Editorial

Pregnancy-related aortic dissection—recognize, mitigate, act

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Aortic dissection (AD) complicating pregnancy is rare, accounting for only a small fraction of all acute ADs (1). Although uncommon, the condition is deserving of attention, as pregnancy is recognized as a potential trigger in almost 20% of dissections occurring in women <35 years old (1), and may lead to significant morbidity and mortality in both the mother and the fetus (2). Pregnancy is an independent risk factor for both aortic growth and AD in women with aortopathy conditions (3), relating to hemodynamic and hormonal changes and their effects on the vasculature (1). Additional factors including hypertension and aortic dilation heighten this risk (3).

Women with heritable thoracic aortic disease (HTAD) including syndromic and non-syndromic conditions are at particular risk of pregnancy-related AD, which is highest during the third trimester and the early postpartum period, but with continued risk for up to several weeks postpartum (1,3,4). Ascending aortic size is an important predictor of dissection risk, although clinicians should be cognizant that type B (descending) AD may occur in the absence of aortic dilation and thus is often unpredictable (5).

How does one mitigate risk of acute AD in susceptible women? For women with Marfan syndrome (MFS), there are some data to guide management. The risk of AD is particularly high in women with MFS and an aortic root size >4.5 cm, reflected in the recent 2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease, which endorses a class I recommendation that these women undergo prophylactic aortic surgery before pregnancy (6). Conversely, women with MFS and aortic root diameter <4.0 cm can be reassured that pregnancy can be carried out and vaginal delivery safely undertaken with reasonably low risk of dissection (5). When the aortic diameter is 4.0 to 4.5 cm, shared decision-making is especially important, recognizing that while uncomplicated pregnancies are reported (4), type A (ascending) AD may occur (1).

Although AD has been widely reported in patients with other genetic aortopathies such as Loeys-Dietz Syndrome (LDS) (7), vascular Ehlers Danlos Syndrome (vEDS) (8) and non-syndromic HTAD (due to pathogenic variants in *ACTA2* and others) (1), there is a paucity of data to guide pregnancy decision-making. In these conditions, guideline recommendations are based on consensus expert opinion and are informed by the specific genetic variant, aortic diameter, family history and other factors—recognizing the uncertainties and emphasizing shared decision-making (6). Importantly, prior aortic root replacement for aneurysm or dissection does not mitigate the AD risk in the descending aorta (1), and individuals who have had prior aortic surgery still require close and careful supervision. Reassuringly, women with bicuspid aortic valve are not at risk for AD related to pregnancy, unless the aortic diameter is aneurysmal or associated with Turner syndrome (9,10).

There are many opportunities to recognize women at risk for pregnancy-related AD beginning with a careful physical exam and family history. Although most women who experience a pregnancy-related AD have an aortopathy condition or a family history of aortic disease (1,2), almost half of these women are not recognized to have the underlying at-risk condition until after the AD has occurred (1,4). Multiple health care providers have the opportunity to recognize the phenotypic features of a syndromic HTAD

or the presence of a concerning family history, which when identified can lead to the diagnosis of an aortopathy condition before a dissection occurs, and enable the timely referral to a medical geneticist, cardiologist and maternal-fetal specialist for counselling and cardiac and obstetric interventions that lessen AD risk.

In women recognized to have an aortopathy condition prior to pregnancy, aortic imaging is performed and prophylactic aortic surgery may be advised for women above certain aortic size thresholds (6). Beta-blocker and anti-hypertensive therapy to treat hypertension (as appropriate) is continued throughout pregnancy and postpartum, and close clinical and imaging surveillance within a multidisciplinary team of cardiologists and maternal-fetal medicine specialists can identify issues if they arise (6). Planning and implementing the method and timing of delivery, in a tertiary center with cardiothoracic surgical support, when appropriate, is a vital part of management (6). The risk period however does not end once the mother has delivered, and continued vigilance is required postpartum.

It can be challenging to distinguish the more common chest and abdominal symptoms of various etiologies that many women experience during pregnancy or postpartum from those due to an acute aortic syndrome. Not all dissections present with classic symptoms (6). When symptoms of concern occur, recognizing women at risk should heighten the clinician's index of suspicion for AD, and prompt rapid assessment and investigation. Women at risk should be educated of the symptoms of acute AD, and when present, pursue an action plan that includes presenting early, advocating for swift and appropriate evaluation, and notifying their providers.

After a pregnancy-related AD, further assessment of the mother with careful clinical evaluation and genetic testing may reveal a previously undiagnosed HTAD, with important implications for cascade testing in the child and other first-degree relatives (6). In the absence of a causative gene variant, first-degree relatives should undergo clinical assessment and aortic imaging, and women should receive counselling regarding risk of subsequent pregnancies.

Recognizing and counselling women at risk of AD are fundamental pillars of effective prevention and management of pregnancy-related AD and its potentially catastrophic sequelae. Improved awareness, multidisciplinary evaluation and management by a team including the medical geneticist, genetic counselor, cardiologist, maternal-fetal medicine specialist, and when appropriate, the aortic surgeon, provides the framework for optimal outcomes.

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Footnote

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