

## **Bicuspid Aortic Valve Behavior in Elite Athletes**

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## Supplemental Methods

### Echocardiography

Echocardiographic studies were performed according to the 1989 guidelines published by the American Society of Echocardiography and then validated in accordance with the revised guidelines from 2015.<sup>1, 2</sup> Echocardiographic and Doppler studies were performed using the Toshiba SSH-140A ultrasound system (Toshiba Medical Systems, Tochigi, Japan) equipped with 2.5- and 3.75-MHz probes; the Philips SONOS 7500 ultrasound system (Philips Medical Systems, Bothell, WA), equipped with a 1-to3-MHz probe; the Toshiba ARTIDA (Toshiba Medical Systems, Tochigi, Japan) echocardiography system, with a 2-to-4.8-MHz probe; and the Philips iE33 system (Philips Medical Systems, Bothell, WA), with a 1-to-5-MHz cardiac sector probe. Images were obtained using standard imaging planes (parasternal long and short axis, apical, subcostal, and suprasternal views). Aortic valve morphology was evaluated in the parasternal long and short axis. The coronary ostium was visualized in all athletes. BAV was confirmed when 2 cusps were clearly identified in short-axis view (Figure 1); once presence of BAV was confirmed, we classified each as type 1 when right-left coronary cusp fusion (anteroposterior BAV with both coronary ostium at the anterior leaflet) was observed, type 2 for right-non-coronary cusp fusion (right-left BAV with right coronary ostium at the right leaflet and left coronary ostium at the left leaflet), or type 3 for left-non-coronary cusp fusion (left-non-coronary BAV with one ostium in each leaflet).<sup>3, 4</sup> Measurements were taken perpendicular to the axis of blood flow and included the largest aortic diameter. Aortic measurements were made from a 2D parasternal long-axis view at the following sites using the inner edge-to-inner edge convention: (1) aortic valve annulus, (2) maximal diameter of the sinuses of Valsalva, (3) sinotubular junction, and (4) maximal diameter of the proximal ascending aorta. The

presence of aortic regurgitation or stenosis was determined using Doppler echocardiography and was classified as mild, moderate, or severe according to published guidelines.<sup>5, 6</sup> Other echocardiographic measurements were performed as described previously.<sup>7</sup>

The pattern of aortic dimensions has been categorized in 3 aortic morphotype based on Z score  $\geq 2$  calculated using our references values for elite athletes<sup>7</sup> as appropriate. When Z score was  $>2$ , row aortic diameters in the 4 planes were evaluated to define the morphotype in case of aortic dilatation:

- no-dilatation morphotype
- root morphotype (dilated root with normal or less dilated ascending aorta)
- ascending morphotype (dilated ascending aorta with normal or less dilated root)

### Cardiopulmonary Testing

The cardiopulmonary testing procedure has been detailed elsewhere.<sup>7</sup> Briefly, athletes performed a maximal exercise test with an incremental protocol. Depending on the specific discipline of each athlete, the test was carried out on a treadmill, cycle, kayak, or rowing ergometer.

## Supplemental Material

**Suppl. Table 1.** Interobserver variability of aortic root size

	Intraclass correlation coefficient	CI 95%	P value
Aortic annulus (mm)	0.858	0.644 – 0.915	0.0001
Sinuses of Valsalva (mm)	0.908	0.814 – 0.948	0.0001
Sinotubular junction (mm)	0.930	0.874 – 0.964	0.0001
Proximal ascending aorta (mm)	0.842	0.724 – 0.916	0.0001

**Suppl. Table 2.** Dilated aortic root in BAV elite athletes according to Mitchell's sports classification based on dynamic components

	BAV elite athletes	Mitchell's sports classification based on dynamic components		
		Type A	Type B	Type C
	n (%)	n (%)	n (%)	n (%)
<b>Total</b>	41 (100)	9 (22)	8 (19.5)	24 (58.5)
<b>(A)</b> Aortic root or ascending aortic diameters measuring $\geq$ 40 mm form men and $\geq$ 36 mm for women	11 (26.8)	1 (11.1)	1 (12.5)	9 (37.5)
<b>(B)</b> Z score $\geq$ 2	18 (43.9)	4 (44.4)	3 (37.5)	11 (45.8)
Athletes with <b>(A)</b> or <b>(B)</b> criteria for dilated aorta	19 (46.3)	4 (44.4)	3 (37.5)	12 (50)
Aortic root morphotype	8 (19.5)	2 (22.2)	2 (25)	4 (16.7)
Ascending aorta mophotype	11 (26.8)	2 (22.2)	1 (12.5)	8 (33.3)
<b>Male</b>	34 (82.9)	6 (17.6)	8 (23.5)	20 (58.8)
<b>(A)</b> Aortic root or ascending aortic diameters measuring $\geq$ 40 mm	9 (26.5)	1 (16.7)	1 (12.5)	7 (35)
<b>(B)</b> Z score $\geq$ 2	14 (41.2)	2 (33.3)	3 (37.5)	9 (45)
Male Athletes with <b>(A)</b> or <b>(B)</b> criteria for dilated aorta	15 (44.1)	2 (33.3)	3 (37.5)	10 (50)
Aortic root morphotype	7 (20.6)	2 (33.3)	2 (25)	3 (15)
Ascending aorta mophotype	8 (23.5)	0	1 (12.5)	7 (35)
<b>Female</b>	7 (17.1)	3 (42.9)	0	4 (57.1)
<b>(A)</b> Aortic root or ascending aortic diameters measuring $\geq$ 36 mm	2 (28.6)	0	-	2 (50)
<b>(B)</b> Z score $\geq$ 2	4 (57.1)	2 (66.7)	-	2 (50)
Female Athletes with <b>(A)</b> or <b>(B)</b> criteria for dilated aorta	4 (57.1)	2 (66.7)	-	2 (50)
Aortic root morphotype	1 (14.3)	0	-	1 (25)
Ascending aorta mophotype	3 (42.9)	2 (66.7)	-	1 (25)

Data are presented as frequency and percentage. BAV: bicuspid aortic valve.

**Suppl. Table 3.** Baseline and follow-up aortic root dilatation in 16 BAV elite athletes according to Mitchell's sports classification based on dynamic components

	BAV elite athletes followed		Mitchell's sports classification based on dynamic components					
			Type A		Type B		Type C	
	n (%)		n (%)		n (%)		n (%)	
<b>Total</b>	16 (100)		2 (12.5)		3 (18.7)		11 (68.8)	
	Baseline	Follow-up	Baseline	Follow-up	Baseline	Follow-up	Baseline	Follow-up
(A) Aortic root or ascending aortic diameters measuring $\geq 40$ mm form men and $\geq 36$ mm for women	4 (25)	6 (37.5)	0	0	1 (33.3)	1 (33.3)	3 (27.3)	5 (45.4)
(B) Z score $\geq 2$	8 (50)	8 (50)	1 (50)	2 (100)	2 (66.7)	1 (33.3)	5 (45.4)	5 (45.4)
Athletes with (A) or (B) criteria for dilated aorta	8 (50)	8 (50)	1(50)	2 (100)	2 (66.7)	1 (33.3)	5 (45.4)	5 (45.4)
Aortic root morphotype	5 (31.2)	4 (25)	1(50)	1(50)	1 (33.3)	0	3 (27.3)	3 (27.3)
Ascending aorta mophotype	3 (18.7)	4 (25)	0	1(50)	1 (33.3)	1 (33.3)	2 (18.2)	2 (18.2)
<b>Male</b>	14 (87.5)		2 (14.3)		3 (21.4)		9 (64.3)	
	Baseline	Follow-up	Baseline	Follow-up	Baseline	Follow-up	Baseline	Follow-up
(A) Aortic root or ascending aortic diameters measuring $\geq 40$ mm	3 (21.4)	5 (35.7)	0	0	1 (33.3)	1 (33.3)	2 (22.2)	4 (44.4)
(B) Z score $\geq 2$	7(50)	7 (50)	1 (50)	2 (100)	2 (66.7)	1 (33.3)	4 (44.4)	4 (44.4)
Male Athletes with (A) or (B) criteria for dilated aorta	7(50)	7 (50)	1(50)	2 (100)	2 (66.7)	1 (33.3)	4 (44.4)	4 (44.4)
Aortic root morphotype	4 (28.6)	3 (21.4)	1 (50)	1 (50)	1 (33.3)	0	2 (22.2)	2 (22.2)
Ascending aorta mophotype	3 (21.4)	4 (28.6)	0	1 (50)	1 (33.3)	1 (33.3)	2 (22.2)	2 (22.2)
<b>Female</b>	2 (12.5)		0		0		2 (100)	
	Baseline	Follow-up	Baseline	Follow-up	Baseline	Follow-up	Baseline	Follow-up
(A) Aortic root or ascending aortic diameters measuring $\geq 36$ mm	1 (50)	1 (50)	-	-	-	-	1 (50)	1 (50)
(B) Z score $\geq 2$	1 (50)	1 (50)	-	-	-	-	1 (50)	1 (50)
Female Athletes with (A) or (B) criteria for dilated aorta	1 (50)	1 (50)	-	-	-	-	1 (50)	1 (50)
Aortic root morphotype	1 (50)	1 (50)	-	-	-	-	1 (50)	1 (50)
Ascending aorta mophotype	0	0	-	-	-	-	0	0

Data are presented as frequency and percentage. BAV: bicuspid aortic valve.

## Supplemental References

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