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Aortopathy Involvement in Patients with Bicuspid Aortic Valve: A Mini Review

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ABSTRACT

Around 0.5% to 2% of the general population have a Bicuspid Aortic Valve disease (BAV), which is considered to be the most common valvular congenital lesion. A marked masculine predominance with a sex ratio of 3:1.

A serious follow-up is required after initial diagnosis, due to high rate of morbi-mortality related to BAV, as most patients develop during life time valvular or aortic complications including aortic stenosis, aortic regurgitation, aortic dissection or secondary aortic dilatation called bicuspid aorthopathy.

The heterogeneous spectrum of related conditions to BAV leads to difficulties in follow-up and treatment options

INTRODUCTION

Around 0.5% to 2% of the general population have a Bicuspid Aortic Valve disease (BAV), which is considered to be the most common valvular congenital lesion [1]. A marked masculine predominance with a sex ratio of 3:1 [2,3].

A serious follow-up is required after initial diagnosis, due to high rate of morbi-mortality related to BAV, as most patients develop during lifetime valvular or aortic complications including aortic stenosis, aortic regurgitation, aortic dissection or secondary aortic dilatation called bicuspid aorthopathy [4,5].

The heterogeneous spectrum of related conditions to BAV leads to difficulties in follow-up and treatment options [6].
Transthoracic echocardiography (TTE) is the first-line imaging modality used for the diagnosis and monitoring of patients with BAV. It provides data regarding concomitant aorthopathy and valvular dysfunction.

EPIDEMIOLOGY

The GenTAC registry presented at the 2015 American Heart Association Scientific Sessions meeting showed that men had more frequently type 1 BAV with fusion between the left and right coronary cusps than women 81.5% versus 69.0%, P=0.03, whereas type 1 BAV with fusion between the right and noncoronary cusps was more frequently observed in women (18.5% versus 31.0%, P=0.03) [7]. No study so far explains sex differences of bicuspid aortic valve morphology and the masculine predominance.

The exact pathogenesis of the formation of bicuspid aortic valves is not yet fully clair and understood.

Classification (Figure 1)

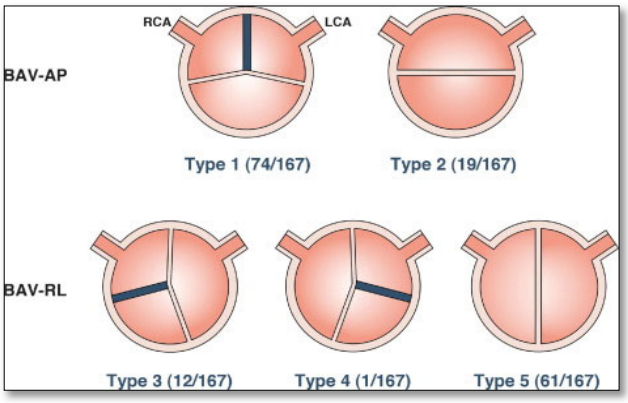

Figure 1. Classification of Bicuspid Aortic Valve disease (BAV).
BAV: Bicuspid Aortic Valve
AP: Anterior-Posterior
RL: Right-Left

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AORTIC INVOLVEMENT

Aortic dissection

Most patients with a bicuspid aortic valve will develop some complication during lifetime involving the aorta, two important complications are aortic dissection and aortic rupture.

The risk of aortic dissection seems to be higher than in the general population (around 8 times more): Even though the relative risk is high, the absolute incidence of aortic dissection remains very low.

No added mortality was noted during follow-up of a large BAV community cohort [8,9].

Aortic dilatation

Bicuspid aortic valve aortopathy is generally an indolent disease, with slow mean growth rates.

Type I BAV is more associated with dilatation at the annulus and sinus of Valsalva with a percentage of 30%. There is relative sparing of this region of the aorta in patients with type II BAVs.

Sinus of Valsalva is often more dilated in males compared to females.

Superimposed valve dysfunction has a great impact and considered as a crucial factor in dilatation progression:

Aortic Regurgitation is associated with increased size of annular SOV and ascending aortic with linear relation of aortic diameters to the severity of AR [10].

BAV is a known cause of Aortic Stenosis, BAV aortopathy is noted independent of AS hemodynamic profile [11]. However, the hemodynamics changes related to AS may increase dimensions of the ascending aorta in the presence of moderate-severe AS [8].

Early surgery has been recommended for patients with aortic diameter increase >2 mm/year [12].

Although typically progressive in its nature (diameters remained stable over time in only 1/3 of patients).

Recently, Michelena et al. [5] suggested that morbidity (including aortic valve and/or ascending thoracic aorta replacement surgeries, the presence of severe AR and/or AS, thoracic aortic aneurysm but also aortic dissections as well as endocarditis) would be more important in man while mortality would be higher in women in the two cohorts analyzed.

Several studies have raised the hypothesis of the BAV phenotype and its role in the rapid progression of valve dysfunction and especially in the evolution of arterial diameters [13], however, the results in the literature are still ambiguous.

The contradictory results of the studies as well as the low numbers of patients studied [13,14] did not allow the creation of a guideline to suggest carrying out a precise valvular phenotyping of the BAV and to organize follow-up intervals specific for different phenotypes, so the use of TEE for this approach was not specifically recommended, except in certain situations [15].

CONCLUSION

Monitoring of patients with BAV is crucial even though long-term outcomes are often reassuring. BAV patients are exposed to an associated aortopathy risk, with risk of aneurysm formation and aortic dissection.

Despite the low incidence of dissection in studied groups, it still higher than general population especially in older patients and progression of aortic dilatation being more rapid with superimposed valvular dysfunction.

New studies should concentrate on identifying markers or clinical/echocardiographic predictive signs for refining risk prediction of aortic dissection and rapid progression of aortopathy in these patients.

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