Heart, Lung and Circulation (2014) **23,** 32–38
1443-9506/04/$36.00
http://dx.doi.org/10.1016/j.hlc.2013.05.644

ORIGINAL ARTICLE

Pattern of Aortic Dilatation in Different Bicuspid Aortic Valve Phenotypes and its Association with Aortic Valvular Dysfunction and Elasticity

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Received 6 December 2012; received in revised form 8 February 2013; accepted 18 May 2013; online published-ahead-of-print 11 July 2013

Objectives We aim to study the pattern of aortic dilatation in different BAV phenotypes and to find any correlations between aortic dilatation, aortic elasticity and AS and/or AR in our local population.

Methods All BAV patients and controls were retrospectively studied. Aortic distensibility and stiffness index of the ascending aorta were calculated.

Results A total of 191 patients with BAV and 180 controls were enrolled. Aortic dilatation involving a single site was more common with dilatation involving only the ascending aorta (R–N phenotype) and aortic root (N–L phenotype). AR was most common in patients with N–L and R–L phenotypes and AS in R–N phenotype. Aortic elasticity had no correlation with the different BAV phenotypes.

Conclusions There are different patterns of aortic dilatation in different BAV phenotypes, which may further contribute to the development of aortic stenosis or regurgitation. Aortic elasticity is independent of the BAV phenotypes and is impaired in BAV patients regardless of AS or AR severity.

Keywords Bicuspid aortic valve • Aortic elasticity • Aortic valve stenosis • Aortic valve regurgitation

Introduction

Bicuspid aortic valve (BAV) is the most common form of adult congenital heart disease [1–4]. BAV includes different morphologic phenotypes [5,6] characterised by various haemodynamic profiles and is usually associated with dilatation of the aorta, aortic stenosis (AS) and aortic regurgitation (AR) [7,8]. When compared to patients with a normal trileaflet aortic valve, patients with BAV have larger aortic dimensions and an increased rate of aortic dilatation over time, with consequent higher risk of developing aortic aneurysm, aortic dissection and rupture [9,10].

Some studies [11–14]] had suggested that different BAV phenotypes may have distinctive genetic inheritance. From clinical observation, we found that different BAV phenotypes had distinctive sites of aortic dilatation. Hence, we aim to study the pattern of aortic dilatation in different BAV phenotypes and its relationship with aortic stenosis, aortic regurgitation and aortic elasticity.

Methods

All congenital BAV patients from our echocardiography database of the outpatient department, National Heart Centre

Abbreviations: BAV, bicuspid aortic valve; AS, aortic stenosis; AR, aortic regurgitation; AA, aortic annulus; SOV, sinuses of Valsalva; STJ, sinotubular junction; Asc Ao, ascending aorta; DIS, distensibility; SI, stiffness index; SBP, systolic blood pressure; DBP, diastolic blood pressure; MAP, mean blood pressure

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Singapore and age-matched normal controls who had echocardiography done for evaluation of murmurs, other cardiac symptoms and subsequently found to be normal were retrospectively studied. Patients with aortic coarctation, transposition of the great arteries or other congenital cardiac defects were excluded. The ethics committee of our hospital and the local research ethics committee had approved this study.

The cuff blood pressure, systolic (SBP) and diastolic (DBP) blood pressure were measured standarised with an automated mercury cuff sphygmanometer after patients were at rest for at least 5 min. All patients underwent full transthoracic echocardiograms (including 2-D M-mode and colour Doppler.) and the blood pressure was measured at the same time. The zoom function was used when the aorta was scanned and the commissures were carefully examined. The diameters of the aortic root (aortic annulus, sinus of Valsalva and sinotubular junction) (Fig. 1) and the ascending aorta (at the site of maximal dilatation of ascending aorta) were measured by 2-D and M-mode in the parasternal long-axis view, perpendicular to the long axis of the vessel, from leading edge to leading edge during end-systole [15]. The dilatations of aortic root (at any one of the three levels) and ascending aorta was defined as a diameter greater than the mean + 2SDs of the values of normal controls [3] which is further subdivided into three groups: less than 20 years, 20–40 years and older than 40 years groups, and each patient's aortic diameter was compared with normal range for their age group. The average of three consecutive measurements was taken.

Aortic distensibility (DIS) and stiffness index (SI) were calculated as [16,17]:

$$\mathrm{DIS} = 2 \times (\mathrm{AoS} - \mathrm{AoD})/\mathrm{AoD} \times (\mathrm{SBP} - \mathrm{DBP}) \times 1000$$

$$\mathrm{SI} = \ln(\mathrm{SBP}/\mathrm{DBP})/(\mathrm{AoS} - \mathrm{AoD})/\mathrm{AoD}$$

where AoS is the dimension of systolic ascending aorta at the end-systole (end of T wave on ECG), AoD is the dimension of diastolic ascending aorta at the end-diastole (end of Q wave on ECG), SBP is the systolic blood pressure, DBP is the diastolic blood pressure, and In is a natural logarithm. AR and AS were graded as mild, moderate and severe using ACC/AHA guidelines [18].

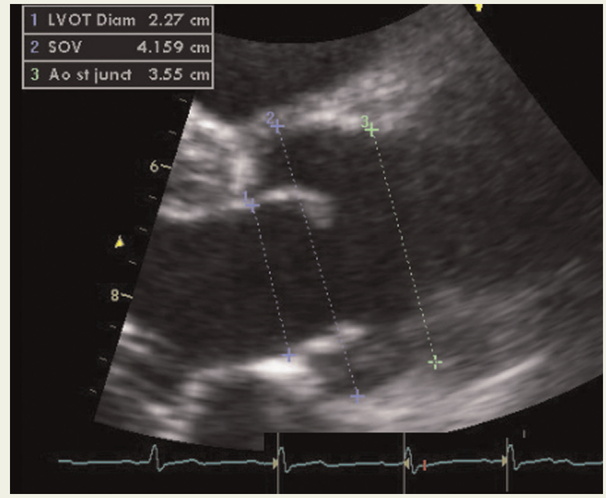

Figure 1 Measurements of aortic annulus, sinus of Valsalva and sinotubular junction.

Three BAV phenotypes were defined as: R–L: fusion of right and left coronary cusp, R–N: fusion of right and non-coronary cusp and N–L: fusion of non-coronary and left coronary cusp (Fig. 2).

Statistic Analysis

Data were summarised as mean ± SD and categorical data were presented as percentages. Independent-samples *t*-test was used to compare aortic distensibility and stiffness index in BAV patients and normal control group. Multivariate analysis was then performed by entering into model variables that were considered significant on univariate analysis.

To assess intra- and inter-observer variability, echocardiographic images from 20 patients were randomly selected. Images were independently presented and read in random order by two cardiologists who were blinded to patients' results. Intra- and inter-observer reproducibility of echocardiographic derived measurement were assessed by calculating the mean difference and standard deviation between results, with percentage variability equal to the mean of

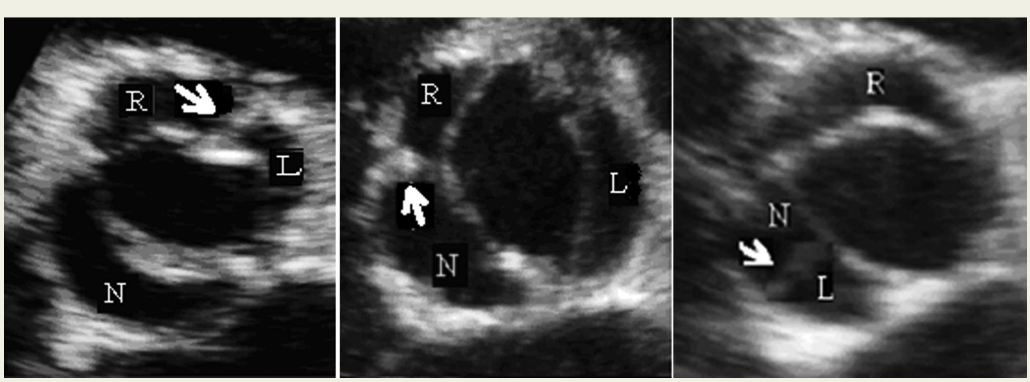

Figure 2 Three different BAV phenotypes. R–L: right–left cusp fusion with raphe (arrow). R–N: right–non cusp fusion with raphe (arrow). N–L: non–left cusp fusion with raphe (arrow).