Aortic Aneurysm in Bicuspid Valve: Is it Genetic Disorder or Caused by Flow Dynamics

Murat Ugur*

Department of Cardiovascular Surgery, Health Sciences University, Sultan Abdulhamid Training Hospital, Istanbul, Turkey

*Corresponding Author: Murat Ugur, Department of Cardiovascular Surgery, Health Sciences University, Sultan Abdulhamid Training Hospital, Istanbul, Turkey.

Received: June 14, 2017; Published: June 22, 2017

Bicuspid aortic valve (BAV) is the most common congenital heart malformation, with a risk factor for the development of aortic valve disease and ascending aortic dilatation [1]. In comparison with normal valve morphology, aortic aneurysm develops in younger ages (49.0 years to 64.2 years) and the rate of operation is higher (72.8% vs 44.8%) with BAV [2].

Due to increased rate of aortic diameter and operation requirement the main cause of aortic pathology has been researched. Genetic and pathogenic mechanism of the aortic dilatation has been advocated by different surgeons.

Some authors reported that decreased fibrillin 1, increased matrix metalloproteinases, cystic medial necrosis, elastic fragmentation and smooth muscle cell reorientation are associated with aortic dilatation in BAV [3,4]. These authors strengthened their thesis with progressive enlargement of the aorta even after successful AVR [5,6]. Borger and colleagues reported an 11% incidence of ascending aortic complications during 10 years of follow-up after AVR for BAV [3]. 1% aortic dissection, %0.9 ascending aortic replacement was reported during the follow up of 1286 AVR patients due to BAV [5]. Aortic dilatation was revealed in one third of first degree relatives of BAV patients, although trileaflet anatomy [7].

Post-valvular flow mechanism is the other factor that effecting aortic dilatation in BAV [1]. Fused cusps changes flow dynamics. Aortic wall shear stress is increased due to changed flow dynamics in BAV. Increased tensile and shear stress affects convex wall of the ascending aorta [4]. Ascending aortic dilatation develops due to transvalvular flow dynamics rather than aortic morphology in aortic stenosis. However, aortic dilatation might present in younger ages in BAV since it is a congenital disorder and ascending aorta is exposed abnormal flow dynamics longer [8]. Regional stress of the wall is higher in non-coronary sinus and sinotubular junction than the rest of the aorta. Non-coronary sinus is mostly dilated segment of the aortic root in BAV due to flow dynamics. Replacement of the degenerated non-coronary sinus in the asymmetric enlargement of the aortic root has acceptable long-term results without further aortic dilatation and reoperation for aortic complications in 12 years [9].

In BAV, surgery indication of the ascending aortic dilatation is important to avoid further enlargement even it is caused by genetic disorder or flow dynamics. Current guidelines recommend surgery to aortic aneurysm in patients with BAV if aortic diameter is greater than 5.0 cm [10]. The main dilemma is the surgical approach to the aortic dilatation with the diameter of 4.0 - 4.5 cm. Since AVR does not prevent the aortic enlargement in mild-moderate aortic dilatation, surgical approach to the ascending aorta at the operation is important to decrease further dilatation. Separate graft replacement to the ascending aorta has lower risk than the composite graft replacement. However, remained asymmetrically dilated aortic sinus might trigger root enlargement in the long term. Replacement of the dilated aortic sinus could avoid further dilatation in mild aortic enlargement in the patients with BAV in follow up period.

Bibliography


Citation: Murat Ugur. “Aortic Aneurysm in Bicuspid Valve: Is it Genetic Disorder or Caused by Flow Dynamics”. EC Cardiology 3.3 (2017): 72-73.
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**Citation**: Murat Ugur. “Aortic Aneurysm in Bicuspid Valve: Is it Genetic Disorder or Caused by Flow Dynamics”. *EC Cardiology* 3.3 (2017): 72-73.