

Metadata list

**Population:** Children <18 years with bicuspid aortic valve and dilated ascending aorta ( $Z>3$ ).

**Overview of collected data:** Demographics, longitudinal echocardiographic data including LVOT measures, longitudinal data on medication, weight and height, outcome data related to LVOT.

**Status:** Data collection ongoing.

Field name	Field description	n	Range of value (categories)	Metadata (continuous)		
				Q1	Median	Q3
<b>SEX</b>	Sex	281	Male (74%) Female (23%) Unknown (3%)			
<b>Type of BAV</b>		281	Left and right coronary cusps fusion (66%) Right and none coronary cusps fusion (25%) True bicuspid aortic valve (no raphe) (1%) Unicuspid aortic or more than one raphe (2%) Unknown/unclear (5%)			
<b>Repaired coarctation</b>	Any personal history of repaired coarctation (surgical or percutaneous)?	277	Yes (4%) No (95%) Unknown (1%)			
<b>Family history of bicuspid aortic valve</b>	Any family history of bicuspid aortic valve in first or second degree family members?	280	Yes (8%) No (56%) Unknown (36%)			
<b>Family history of BAV-associated aortic</b>	Any family history of BAV-associated aortic dissection	280	Yes (2%) No (63%) Unknown (35%)			

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Field name	Field description	n	Range of value (categories)	Metadata (continuous)		
				Q1	Median	Q3
<b>dissection and aneurysm</b>	and aneurysm (first or second degree family members)?					
<b>Date of last clinic visit</b>	Last known to be alive (date of last clinic visit in cardiology with ascertainment of medication and echocardiography measurements)	275				
<b>ECHO. Measurement (repeated measurement for a single patient)</b>						
<b>Heart rate</b>	Heart rate recorded at the time of echocardiography or the closest clinic visit (less than 1 month apart)	1213		69	79	90
<b>Systolic blood pressure</b>	Systolic blood pressure recorded at the time of echocardiography or the closest clinic visit (less than 1 month apart)	1189		96	105	115
<b>Diastolic blood pressure</b>	Diastolic blood pressure recorded at the time of echocardiography or the closest clinic visit (less than 1 month apart)	1169		59	65	71

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Field name	Field description	n	Range of value (categories)	Metadata (continuous)		
				Q1	Median	Q3
<b>Weight</b>	Weight (in kg) at the time of echocardiography	1280		21.0	35.6	57.0
<b>Height</b>	Height (in cm) at the time of echocardiography	1246		117,0	142.0	165.0
<b>Aortic annulus diameter</b>	(mm)	1014		17,1	20,0	23,0
<b>Sinus of Valsalva diameter</b>	(mm)	1137		22,4	26,7	30,4
<b>Ascending aorta diameter</b>	Ascending aorta diameter (mm) taken at the level of the RPA	1286		25,0	30,0	34,0
<b>Maximal recorded gradient</b>	Maximal recorded gradient at the level of the aortic valve by continuous Doppler (mmHg)	1214		16,0	27,0	45,7
<b>Aortic regurgitation</b>	Aortic regurgitation (record the WORST regurgitation in the report. If mentioned mild to moderate, choose moderate)	1418	None (or 0 / IV) (15%) Trivial (or I / IV) (31%) Mild (or II / IV) (33%) Moderate (or III / IV) (18%) Severe (or IV / IV) (2%) Unknown (1%)			
<b>Left ventricular shorting fraction</b>	(%)	1243		35,0	39,0	43,0

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				Q1	Median	Q3
<b>Left ventricular ejection fraction</b>	(%)	1021		65,0	70,0	74,6
<b>Left ventricular function</b>	Left ventricular function by qualitative evaluation	1406	Normal (99%) Mildly reduced (<1%) Moderately reduced (<1%) Severely reduced (<1%) Unknown (<1%)			
<b>Medication</b>	Is the participant on any medication at the time of or nearing the current echocardiogram ?	1422	Yes (7%) No (88%) Unknown (5%)			
<b>2<sup>nd</sup> medication</b>	Is there a 2 <sup>nd</sup> medication?	99	Yes (6%) No (92%) Unknown (2%)			
<b>3<sup>rd</sup> medication</b>	Is there a 3 <sup>rd</sup> medication?	6	Yes (33%) No (67%) Unknown (0%)			
<b>4<sup>th</sup> medication</b>	Is there a 4 <sup>th</sup> medication?	2	Yes (50%) No (50%) Unknown (0%)			
<b>5<sup>th</sup> medication</b>	Is there a 5 <sup>th</sup> medication?	1	Yes (0%) No (100%) Unknown (0%)			
<b>Aortic Intervention follow-up</b>	Any aortic valve intervention during follow-up (surgery or percutaneous)?	275	Yes (7%) No (93%) Unknown/unclear (<1%)			

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				Q1	Median	Q3
<b>Date of aortic valve intervention</b>		18				
<b>Aortic root or ascending aorta intervention</b>	Any aortic root or ascending aorta intervention?	275	Yes (3%) No (96%) Unknown / unclear (1%)			
<b>Date aortic root or ascending aorta intervention</b>		7				
<b>Aortic dissection follow-up</b>	Any aortic dissection during follow-up?	275	Yes (0%) No (99%) Unknown / unclear (<1%)			
<b>Date of aortic dissection</b>		0				
<b>Circumstances around aortic dissection</b>	Please describe circumstances around aortic dissection	0				
<b>Surgery for aortic dissection</b>	Any surgery for aortic dissection?	0				
<b>Date of aortic dissection surgery</b>		0				
<b>Date of death</b>	Date of death (if applicable)	0				