Comprehensive morphological classification of bicuspid aortic valve by cine CMR in 368 patients.

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Purpose: Bicuspid aortic valve (BAV) has been shown to encompass large heterogeneity in valve morphology and aortopathy expression. A genetic or hemodynamic contribution has been postulated to contribute to progressive remodeling and growth of the ascending aorta; however a dominant prognostic marker has yet to be identified. While pilot studies have shown the fusion of the right-left (RL) or right-non coronary (RN) valve leaflets to be associated with different aortopathic phenotypes, the current 'valve fusion models' are often highly simplified and do not comprehensively describe the large variability in lesion morphology (Fig 1). In the context of the heterogenous expression of aortopathy, a comprehensive method to describe valve morphology is necessary to further understand the relationship between BAV morphology and aortopathy expression.

Methods: BAV patients receiving standard functional CMR and 4D flow MRI between November 2012 and August 2014 were prospectively added to a database for valve morphology analysis. The valve was categorized using the following criteria (Fig 1):
1) Existence of valve raphe: as proposed by Sievers et al (2007): type 0 = no visible raphe; type 1 = presence of a raphe, as identified by leaflet 'fusion', e.g. 1 RL, 1 RN or 1 left-noncoronary (LN); type 2 = presence of 2 raphes (Fig 1b and 1c).
2) Orientation of the valve orifice opening as using a clock face scheme. The inter-atrial septum was used to orientate the 12-6 axis of the clock face (Fig 1b).
3) If present, the position of an eccentric / dominant leaflet (i.e. a patently larger leaflet visible during opening, Fig 1b and 1c).
4) If present, the location of partially fused leaflets were noted.

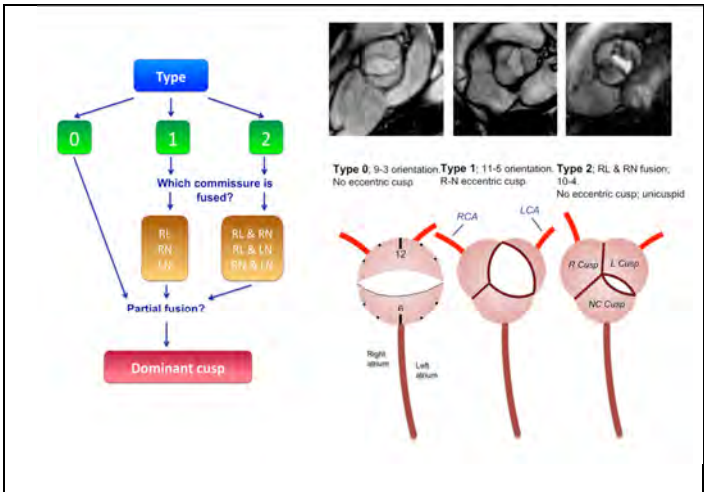

Fig 1. Classification parameters, and diagrammatic and bSSFP images of type 0, type 1 RL and type 2 RL RN.

Results: Of 368 eligible patients, the type 1 RL valve phenotype was the most prevalent morphology with an opening orientation between 8-10 o'clock (table 1, n=185, 50%). When type 0 subjects with a similar opening orientation are included, the 68% incidence of the opening orientation aligns with previous surgical reports (73%, n=304). When combined, the 24% incidence of Type 1 RN and type 0 between 5-7 o'clock aligns well with surgical reports (19%). Of note, eccentric valve leaflets were seen in 213 (58%) and partial fusions were found in 123 (33%) of all cases.

Type	0	1			2		
		RL	RN	LN	RL RN	RL LN	RN LN
Total (368)	111	185	30	5	31	1	5
Eccentric leaflet (213)	64	110	26	4	7	0	2
Clock face							
12 – 6 (38)	19	0	16	0	1	0	2
11 – 5 (48)	25	0	11	1	9	1	1
10 – 4 (42)	14	9	3	0	16	0	0
9 – 3 (229)	50	173	0	1	5	0	0
8 – 2 (10)	2	3	0	3	0	0	2
7 – 1 (4)	1	0	0	0	0	0	0
Partial fusion (123)	19	59	10	4	26	1	4

Table 1: Classification of BAV subtypes using four different criteria specified above.

Discussion and conclusions: The presented method to categorize valve phenotypes aligns well with surgical specimen studies. In addition, MRI offers the capability to categorize the opening dynamics of the leaflets, which may play a significant role in outflow hemodynamics. Regardless of a genetic or hemodynamic contribution, it is important to understand if a relationship exists between valve morphology and aortopoathy. With this in mind, a comprehensive, non-invasive MRI-based method to categorize BAV morphology was demonstrated. The results highlight the considerable heterogeneity of valve morphology.

References: 1) Barker AJ et al. Circ Cardiovasc Imaging. 2012 Jul;5(4):457–66. 2) Sievers H-H, Schmidtke C. J Thorac Cardiovasc Surg. 2007 May;133(5):1226–33.