

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: Analysis ongoing.

Field name	Field description	n	Range of value (categories)	Metadata (continuous)		
				Q1	Median	Q3
SEX	Sex	579	Male (77%) Female (21%) Unknown (2%)			
Type of BAV		577	Left and right coronary cusps fusion (56%) Right and none coronary cusps fusion (28%) True bicuspid aortic valve (no raphe) (2%) Unicuspid aortic or more than one raphe (5%) Unknown/unclear (9%)			
Repaired coarctation	Any personal history of repaired coarctation (surgical or percutaneous)?	575	Yes (4%) No (95%) Unknown (1%)			
Family history of bicuspid aortic valve	Any family history of bicuspid aortic valve in first or second degree family members?	579	Yes (10%) No (60%) Unknown (30%)			
Family history of BAV-associated aortic	Any family history of BAV-associated aortic dissection	579	Yes (2%) No (63%) Unknown (35%)			

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				Q1	Median	Q3
dissection and aneurysm	and aneurysm (first or second degree family members)?					
Last known to be alive	Last known to be alive (date of last clinic visit in cardiology with ascertainment of medication and echocardiography measurements)	577				
ECHO. Measurement (repeated measurement for a single patient)						
Heart rate	Heart rate recorded at the time of echocardiography or the closest clinic visit (less than 1 month apart)	2850		69	79	90
Systolic blood pressure	Systolic blood pressure recorded at the time of echocardiography or the closest clinic visit (less than 1 month apart)	2842		96	105	115
Diastolic blood pressure	Diastolic blood pressure recorded at the time of echocardiography or the closest clinic visit (less than 1 month apart)	2819		59	65	71

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Field name	Field description	n	Range of value (categories)	Metadata (continuous)		
				Q1	Median	Q3
Weight	Weight (in kg) at the time of echocardiography	3029		21.0	35.6	57.0
Height	Height (in cm) at the time of echocardiography	2996		117,0	142.0	165.0
Aortic annulus diameter	(mm)	2518		17,1	20,0	23,0
Sinus of Valsalva diameter	(mm)	2994		22,4	26,7	30,4
Ascending aorta diameter	Ascending aorta diameter (mm) taken at the level of the RPA	2994		25,0	30,0	34,0
Maximal recorded gradient	Maximal recorded gradient at the level of the aortic valve by continuous Doppler (mmHg)	2746		16,0	27,0	45,7
Aortic regurgitation	Aortic regurgitation (record the WORST regurgitation in the report. If mentioned mild to moderate, choose moderate)	3161	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%)			
Left ventricular shorting fraction	(%)	2946		35,0	39,0	43,0

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				Q1	Median	Q3
Left ventricular ejection fraction	(%)	2529		65,0	70,0	74,6
Left ventricular function	Left ventricular function by qualitative evaluation	3155	Normal (99%) Mildly reduced (<1%) Moderately reduced (<1%) Severely reduced (<1%) Unknown (<1%)			
Medication	Is the participant on any medication at the time of or nearing the current echocardiogram ?	3171	Yes (7%) No (88%) Unknown (5%)			
Aortic Intervention follow-up	Any aortic valve intervention during follow-up (surgery or percutaneous)?	275	Yes (7%) No (93%) Unknown/unclear (<1%)			