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Maternal and fetal outcomes of bicuspid aortic valve with early-onset complications

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Brief Report

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Tate Truly, Michael Huang, and Siddharth K. Prakash take responsibility for all aspects of the reliability and freedom from bias of the data presented and their discussed presentation.

Abstract

Bicuspid aortic valve is the most common congenital heart malformation and predisposes patients to thoracic aortic aneurysms and aortic dissections. Current peripartum guidelines are extrapolated from other heritable causes of thoracic aortic disease and do not account for unique characteristics of bicuspid aortic valve patients. We therefore evaluated the prevalence of maternal and fetal complications of women with early-onset complications of bicuspid aortic valve disease in the UTHealth Bicuspid Aortic Valve Research Registry. We found that the rate of cardiovascular complications was high and that relatively few women received guideline-recommended care.

Bicuspid aortic valve (BAV) is the most common adult congenital cardiac malformation, affecting 1–2% of the population. BAV predisposes to aortic valve disease, infective endocarditis, and thoracic aortic aneurysms with an increased lifetime risk for acute aortic dissections.[1] In the general population, adverse cardiovascular outcomes are increased in pregnancy due to physiologic changes including left ventricular dilation and increased cardiac output.[2] Pregnancy outcome studies involving BAV have focused on risks of peripartum aortic dissection, which is relatively low compared to other causes of heritable thoracic aortic disease.[3] Clinical guidelines recommend imaging of the aorta prior to pregnancy and at specified intervals during pregnancy depending on WHO risk stratification.[4] However, current guidelines make no significant distinctions between aortic disease subtypes in pregnancy.[2] We hypothesise that pregnancy may exacerbate peripartum cardiovascular risk in women with BAV and significant cardiac or vascular disease. We therefore analysed peripartum cardiovascular outcomes in a cohort of high-risk women who experienced early-onset complications of BAV.

Methods

There were 132 eligible participants with early-onset complications of BAV disease in the UTHealth BAV Research Registry.[5] Early-onset complications of BAV are defined as moderate or severe aortic stenosis or regurgitation, large thoracic aortic aneurysms (Z-score > 4), or valve or aortic interventions prior to the age of 30 years. Participants with syndromic causes of BAV such as Loeys–Dietz syndrome were excluded. They were invited to complete an online REDCap survey about their medical history, family history of congenital heart lesions, and pregnancy history, including maternal and fetal cardiovascular complications, treatments, and outcomes. In a subset of participants with available data, pre- and post-pregnancy images were analysed to assess changes in aortic dimensions during pregnancy.

Results

Sixty registry participants completed the online survey, and 44 (73%) reported at least one pregnancy. These patients constituted the study cohort (Table 1). The mean age of participants was 49 years (SD 14), and the mean body mass index was 28.5 kg/m^2 (SD 6). A total of 17 participants (39%) reported at least one cardiac or aortic surgery, including aortic valve replacement, aortic aneurysm repair, coarctation repair, or mitral valve surgery. More than one in four participants reported at least one relative who had been diagnosed with BAV, thoracic aortic aneurysm, or aortic dissection. In addition, five participants (11%) reported at least one other congenital lesion: mitral valve prolapse (4) or coarctation of the aorta (1).

There were 101 pregnancies in total: 15 participants reported a single pregnancy and 29 reported multiple pregnancies (Table 2). The mean age at first pregnancy was 25 (SD 5) years. Thirty-nine women (89%) reported at least one pregnancy with uneventful term delivery, which was the most frequent delivery outcome, for a total of 75 pregnancies. In the antepartum period, 11 pregnancies (11%) in 10 participants (23%) were affected by cardiovascular events, including newly diagnosed aortic valve disease (1), newly diagnosed thoracic aortic aneurysm (1), newly diagnosed congestive heart failure (1), hypertension requiring treatment (2), pre-eclampsia (5),

Table 1. Characteristics of the study cohort (N = 44)

Age (SD)	49 (14)
BMI (SD)	28.5 (6)
Race/ethnicity	
White/non-Hispanic	33 (75)
Medical history	
Hypertension	13 (30)
Diabetes mellitus	1 (2)
Tobacco use	12 (27)
Previous cardiac or aortic surgery	
Aortic aneurysm repair with aortic valve replacement	7 (16)
Aortic valve replacement	7 (16)
Aortic aneurysm repair	1 (2)
Coarctation repair	1 (2)
Mitral valve repair	1 (2)
Other congenital lesions	
Coarctation	1 (2)
Mitral valve prolapse	4 (9)
Family history	
Bicuspid aortic valve	13 (30)
Aortic aneurysm	8 (18)
Cerebral aneurysm	3 (7)
Myocardial infarction	15 (34)
Stroke or transient ischaemic attack	6 (14)
Ventricular septal defect	2 (5)

This table depicts the demographics, family history, and medical history of women with early-onset complications of BAV disease who reported at least one pregnancy. Unless otherwise specified, percentages are in parentheses.

Table 2. Maternal and fetal outcomes

Delivery outcomes (% of all pregnancies)	
Term	75 (74)
Term in hospital	4 (4)
Pre-term	1 (1)
Pre-term in hospital	4 (4)
Spontaneous abortion	13 (13)
Stillbirth	2 (2)
Elective abortion	2 (2)
Pregnancy cardiovascular complications (% of all participants)	
Development of new > mild valve disease	1 (2)
Aortic aneurysm	1 (2)
Hypertension	2 (5)
Heart failure	1 (2)
Preeclampsia	5 (11)
HELLP syndrome	1 (2)
Pregnancy non-cardiovascular complications (% of all participants)	
Placenta previa	1 (2)
Kidney disease	1 (2)
Cholestasis of pregnancy	1 (2)
Fetal cardiovascular outcomes	
Aortic valve abnormality	3 (3)
Aortic root dilation	2 (2)
Echocardiographic data	
Maximum aortic diameter mm/year (SD)	1.6 (1.9)
Sinuses of Valsalva diameter mm/year (SD)	1.2 (1.3)

This table depicts fetal delivery outcomes, maternal complications during pregnancy, and fetal congenital heart malformations in a cohort of women who experienced early complications of bicuspid aortic valve disease. Echocardiographic data are reported as the annualised mean change in aortic diameter.

and HELLP syndrome (1). There were also non-cardiovascular complications in three participants: placenta previa (1), cholestasis of pregnancy (1), and acute kidney disease (1). In the postpartum period, three participants had a total of five children who were diagnosed with a congenital heart malformation, including bicuspid aortic valve (2), thoracic aortic aneurysm (2), and aortic regurgitation (1). Only four (9%) participants received guideline recommended imaging prior to and during their pregnancies. In this subgroup, the normalised increase in maximum aortic diameter and sinus of Valsalva diameter was 1.6 mm/year (SD 1.9) and 1.2 mm/year (SD 1.3), respectively. Pre-existing aortic regurgitation increased from mild to moderate (1) or severe (1) in two participants.

Discussion

The primary objective of this study was to evaluate maternal and fetal outcomes of women with BAV who experienced early-onset vascular or aortic complications. We identified three key takeaways. First, relatively few women (9%) received guideline-recommended cardiac imaging surveillance during their pregnancies. Second, the prevalence of cardiovascular complications was high (23%). Hypertensive diseases of pregnancy were significantly increased in the study cohort (18%) compared to contemporary data (10%).[6] Hypertension and pre-eclampsia are the seventh leading cause of maternal mortality in the United States, accounting for 7% of pregnancy-related deaths.[7] The increased burden of hypertension in women with early-onset complications related to BAV is consistent with the severity of vascular disease in this cohort and corresponds to progressive aortic dilation during pregnancy. Image analysis confirmed that maximum aortic diameters increased relatively rapidly compared to longitudinal dilation rates of contemporary BAV cohorts.[8] These characteristics are consistent with a "complex valvulo-aortopathy" phenotype with more aggressive aortic disease, as may be anticipated for patients with early-onset complications of BAV.[9] Third, we found that maternal outcomes for women with BAV are generally favourable. There were no maternal deaths, and the prevalence of pre-term delivery (5%) or spontaneous abortion (13%) was not significantly increased in comparison to the general population.[10]

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Conclusions

We found that the risks of pre-term delivery (5%) and adverse fetal outcomes such as spontaneous abortion (13%) are not significantly increased in women with BAV compared to women in the general population. However, nearly one-fourth of women in this series developed new or worsening cardiovascular conditions during pregnancy. The prevalence of hypertensive diseases of pregnancy (18%) was also higher than reported rates in the United States (10%).[6] We conclude that the increased baseline risk for cardiac and aortic disease in women with BAV may be exacerbated by peripartum hypertension, emphasising the need for careful cardiovascular surveillance of women with BAV who are of reproductive age. For women with established vascular disease, coordinated peripartum cardiovascular care by a multispecialty team that includes a cardiologist, and a maternal–fetal medicine specialist can reduce the incidence of obstetric complications.[11]

Limitations

Early-onset complications due to BAV are relatively rare. Additional studies in larger groups may be needed to validate our observations. Other potential limitations include selection or recall bias in the survey data. Images were only available for a subset of cases, and this limited our ability to detect aortic changes during pregnancy.

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Conflicts of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant guidelines on human experimentation at the University of Texas Health Science Center at Houston and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the institutional committees at the University of Texas Health Science Center at Houston.

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