnancy, polycystic kidney disease, chronic corticosteroid, and immunosuppressive agents use
Infections	Syphilis, tuberculosis, pneumonia, pericarditis, osteomyelitis, sepsis

cardiac output and left ventricular dimensions, heart rate, all of them require the aortic wall remodeling [9]. The aortic diameter and compliance increase throughout pregnancy, considerably during the third trimester and the post-partum period. This effect is more enhanced in hypertensive and preeclamptic women [10]. The structural remodeling changes of the aortic wall observed within the tunica media and intima and induced by estrogen and progesterone as well as by circulating angiogenic factors, start early in pregnancy. They have been suggested as relevant to the aorta degeneration including the hyperplasia of aortic smooth muscle, the decrease in the amount of acid mucopolysaccharides, and the disintegration of elastic fibers [10]. These hormonal-induced changes may result in the significant impairment of the vascular wall, thereby being a relevant reason for the AoD development during gestation [8].

PRECONCEPTION COUNSELLING

The preconception counselling is vital for patients with aortopathies to assess the risk of AoD and to establish the most appropriate care plan. Young women with the family history of AoD or diagnosed connective tissue disorders and congenital aortic abnormality should be proposed the diagnostic evaluation prior conception. The preconception counselling is mandatory for patients with the enlarged aortic root in order to prevent AoD in pregnancy [11].

The assessment of the entire aorta with preferably magnetic resonance imaging (MRI) should be obligatory for patients considering pregnancy with the AoD risk factors in order to perform the elective root replacement if necessary [12, 13].

Although the aortic root of more than 4 cm in diameter and/or fast increase in the aorta size or AoD of ascending aorta in the past, mostly in the patients with connective tissue disorders are the more frequent risk factors for AoD, the aortic dissection may develop in pregnant women without

evident risk factors. Hence, AoD should be taken into consideration while diagnosing chest pain in every pregnant woman [14].

In patients with the high risk of AoD or the aortic rupture during pregnancy, the need for the surgical intervention should be considered prior to the conception. The diameter of the aorta when the scheduled surgical intervention should be offered depends on concomitant genetic diseases, a pace of the aorta diameter increase, and symptoms. Due to the extremely high risk of death exceeding 10%, pregnancy should be avoided by women with Marfan syndrome with the dilated aortic root ≥ 4 cm, when the rapid growth of the aorta dimension (> 0.5 cm per year) is observed in patients with bicuspid aortic valve and the dilation of the aorta ≥ 5 cm and by women with previous AoD type A. The surgical repair of the aorta should be performed even before considering conception in these cases [15, 16].

The safe diameter of the aorta in Marfan syndrome below which the dissection will not develop in pregnancy has not been established, although AoD is uncommon when the aortic diameter is below 4 cm. In general, the patients with the prior aortic dissection should be discouraged from pregnancy. Women with the dilated aortic root ≥ 4.0 – 4.5 cm, with concomitant connective tissue diseases, the family history of AoD, or the indications for the surgical aortic valve replacement, should be offered the surgery before pregnancy planning [15]. Due to their teratogenicity, ACE inhibitors (angiotensin converting enzyme inhibitors) and ARBs (angiotensin receptor blockers) should be discontinued in favor of β -blockers prior conception.

The genetic assessment should be ensured for the patients with the possible genetic diseases of the connective tissue [17].

PREGNANCY MANAGEMENT

The patients with risk factors for AoD during pregnancy abovementioned require the constant and close surveillance of the multidisciplinary medical team consisting of a specialist in perinatal and neonatal medicine, cardiologist, cardiothoracic surgeon and anesthesiologist in tertiary center of perinatal care [4]. The management options in gestation depend on the causes of the aortopathy, aortic dimension and concomitant diseases. The fundamental goal is the prevention of AoD and the aorta rupture which are life-threatening conditions with the maternal blood pressure and heart rate control in order to reduce their negative influence on the aortic wall. Beta-blockers, first of all, metoprolol and labetalol which possesses the activity of both β - and α -adrenergic receptors blocker, are commonly safe and efficient agents during pregnancy and should be administered to alleviate aortic wall stress and to decrease the risk of the aorta enlargement. Atenolol should be avoided due to its association with the fetal growth re-

striction [5]. The doses of β -blockers should be adjusted to gain the maximum effect, optimally the resting heart rate of < 60 bpm is desirable and needs to be maintained if well tolerated by the patient [18]. The rigorous blood pressure control, with the systolic blood pressure < 140 mmHg and the diastolic < 90 mmHg is advised. Patients with chronic hypertension and preeclampsia need special attention due to the increased risk of AoD during gestation [16]. Since the high risk for aortic complications decreases gradually after delivery, the therapy should be prolonged until the completed 3rd post-partum month [9]. If β -blockers are contraindicated, calcium channel blockers may be administered [7].

During pregnancy, the regular assessment of the aortic diameter with the echocardiographic measurement is mandatory every 4 to 12 weeks till the 6th month post-partum. MRI without gadolinium contrast is recommended for the pregnant women with the aorta anomalies such as the dilatation of the aortic arch, descending aorta, and/or abdominal aorta [19]. This diagnostic tool is favored to the computed tomography (CT) in order to avoid fetal exposure to the ionizing radiation. The initial MRI assessment of the entire aorta with subsequent regular echocardiographic monitoring in women with the aorta abnormalities who have never had pre-pregnancy examination is urged. However, CT due to its availability, particularly in an urgent situation, is often used and may be performed in pregnant patients in justified cases [7].

In addition to the routine fetal ultrasound examinations, the fetal echocardiography should be offered for the prenatal diagnosis of congenital abnormalities related to the connective tissue disorders [15].

In the case of diagnosed or suspected genetic connective tissue disease in parents such as Marfan syndrome, Loeys-Dietz, and vascular Ehlers-Danlos syndrome, which are characterized by an autosomal dominant inheritance with the 50% risk of the transmission on offspring, the invasive genetic prenatal testing should be offered (chorionic villus sampling or amniocentesis) [12].

The management of women of childbearing age with aorta diseases is presented in Table 2 [7].

The clinical symptoms of AoD are unspecific — it should be considered in patients reporting the acute chest pain (70%), back pain (21.7%) or epigastric and abdominal pain (2.5%) and/or with dyspnea, persistent cough, syncope and circulatory collapse [4]. The neurological injury or the pericardial tamponade may also be present and usually manifest as syncope. The typical thoracic pain is rarely manifested in patients with dissection-related neurological events [20]. In order to confirm the diagnosis, the immediate MRI or CT should be carried out. The prompt surgical treatment for AoD is mandatory. The urgent surgery of AoD during pregnancy results in high maternal mortality and morbidity. One month after the surgical intervention, the reported

Table 2. The management of women of childbearing age with aorta diseases [7]

Before pregnancy	Maternal risk assessment — echocardiography — cardiac MRI Surgical procedure if necessary Genetic testing Management of concomitant diseases: diabetes, hypertension, obesity
Pregnancy	Management in a tertiary center of perinatal care by the team of specialists in perinatal medicine, cardiology and cardiothoracic surgery, anesthesiology, neonatology Echocardiography every 4–6 weeks Hypotensive treatment — β -blockers Genetic testing if indicated Fetal echocardiography (18–20 th week) Regular fetal growth and wellbeing assessment Treatment of comorbidities: diabetes, anemia, etc.
Delivery	Cesarean section — aortic diameter ≥ 4.0 cm: Marfan syndrome, AoD, severe aortic regurgitation, heart failure — aortic diameter ≥ 4.5 cm: other thoracic aortic aneurysms Vaginal delivery with the assisted second stage — aortic diameter < 4.0 cm — regional/epidural anesthesia

mortality rate is as high as 40–50%; the survivors suffer from significant complications such as renal and respiratory compromise, and severe neurologic impairment [21]. The possible surgical options in the case of AoD diagnosed during pregnancy depend on the gestation age. If necessary, the surgical repair of AoD with the continuation of pregnancy might be considered below the 28th week of gestation. Between the 28th and 32nd week, the decision of the aortic repair with the cesarean delivery depends on the mother and fetus condition. After the 32nd week of gestation, the cesarean delivery with the consecutive aorta repair in the same surgical session offers the best outcomes [22]. The main obstetric issue for these cases is the high risk of the peripartur hemorrhage mainly due to the use of large quantities of anticoagulants required during the extracorporeal circulation. Haas et al. [23] recommend that the hysterectomy just after the cesarean section should be considered as a prevention of the massive bleeding caused by anticoagulants. The peripartur hysterectomy is the highest risk surgical procedure, including the risk of hemorrhage and should be considered only if additional indications like the uterine atony or placental pathology, exist. According to the data presented by Guo C. and al. [24], the Bentall procedure instead of repairing the arch with the deep hypothermia and the circulation arrest results in favorable outcomes. In the

large study on AoD in pregnancy, it has been reported that two types of procedures were performed the most often: the ascending aorta replacement (40%) and the Bentall operation (31.2%) with advantageous results [4].

The acute dissection of the descending aorta could be treated with vigorous hypotensive therapy unless severe, life-threatening complications occur [25].

DELIVERY

The decision on the mode of delivery for the women with the aortopathy should be based on the thorough analysis of the present maternal and fetal condition, medical history and should be made by the multidisciplinary team consisting of the obstetrician, cardiologist, anesthesiologist, and neonatologist. The recommendations on cesarean section and vaginal delivery for the patients with aortopathies are presented in Table 2 [7].

POST-PARTUM

The AoD risk for patients with aortopathies remains high in post-partum thus they require close surveillance and care with β -blockers treatment for 3 months after delivery [10].

SUMMARY

The pregnant patients with the aortopathy, mainly Marfan syndrome, should be assigned to the group of highest risk of AoD during pregnancy and the post-partum period. The preconception counselling is vital for the patients with aortopathies to assess the risk of AoD and to establish the most appropriate care plan: in patients with the high risk of AoD or the aortic rupture during pregnancy, the need for the surgical intervention should be considered prior to the conception. In the case of chest pain and dyspnea during pregnancy, the possibility of AoD should be strongly considered even regardless of other risk factors. For the pregnant patients with the known risk factors for AoD and for those presenting clinical symptoms of AoD, the close monitoring of pregnancy and the aorta condition should be offered in the tertiary perinatal care center where the cardiovascular procedures are immediately available.

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