Significant Diameter Heterogeneity Between Aortic Root and Aortic Annulus in Bicuspid Aortic Valve

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Introduction

- Bicuspid aortic valve (BAV) is potentially associated with Aortic dilatation and Aortic dissection
- Besides true BAV, there are 3 morphologies of fusion BAV
    - BAV-1: Left and right coronary leaflets fusion (RC-LC)
    - BAV-2: Right and non-coronary leaflets fusion (NC-RC)
    - BAV-3: three-leaflet fusion
    - BAV-4: True bicuspid

Aims

- Retrospectively determine the pattern of dilatation according to the commonest types of fusion BAV
- Assess the possibility to modify patient management according to findings

Method

- Patients with fusion BAV 18 years old or younger
- Retrospective review of echocardiography reports between March 2009 and January 2014
- Availability of fusion morphology described
- Availability of echocardiography diameters of aortic valve, aortic root and ascending aorta
- Exclusion criteria:
    - Diagnosis other than BAV retained at discharge
    - True bicuspid valve (embryologically different)
    - Missing echocardiography data
    - Pathologies that could affect size of the proximal aorta (ex: Turner syndrome, large interventricular defects, Kawasaki disease, Shone's syndrome, …)

❖ *Proximal aortic dimensions were normalized to Z-scores according to equations by Dallaire et al., from «Bias related to body mass index in pediatric echocardiographic Z scores »*

Results

Table 1. Study Population

Basic characteristics	All (n=216)	BAV-1 (n=155)	BAV-2 (n=61)
Age echo 1 (yr)	7.19 ± 5.56	7.08 ± 5.55	7.46 ± 5.63
Male n(%)	161 (74.5)	117 (75.5)	44 (72.1)
Height (cm)	115.4 ± 40.7	115.2 ± 40.9	116.0 ± 40.7
Weight (Kg)	28.5 ± 21.5	28.2 ± 21.5	29.5 ± 21.8
BSA (m^2)	0.93 ± 0.52	0.92 ± 0.52	0.95 ± 0.53
AV sten. n(%)*	66 (30.6)	39 (25.2)	27 (44.3)
Coarctation n(%)	36 (16.7)	26 (16.8)	10 (16.4)

* P = 0,006 RC-LC vs NC-RC

Table 2. Z-Scores According to Fusion Type (mean ± SD)

	RC-LC	NC-RC	p value
Aortic Valve (AV)	0.43 ± 1.71	0.88 ± 1.90	0.11
Aortic Root (AR)	**0.70 ± 1.50**	**-0.08 ± 1.34**	**0.0007**
Ascending Ao (AA)	1.45 ± 2.13	2.04 ± 2.15	0.07

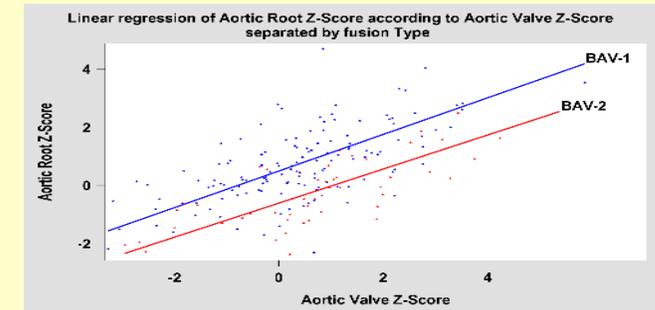### Table 3. Z-Scores According to Fusion Type (mean ± SD) and Presence (st) or Absence of Valve Stenosis (n-st)

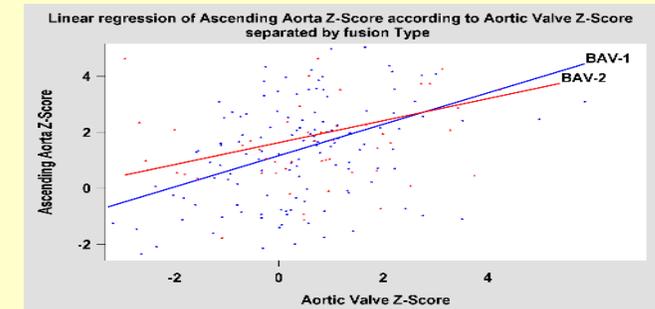|	RC-LC		NC-RC		p-value (inter BAV)	
	st	n-st	St	n-st	st	n-st
AV	0.38 ±1.72	0.39 ±1.79	1.03 ±2.32	0.78 ±1.54	0.34	0.28
AR	0.67 ±1.50	0.79 ±1.57	0.17 ±1.73	-0.25 ±0.98	0.48	**0.00035**
AA	1.42 ±2.11	1.25 ±2.16	2.89 ±2.19	1.38 ±1.90	0.097	0.75

Interpretations

- *No statistically significant difference in AA Z-scores*
- *Higher aortic root Z-score in RC-LC (significant in absence of stenosis) (or absence of coarctation of the aorta; 0,88±1,53 in RC-LC vs 0,04±1,36 respectively; p = 0,0011)*
- *Trend towards larger aortic annulus in NC-RC*

Results

Figure 1.

Linear regression of Aortic Root Z-Score according to Aortic Valve Z-Score separated by fusion Type

Figure 2.

Linear regression of Ascending Aorta Z-Score according to Aortic Valve Z-Score separated by fusion Type

Limitations

1. Retrospective nature of the study
2. Current study did not evaluate follow-up measurements

Conclusion

- Although NOT necessarily dilated (mean Z < 2.0) type-1 BAV (RC-LC) aortic roots are larger than in type-2 (NC-RC)
- Aortic roots represent significant heterogeneity with the aortic valve (unlike type-2 fusion)
- These difference could potentially lead to modifications in patient approach according to his/her BAV morphology.