Should Aortas in Patients with Bicuspid Aortic Valve Really Be Resected at an Earlier Stage than Tricuspid? CON

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• Aorta • Bicuspid • Tricuspid • Aortic resection

Bicuspid aortic valve (BAV) is the most common congenital heart malformation, affecting 1% to 2% of the population, and constitutes an important risk factor for the development of aortic valve disease. Unlike patients with normal tricuspid aortic valves (TAVs), only 20% of patients with congenitally BAV will maintain a normally functioning valve, and most will develop BAV-related complications.[1] BAV results in early valvular calcification and stenosis in three-quarters of patients, valvular insufficiency in 15%, and a mixed lesion in 10%.[2] Thirty percent of patients with BAV will also develop infective endocarditis. In addition, BAV is intimately associated with abnormalities of the aortic wall, such as ascending aortic dilatations and aneurysm formation.[3] The prevalence of aneurysmal dilatation in the literature ranges from 33% to 80%. This variation is based on aortic size cutoffs for definitions of aortic aneurysms and normative values for age and body surface area.[4]

Given the high incidence of sequelae, BAV accounts for considerable morbidity and mortality compared with other congenital cardiac abnormalities. The aortic dilatation that occurs with BAV arises more frequently and at a younger age than it does in patients with TAV, and the clinical significance of the correlation between BAV and ascending aortic dilatation is based on the potential for aortic dissection and rupture.[5] Indeed, the associated aortic pathology has significantly complicated the surgical treatment regimens in patients with BAV, because clinicians have realized that the disease process between the valve and aortic wall is intimately associated, yet the underlying etiologic link, whether physiologic or genetic, is still not completely understood. Defining the physiologic or potential molecular biologic basis for aneurysm formation in BAV is critical for understanding disease progression and designing appropriate evidence-based strategies for intervention.

This review explores the pathologic process in the development of BAV, examines the relationship between ascending aortic dilatation and aneurysm formation in the setting of valvular stenosis, explores the relevant genetic basis for aneurysm formation in BAV, and evaluates the current recommended treatment guidelines for

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concomitant aneurysms in patients with BAV. Based on the available evidence, the authors extend a strong argument for a more conservative approach to treatment of concomitant ascending aortic dilatation in BAV disease.

THE BAV

Normal development of the semilunar valves occurs with the advent of valve swellings of subendocardial tissue located near the luminal surface of the aorticopulmonary trunk. Morphologic changes occur during valvulogenesis, which include tissue resorbtion and remodeling, ultimately leading to formation of the thin-walled trileaflet semilunar cusps of the TAV. In BAV development, there is a disruption of this developmental process.

The normal aortic valve is a TAV, with 3 equal-sized leaflets or cusps and 3 clear lines of coaptation. Congenital BAVs are a genetic variant in 1% to 2% of the population, with 2 functional cusps, unequal in size due to fusion during valvulogenesis. This variant is the result of a complex developmental process, not simply the fusion of 2 normal cusps. The larger leaflet in BAV is referred to as the conjoined leaflet. This fused leaflet contains a central raphe, or fibrous ridge, representing the site of congenital fusion. The central raphe is identifiable in most patients with BAV, and has been shown not to contain valve tissue on pathologic examination.[6] In contrast, the TAV is characterized by 3 distinct cusps without a raphe (**Fig. 1**).

In congenital BAV, the cusps are conjoined. Two commissures are present, and neither is partially fused. The shallow, upper aspect of the raphe is usually distinctly below the sinotubular junction, terminating well below the cuspid line of closure.[7,8] The circumferential distances between each of the 3 commissures are approximately equal in the normal TAV, and in congenital BAV the distances between the 2 commissures are also similar.

In a minority of cases, BAV occurs as an acquired pathologic process related to rheumatic heart disease or a degenerative, nonrheumatic inflammatory process. In these cases, the true commissure is fused.[8] This commonly originates at the level of the sinotubular junction, and is maintained at a level similar to the closing edge of the remainder of the valve.

In congenitally bicuspid valves, fusion of the raphe is typically observed between the right and left cusps in 86% of BAV cases.[2] The coronary arteries also tend to arise from the front of the cusps in which the raphe is present. In congenital BAV, the left main coronary artery is frequently up to 50% shorter and the coronary circulation tends to be a left-dominant pattern, more than in normal TAV hearts.[6,8]

BAV in the General Population

Prevalence of BAV in the population is 1% to 2%. The numbers are surprising when the potential clinical consequences are considered in light of a current population of the United States of approximately 305 million. Thus, there are 3 million to 6.1 million United States citizens with BAVs.

Most of the information available regarding prevalence of BAV in the population originated from pathology centers with a wide range of variation. BAV has not been considered rare since 1886, when Osler reported that 1.2% of 800 routine autopsies revealed this congenital malformation, excluding infective endocarditis (**Table 1**).[1,9] In 1970, Roberts[9] found 13 BAVs in 1440 routine post mortems on adults (0.9%), but he argued that this could reach 2% by including patients with cardiac disease. The most reliable

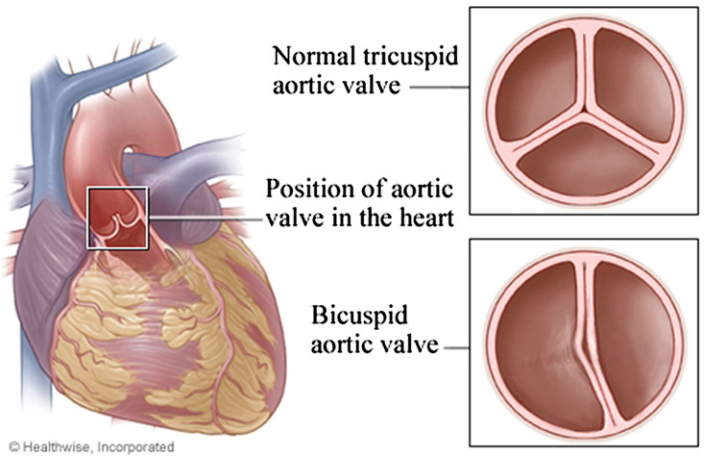

Fig. 1. The 3 distinct cusps, equal in size in the normal TAV. In BAV, 2 functional cusps are present with the conjoined leaflet containing a central raphe or fibrous ridge (Bicuspid aortic valve image provided courtesy of Healthwise, Incorporated. All rights reserved.).