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Short Communication

Current Approach to Bicuspid Aortic Valve: Review

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Bicuspid aortic valve (BAV) is the most common congenital cardiac anomaly. The most frequent associated finding is dilation of the proximal ascending aorta secondary to abnormalities of the aortic media. The BAV may function normally throughout life, may develop complications can include aortic valve stenosis or incompetence, endocarditis, aortic aneurysm formation, and aortic dissection. Aortic root dilatation is common in BAV, even when the valve is haemodynamically normal, and consequently aortic dissection usually occurs in previously asymptomatic patients. All patients should therefore be regularly reviewed to identify progressive root dilatation with a view to preempting dissection by prophylactic surgery.

Keyword: bicuspid aortic valve; aortic media abnormality; aortic aneurysm and dissection.

INTRODUCTION

The bıcuspıd aortıc valve

Bicuspid aortic valve (BAV) represents the most common cardiac congenital malformation in the adult age. The BAV is seen in 1% to 2% of the population and may be complicated by aortic stenosis or aortic insufficiency and infective endocarditis. It may be associated with abnormalities of the aortic wall such as coarctation of the aorta, aortic dissection, and aortic aneurysm. Most patients with a BAV develop some complications during life (Braverman et al., 2005; Yener et al., 2002).

Congenital coronary anomalies, coronary atherosclerosis, and calcification have been described in association with bicuspid aortic valve (Ward, 2000).

The congenitally BAV may function normally throughout life, may develop progressive calcification and stenosis or may develop regurgitation with or without infection. Aortic root dilatation is common in BAV, even when the valve is haemodynamically normal, and consequently aortic dissection usually occurs in previously asymptomatic patients (Tadros et al., 2009; Pachulski et al., 1991).

BAV are in most cases remain undetected until infection or calcification supervenes (Lamas and Eyky, 2000).

Aortic stenosis and regurgitation, infective endocarditis and aortic dissection are the most common complications. Left coronary artery dominance is more common in patients with a BAV (29-56.8%) and in 90% of cases, the left main coronary artery is less than 5 mm in length (Hutchins et al., 1978; Higgins and Wexler, 1975). The ignorance of these associations may cause an inadequate myocardial preservation and an increased risk of myocardial infarction (Presbitero et al., 1987; Demir, 2009).

The histological findings of BAV are nonspecific, and had been described by several authors in patients with Marfan syndrome (McKusick, 1972; Keane et al., 2000; Sá et al., 1999). The histopathological appearance of thoracic aortic aneurysm in Marfan and BAV is similar, and includes evidence of vascular smooth muscle cell (VSMC) apoptosis and extracellular matrix degeneration in the absence of a significant inflammatory response

(Schmid et al., 2004).

Abnormalities in the ascending aorta of the patients with BAV, specifically premature medial layer VSMC apoptosis, have been described, explaining the higherthan-expected prevalance of aortic dissection in these patients (Nistri et al., 2002).

Also recently studies show less elastic tissue in the aortas of BAV patients (Parai et al., 1999; De Backer et al., 2006).

In patient with BAV there are fibrillin, fibronectin, and tenascin abnormality. Additionally Bonderman et al suggested that a primary role for VMSC apoptosis in the development of aneurysm these patients (Bonderman et al., 1999).

The FBN1 gene encodes fibrillin-1, a large glycoprotein that is secreted from cells and deposited in the extracellular matrix in structures called microfibrils. Microfibrils are found at the periphery of elastic fibers, including the elastic fibers in the medial layer of the ascending aorta, and in tissues not associated with elastic fibers (Hiratzka et al., 2010).

Fedak et al suggested that fibrilin-1 content was reduced in patient with BAV (Fedak et al., 2003).

The histopathological appearance of thoracic aortic aneurysm in Marfan and BAV is similar, and includes evidence of VSMC apoptosis and extracellular matrix degeneration in the absence of a significant inflammatory response .Abnormalities in the ascending aorta of the patients with BAV, specifically premature medial layer smooth muscle cell apoptosis, have been described, explaining the higherthan-expected prevalance of aortic dissection in these patients (Schmid et al., 2004; Parai et al., 1999).

Recently Kiotsekoglou et al. demonstrated significant biventricular diastolic and biatrial systolic and diastolic dysfunction in Marfan syndrome patients. Also speculated that these findings suggest that Marfan syndrome affects diastolic function independently. Diastolic abnormalities could be attributed to fibrillin-1 deficiency and dysregulation of transforming growth factor-b activity in the cardiac extracellular matrix (Kiotsekoglou et al., 2009). Recently Santarpia et al demonstrated that left ventricular longitudinal ($p=0.01$), circumferential ($p=0.01$) and radial ($p<0.001$) strain (%) were lower in BAV (Santarpia et al., 2011).

Recently published Guidelines (Hiratzka et al., 2010) for the diagnosis and management of patients with thoracic aortic disease recommendations for BAV are summarized below:

CLASS I

1. First-degree relatives of patients with a BAV, premature onset of thoracic aortic disease with minimal risk factors, and/or a familial form of thoracic aortic aneurysm and dissection should be evaluated for the presence of a BAV and asymptomatic thoracic aortic disease. (Level of Evidence: C)

2. All patients with a BAV should have both the aortic root and ascending thoracic aorta evaluated for evidence of aortic dilatation (Level of Evidence: B)

3. Should undergo elective operation at smaller diameters (4.0 to 5.0 cm) to avoid acute dissection or rupture. (Level of Evidence: C)

4. Patients with a growth rate of more than 0.5 cm/year in an aorta that is less than 5.5 cm in diameter should be considered for operation. (Level of Evidence: C)

5. Patients undergoing aortic valve repair or replacement and who have an ascending aorta or aortic root of greater than 4.5 cm should be considered for concomitant repair of the aortic root or replacement of the ascending aorta. (Level of Evidence: C)

6. Elective aortic replacement is reasonable for patients with BAV when the ratio of maximal ascending or aortic root area (Tr2) in cm2 divided by the patient's height in meters exceeds 10 (CLASS IIa, Level of Evidence: C).

Consequently aortic stenosis and regurgitation, infective endocarditis and aortic dissection are the most common complications of BAV additionaly this process continues after valve replacement. The person with BAV requires continuous surveillance to treat associated lesions and prevent complications. Arterial hypertension should be meticulously controlled. Smoking should be discouraged and control of hypercholesterolaemia considered, in view of the impact of these factors on the development of aortic stenosis. Aortic root dilatation is common in BAV, even when the valve is haemodynamically normal, and consequently aortic dissection usually occurs in previously asymptomatic patients. All patients should therefore be regularly reviewed to identify progressive root dilatation.

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