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Original Article

Bicuspid aortic valve in pregnancy

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ABSTRACT

Objective: The outcomes in pregnant patients with bicuspid aortic valves (BAVs) are rarely reported, despite the potentially critical nature of the condition. The aim of this study is to present the clinical complications of BAV in pregnancy.

Materials and methods: A MEDLINE database search and a Google internet search were conducted to find literature on BAV in pregnancy published between 1980 and 2012.

Results: BAV in pregnancy can lead to critical cardiovascular events including aortic dissection, aortic valve disorders, and infective endocarditis; some of these complications may lead to poor maternal outcomes or fetal demise. No differences were noted in either maternal or fetal mortality between syndromic and nonsyndromic pregnant patients with BAV (maternal: 50% vs. 28.6%; $p = 0.4959$; fetal: 25% vs. 0%; $p = 0.1987$). The peak and mean pressure gradients across the aortic valve increased significantly with advancing gestational trimester; a remarkable decrease in peak pressure gradients was seen postpartum. The calculated aortic valve area showed a significant decrease in the third trimester compared with the prepregnancy value, as well as a considerable postpartum decrease.

Conclusion: Syndromic and nonsyndromic BAVs may have similar importance for maternal and fetal mortality. Aortic valve stenosis may become more severe with advancing pregnancy, with attenuation after delivery. Patients may require surgical intervention for the complications of BAV during pregnancy.

Introduction

Bicuspid aortic valve (BAV) is the most common congenital cardiac malformation, occurring in 1–2% of the population [1]. The malformation involves both the valve and the aorta, and patients require lifelong surveillance [2]. The critical complications of this condition are well described and include aortic stenosis, aortic regurgitation, infective endocarditis, and aortic dissection [3]; of these, isolated aortic stenosis is the most common associated disorder [4]. Pregnant women with a severely stenotic BAV may experience cardiovascular deterioration, with 10–30% of this patient subset experiencing complications that, in turn, lead to a high therapeutic abortion rate. Women with mildly to moderately stenotic BAVs may not experience these complications. Cardiac issues during pregnancy, such as deteriorating heart function, angina, and arrhythmias, may require prompt surgical intervention, posing risks to both mother and fetus [5]. However, the natural progression of this condition during pregnancy and its effects on maternal and fetal outcomes have not been sufficiently elucidated at this point.

Materials and methods

A MEDLINE database search and a Google internet search was conducted of the English literature for articles published between 1980 and 2012. “Bicuspid aortic valve” and “pregnancy” were the two main search terms; once articles were collected, the reported complications of BAV were carefully scrutinized. A total of 100 publications were obtained through these searches, but 50 were unrelated to BAV and pregnancy and were therefore excluded. Review articles with no substantial data (8 articles), original articles or case reports with missing or obscure data (12 articles), articles oriented toward the fetus rather than the parturient (6 articles), and articles describing the onset of cardiac events during the postpartum period (1 article) were excluded. A total of 23 reports were included in this study.

The quantitative data reported for patients with a stenosed BAV were analyzed and compared using the paired or unpaired *t* test.

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Fisher exact test was also used for comparison of frequency. A p value <0.05 was considered statistically significant.

Results

A total of 33 pregnant women, described in 23 reports, developed cardiovascular sequelae of BAV including aortic dissection, aortic stenosis, aortic regurgitation, and infective endocarditis. Aortic stenosis was the most common complication, with aortic dissection the second most common (Table 1).

Aortic dissection

Aortic dissection occurred in 11 pregnant women, described in 10 reports [6–15]. Their age was 30.4 ± 5.1 years (range, 19–36 years; median, 30 years). Nine patients developed aortic dissection during pregnancy, at a mean of 32.7 ± 5.6 weeks' gestation (range, 21–38 weeks; median, 32 weeks), one patient developed the complication 2 weeks after a vaginal delivery, and the final patient developed a dissection on Day 2 after cesarean section. Four of the 11 patients (36.4%) were primigravid. The onset symptoms were described in eight patients: chest pain in four patients (50%), and one patient (12.5%) each with epigastric pain, chest discomfort, syncope, and shortness of breath. Five patients (45.5%) had a delay of 61.2 ± 91.2 hours (range, 3–216 hours; median, 12 hours) from symptom onset to hospital admission, and four patients (36.4%) had a delay of 16 ± 10.3 hours (range, 1–24 hours; median, 19.5 hours) from admission to diagnosis. The diagnosis was established by noninvasive imaging methods (echocardiography, ultrasound, computed tomography, or magnetic resonance imaging) in 10 patients (90.9%); in one patient (9.1%), the diagnosis was made at autopsy. The aortic dissection was type A in 10 patients (90.9%) and type B in one patient (9.1%). The diameter of the aorta at the widest part of the dissection was recorded in four patients, with a mean value of 46 ± 17.5 mm (range, 29–70 mm; median, 42.5 mm). Three patients (27.3%) had associated aortic valve regurgitation, and one patient (9.1%) had mitral valve prolapse.

BAV was the sole risk factor for aortic dissection in six patients (54.5%), three patients (27.3%) had Turner syndrome (1 with associated coarctation of the aorta), one patient (9.1%) had Marfan syndrome, and one patient (9.1%) used tobacco. Urgent aortic repair was performed upon dissection diagnosis in nine patients (81.8%), a 2-hour delay occurred in one patient (9.1%), and one patient (9.1%) experienced sudden death before surgery could be performed. Ascending aorta replacement was performed in four patients (36.4%), ascending aorta replacement with aortic valve replacement in one patient (9.1%), the Bentall operation in three patients (27.3%), and aortic valve replacement with aortic root plication and descending aorta replacement in one patient (9.1%). Urgent cesarean section for aortic dissection was performed in five patients (45.5%) at 33 ± 3.6 weeks' gestation (range, 28–37 weeks; median, 32 weeks). Aortic surgery was performed at a mean of 32.3 ± 3.8 weeks' gestation (range, 28–37.3 weeks; median, 32 weeks). Seven (63.6%) patients survived; one patient (9.1%) died suddenly before the diagnosis could be made, and three patients (27.3%) died suddenly after aortic repair. The mortality was 36.4%.

Patients with aortic dissection were further divided into two subgroups: syndromic and nonsyndromic. The former included patients with Marfan or Turner syndrome ($n = 4$), and the latter had patients with no features of any syndrome ($n = 7$). No differences were noted in either maternal or fetal mortality between the syndromic and nonsyndromic pregnant patients (maternal: 50% vs. 28.6%; $p = 0.4959$; fetal: 25% vs. 0%; $p = 0.1987$).

Table 1
Complications of bicuspid aortic valve during pregnancy.

Complication	Report, *n* (%)	Case, *n* (%)
Aortic stenosis	9 (37.5)	18 (52.9)
Aortic dissection	10 (41.7)	11 (32.4)
Infective endocarditis	3 (12.5)	3 (8.8)
Aortic regurgitation	2 (8.3)[a]	2 (5.9)[a]

[a] One patient from one report had aortic valve regurgitation caused by infective endocarditis.

Aortic valve stenosis

The transvalvular pressure gradients and calculated aortic valve areas in 18 pregnant patients with stenotic BAVs were collected from nine reports [16–24]. The mean patient age was 27.0 ± 6.9 years (range, 16–38 years; median, 28 years). Severe aortic stenosis was present in 15 patients (83.3%) and moderate stenosis in three patients (16.7%). Four patients (25%) manifested congestive heart failure. Six patients required cesarean section at a mean gestational age of 33.8 ± 4.5 weeks (range, 28–39 weeks; median, 33.5 weeks). The fetal monitoring was described for eight patients: seven tracings (87.5%) indicated good fetal well-being but one fetus (12.5%) exhibited "distress" and an urgent cesarean delivery was performed [19].

Surgical intervention for aortic valve stenosis was required in six patients (33.3%) of which four (66.7%) had balloon aortic valvuloplasty during pregnancy [18,20,23], and two (33.3%) underwent aortic valve replacement after delivery [16,22]. The peak and mean pressure gradients across the aortic valve increased significantly with advancing pregnancy trimester (peak: 92.8 ± 13.9 mmHg vs. 65.3 ± 19.6 mmHg; $p = 0.0232$; mean: 48.5 ± 7.7 mmHg vs. 35.0 ± 4.8 mmHg; $p = 0.0249$). A remarkable decrease in the peak pressure gradients was seen postpartum, compared with the values noted in the third trimester (54.8 ± 29.4 mmHg vs. 92.8 ± 13.9 mmHg; $p = 0.0310$; Fig. 1). The calculated aortic valve area showed a significant decrease in the third trimester compared with its prepregnancy value (0.74 ± 0.04 cm^2 vs. 1.03 ± 0.26 cm^2; $p = 0.0343$); this value again decreased considerably in the postpartum period, although the reduction in area did not reach statistical significance (Fig. 2).

Aortic valve regurgitation

Two young pregnant patients with BAV developed aortic regurgitation during pregnancy. The first patient was 16 years old and presented with dyspnea and orthopnea at 32 weeks' gestation, then with breathlessness at 34 weeks' gestation [25]. She underwent cesarean delivery at 34 weeks due to the deterioration in her heart function. Her female baby did well at birth, with Apgar scores of 3 and 9 at 1 minute and 5 minutes, respectively. A maternal cardiac function evaluation performed 3 weeks after delivery showed a left ventricular diastolic dimension of 60 mm and a valvular pressure gradient of 33 mmHg.

The second patient was a 23-year-old woman with severe aortic valve regurgitation present at 21 weeks' gestation. Urgent aortic valve replacement was performed but the patient experienced subsequent intrauterine fetal demise [26].

Infective endocarditis

Three pregnant women with BAV experienced infective endocarditis (Table 2) [26–28]. One patient died suddenly, and the diagnosis was made by autopsy. The other two patients underwent aortic valve replacement for decreased heart function and both