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Association between leaflet fusion pattern and thoracic aorta morphology in patients with bicuspid aortic valve

Bryce Merritt^{1*}, Alexander Turin¹, Michael Markl^{1,2}, James Carr^{1,3}

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Background

Bicuspid Aortic Valve (BAV) is the most common congenital heart defect and is associated with a high risk of aortic dilation. We sought to determine if patients with certain BAV phenotypes are predisposed to particular morphological abnormalities of the thoracic aorta.

Methods

One hundred and ninety-two patients with BAV who underwent magnetic resonance angiography between January 2007 and July 2010 were retrospectively identified. Aortic morphology was examined through measurements of diameter at nine levels along the thoracic aorta, 3D volume of the ascending aorta, vessel eccentricity, and assessment of aortic root and arch morphology. Diameters and volumes were normalized to body surface area. BAV phenotypes were defined by fusion of the right and left coronary cusps (Type 1), right and noncoronary cusps (Type 2), and left and non-coronary cusps (Type 3).

Results

We found 140 patients (73%) with Type 1 fusion, 46 patients (24%) with Type 2 fusion, and 6 patients (3%) with Type 3 fusion. Mean diameter at the sinus of Valsalva was significantly larger in Type 1 patients (1.97 vs. 1.77 cm/m2; p<0.001). Mean aortic volume in the proximal ascending aorta was significantly greater in Type 2 patients (0.93 vs. 0.60 cm3/m2; p<0.01). Type 2 patients possessed greater dimensions in the distal ascending aorta and proximal aortic arch, and were also more likely to have ascending distention of the aortic root.

 $^{\overline{1}}\mbox{Radiology},$ Northwestern University, Feinberg School of Medicine, Chicago, IL, USA



Figure 1 Cardiac magnetic resonance image annotated to illustrate the 9 levels at which the aortic diameter was measured. 1. Aortic annulus; 2. Sinus of Valsalva; 3. Sinotubular junction; 4. Proximal ascending aorta; 5. Mid-ascending aorta; 6. Distal ascending aorta; 7. Proximal to the innominate trunk (proximal aortic arch); 8. Distal to the left subclavian artery (distal aortic arch); 9. Descending aorta at the level of the diaphragm.



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| | 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 |
|--------|--------------------------------------|---|--|---|---|--|--|--|------------------------------|
| Type 1 | 1.60* | 1.97‡ | 1.82* | 2.03 | 2.01 | 1.78 | 1.65 | 1.27 | 1.15* |
| Type 2 | 1.49 | 1.77 | 1.71 | 1.97 | 2.03 | 1.91† | 1.77 ⁺ | 1.22 | 1.07 |
| Type 3 | 1.56 | 1.72 | 1.83 | 1.87 | 1.88 | 1.68 | 1.55 | 1.23 | 1.07 |
| | *p < 0.05 †p < 0.01 ‡p < 0.001 | Aortic annulus Ascending Aorta Descending ac | ; 2. Sinus of Vals ; 7. Proximal to t orta at the level of | alva; 3. Sinotub he innominate tr f the diaphragm | ular junction; 4. F unk (proximal ao | roximal Ascendin rtic arch); 8. Dista | g Aorta; 5. Mid-) I to the left subel | Ascending Aorta; avian artery (dist | 6. Distal al aortic arch) |
| | 7 1/2r | intions | in an | tic dia | motor | with h | icucni | d porti | |

Conclusions

Our results suggest that BAV with Type 1 fusion is associated with dilation of the aortic root, while BAV with Type 2 fusion is associated with dilation of the distal ascending aorta and proximal arch. Our findings support the role of hemodynamics in the aortic dilation seen in the presence of BAV, and illustrate the morphological heterogeneity that exists among BAV phenotypes.

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Author details

¹Radiology, Northwestern University, Feinberg School of Medicine, Chicago, IL, USA. ²Biomedical Engineering, Northwestern University, Chicago, IL, USA. ³Bluhm Cardiovascular Institute, Northwestern Memorial Hospital, Chicago, IL, USA.

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