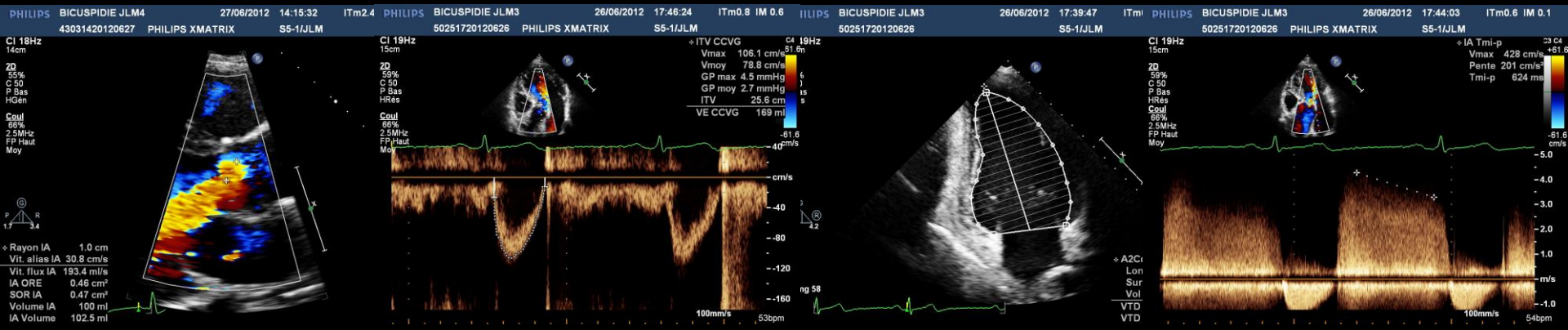
Bicuspid aortic valves : Valvular and aortic issues



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Disclosures related to this talk:

none

What are the prevalence and Clinical implications of Bicuspid AV ?

- Bicuspid AV : most common congenital cardiac malformation, occurring in 0.7-2 % of the population (males are affected 4:1)
- BAV is present in >50% of patients with aortic coarctation and 10% of women with Turner syndrome
- The majority of patients with BAV will develop complications requiring surgery (mostly in their 4th to 6th decades)
- BAV might be responsible for more deaths and morbidity than the combined effects of all other congenital heart defects

Fedak et al. *Circulation*. 2002; 106:900-4

Diagnosis of a BAV: When and how?

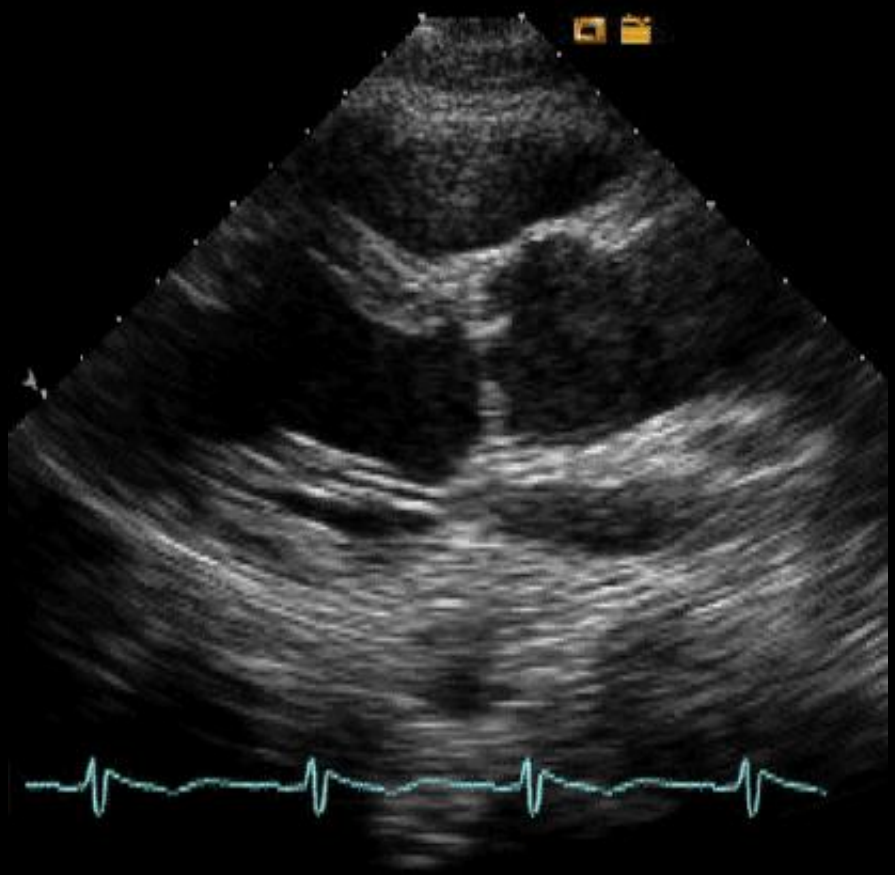
When to suspect a bicuspid aortic valve ?

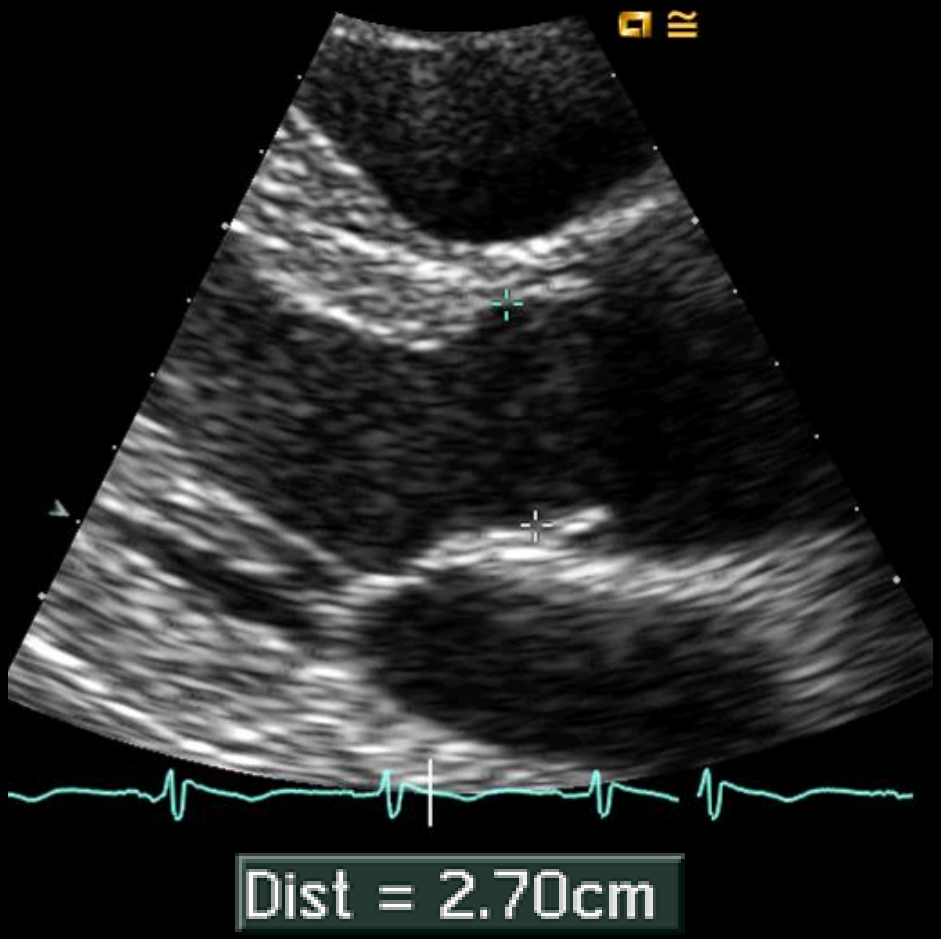
- Coarctation of the aorta
- Cardiac murmur in adolescent / young adult
- Aortic dilatation/ regurgitation <60 years
- Aortic valve stenosis in the 4th to 6th decade
- First relative of a patient with a bicuspid valve
- Turner syndrome

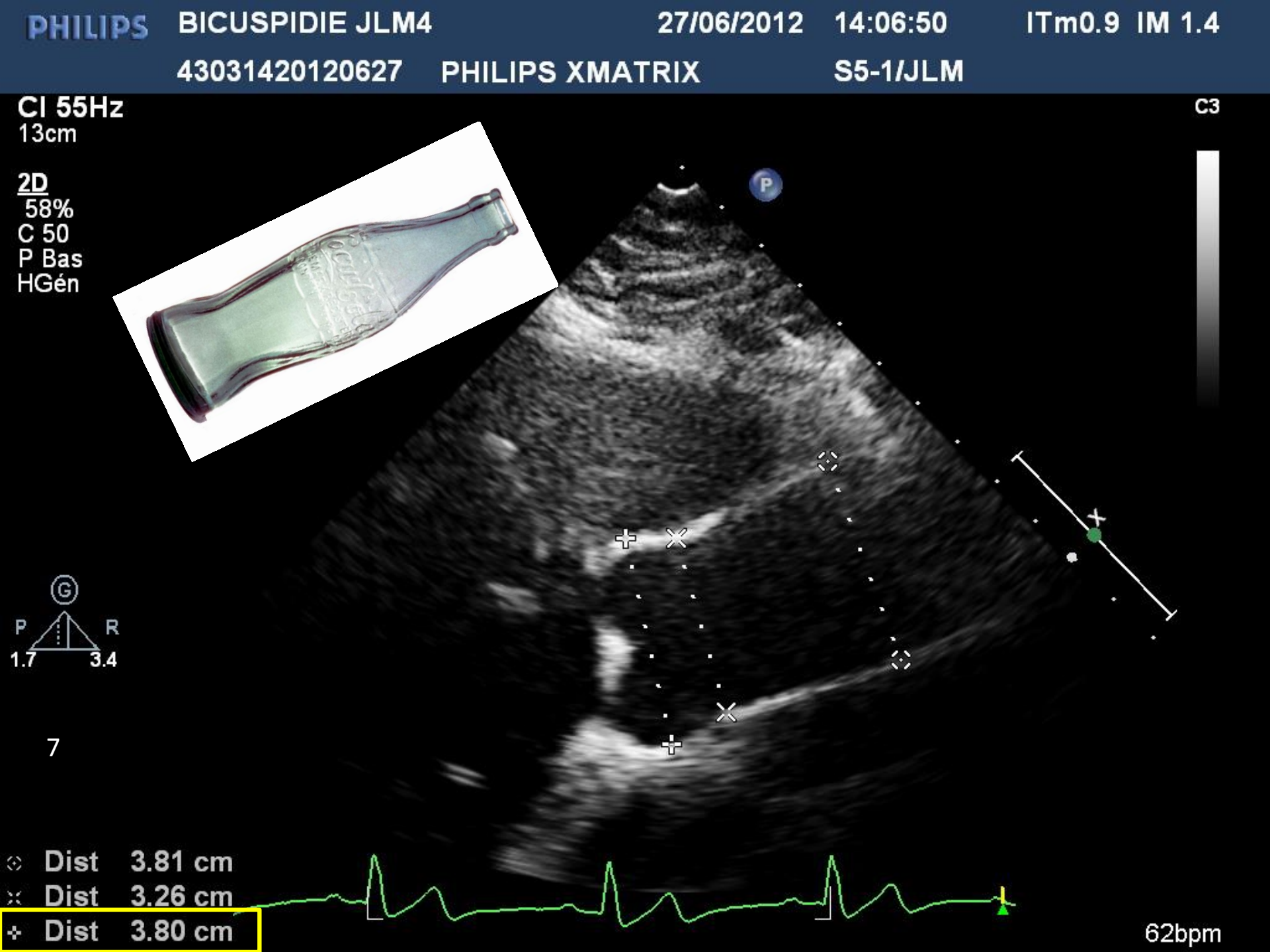
How to diagnose a bicuspid valve ?

Male patient, 48 years

1/ Deformed aortic Root; 2/ Asymmetric opening; 3/ Large ring



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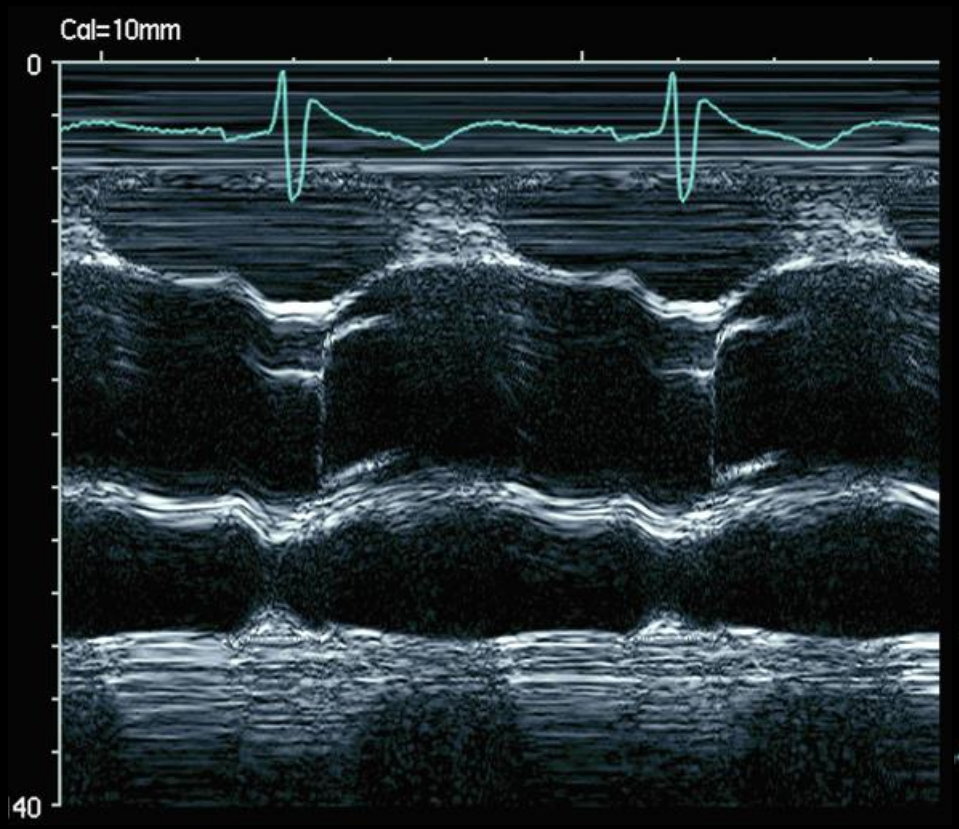
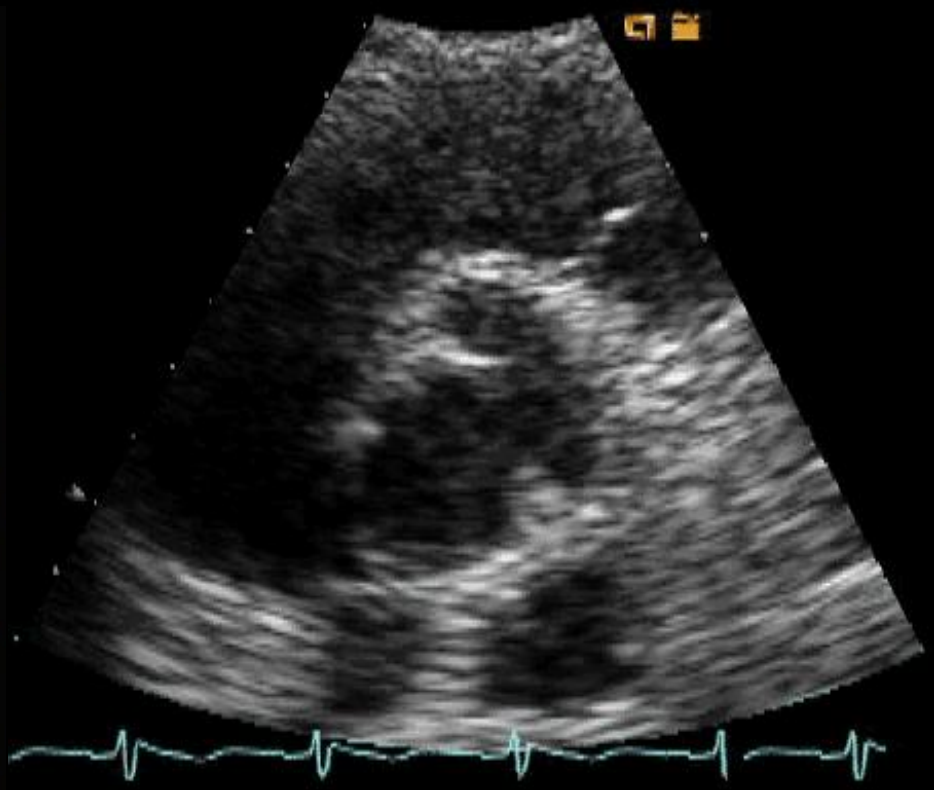
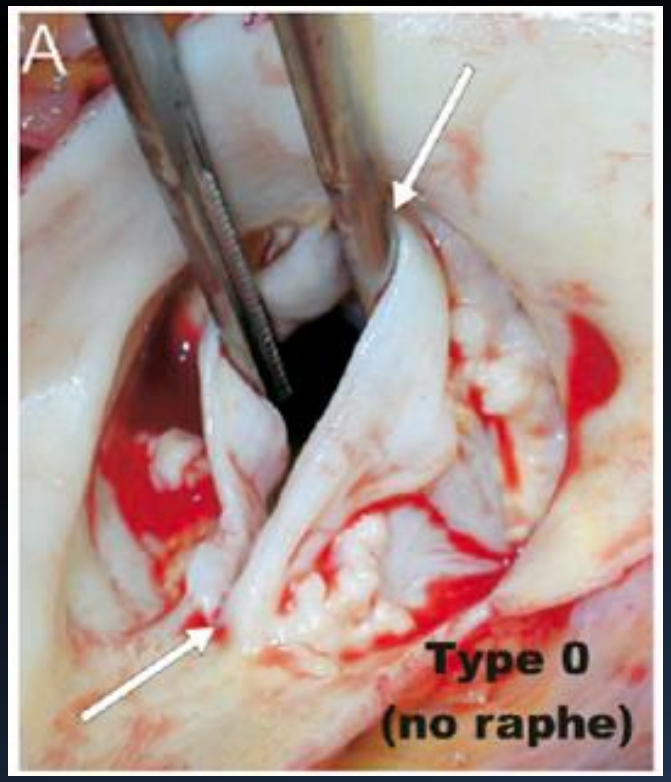
Dist 3.81 cm
Dist 3.26 cm
Dist 3.80 cm
62bpm

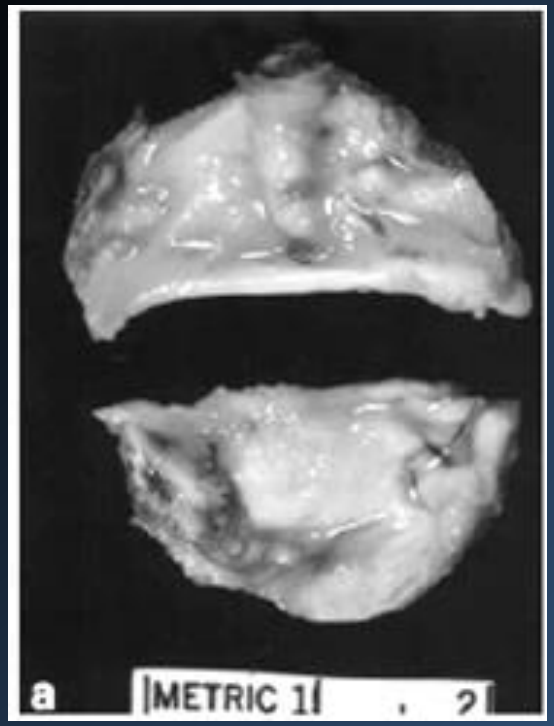
En-face view: No raphe (Type 0)





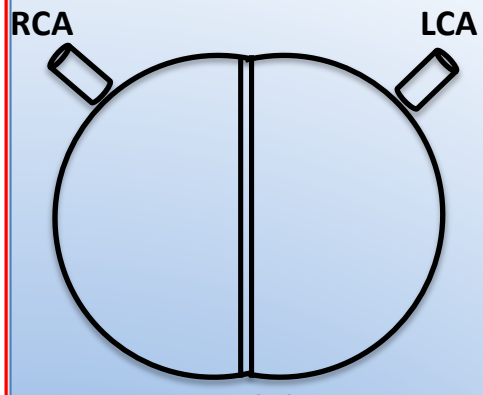
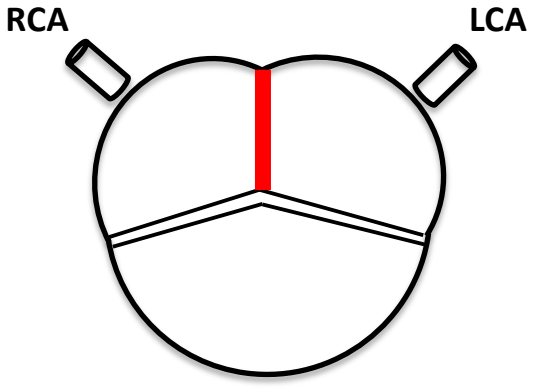
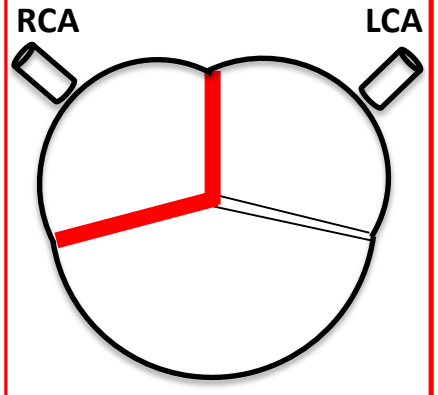
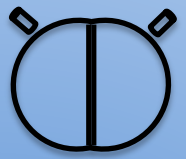
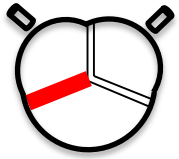
Bicuspid aortic valve/ Type 0: Two Sinuses of Valsalva / 2 cusps





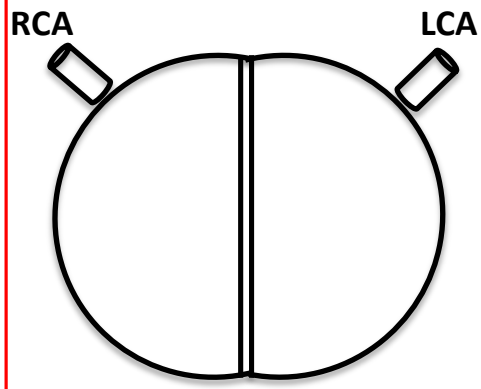
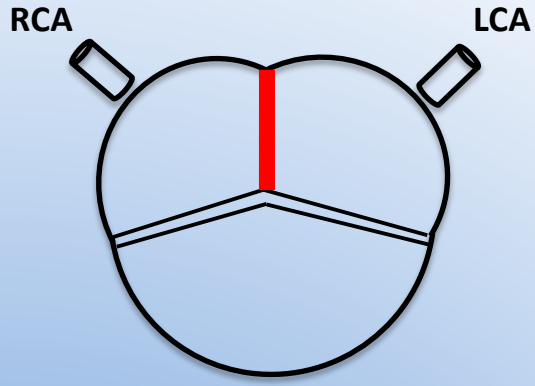
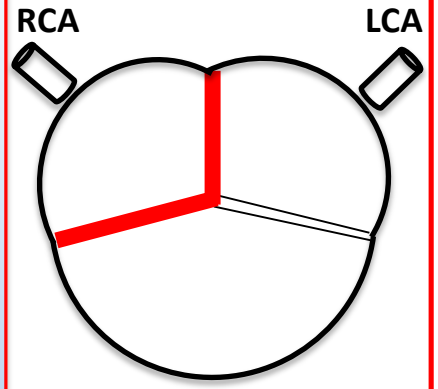
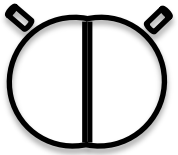
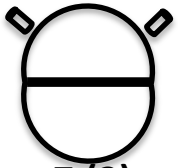
Type O: 6% of bicuspid valves

Roberts & Ko. *Circulation*. 2005;111: 920-925

Main category: Number of raphes	0 raphe / Type 0 True bicuspid		1 raphe / Type 1 (Bicommisural)			2 raphes / Type 2 (Unicommissural)
	 21 (7)		 269 (88)			 14 (5)
1. subcategory: spatial position of cusps in Type 0 and raphes in Types 1 and 2	Lat  13 (4)	AP  7 (2)	LR  216 (71)	NR  45 (15)	NL  8 (3)	NR-LR  14 (5)
2. subcategory: Valvular function	I	6 (2)	1 (0.3)	79 (26)	22 (7)	3 (1)	6 (2)
	S	7 (2)	5 (2)	119 (39)	15 (5)	3 (1)	2 (1)
	B (I + S)		1 (0.3)	15 (5)	7 (2)	2 (1)	2 (1)
	No			3 (1)	1 (0.3)		

Adapted from Sievers & Schmidtke. *J Thorac Cardiovasc Surg*. 2007; 133:1226-33

Type 1/LR vs. NR: Does it matter?

Main category: Number of raphes	0 raphe / Type 0 True bicuspid		1 raphe / Type 1 (Bicommisural)			2 raphes / Type 2 (Unicommissural)
	 21 (7)		 269 (88)			 14 (5)
1. subcategory: spatial position of cusps in Type 0 and raphes in Types 1 and 2	Lat  13 (4)	AP  7 (2)	LR  216 (71)	NR  45 (15)	NL  8 (3)	NR-LR  14 (5)
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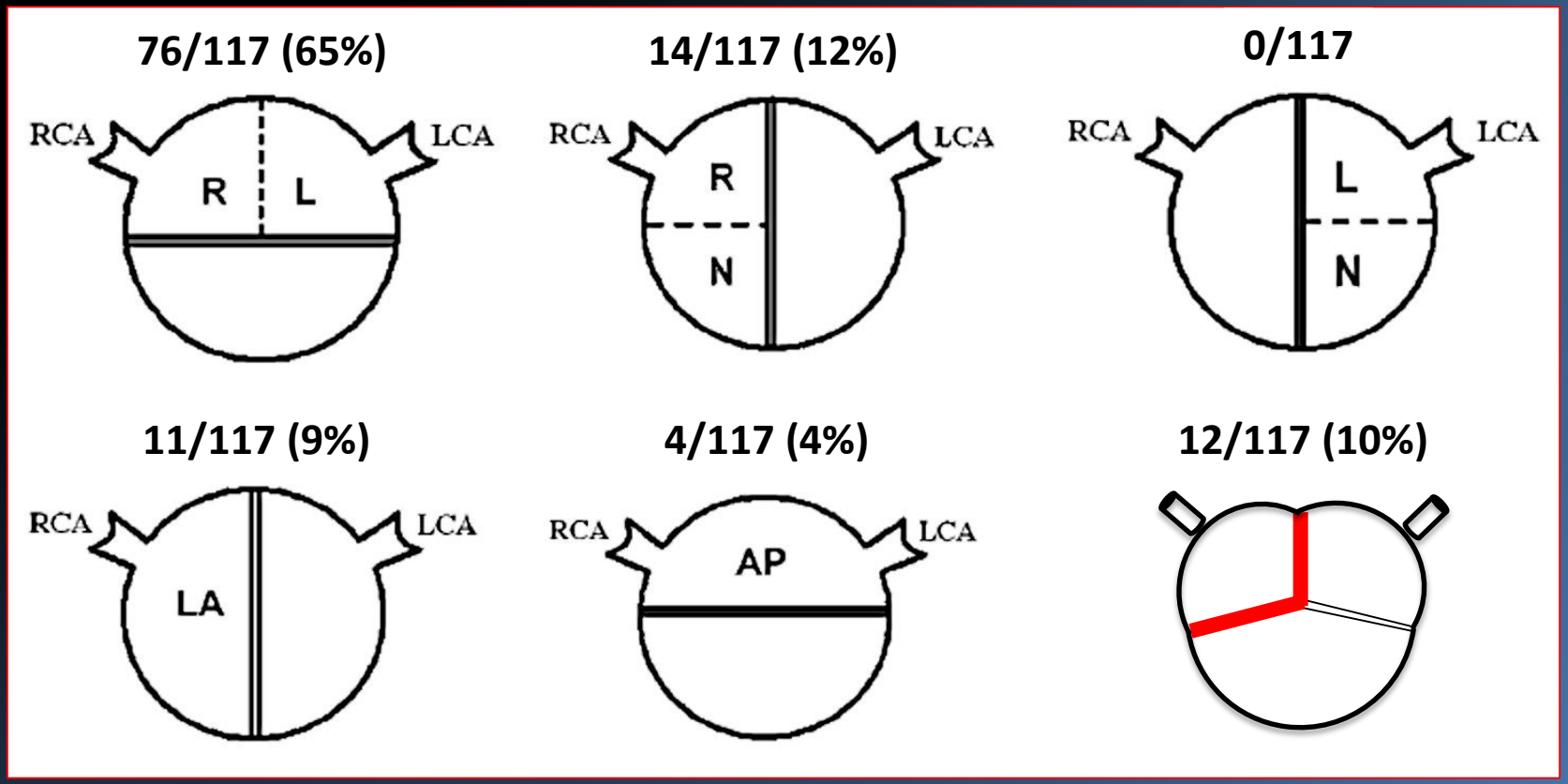
	S	7 (2)	5 (2)	119 (39)	15 (5)	3 (1)	2 (1)
	B (I + S)		1 (0.3)	15 (5)	7 (2)	2 (1)	2 (1)
	No			3 (1)	1 (0.3)		

Adapted from Sievers & Schmidtke. *J Thorac Cardiovasc Surg*. 2007; 133:1226-33

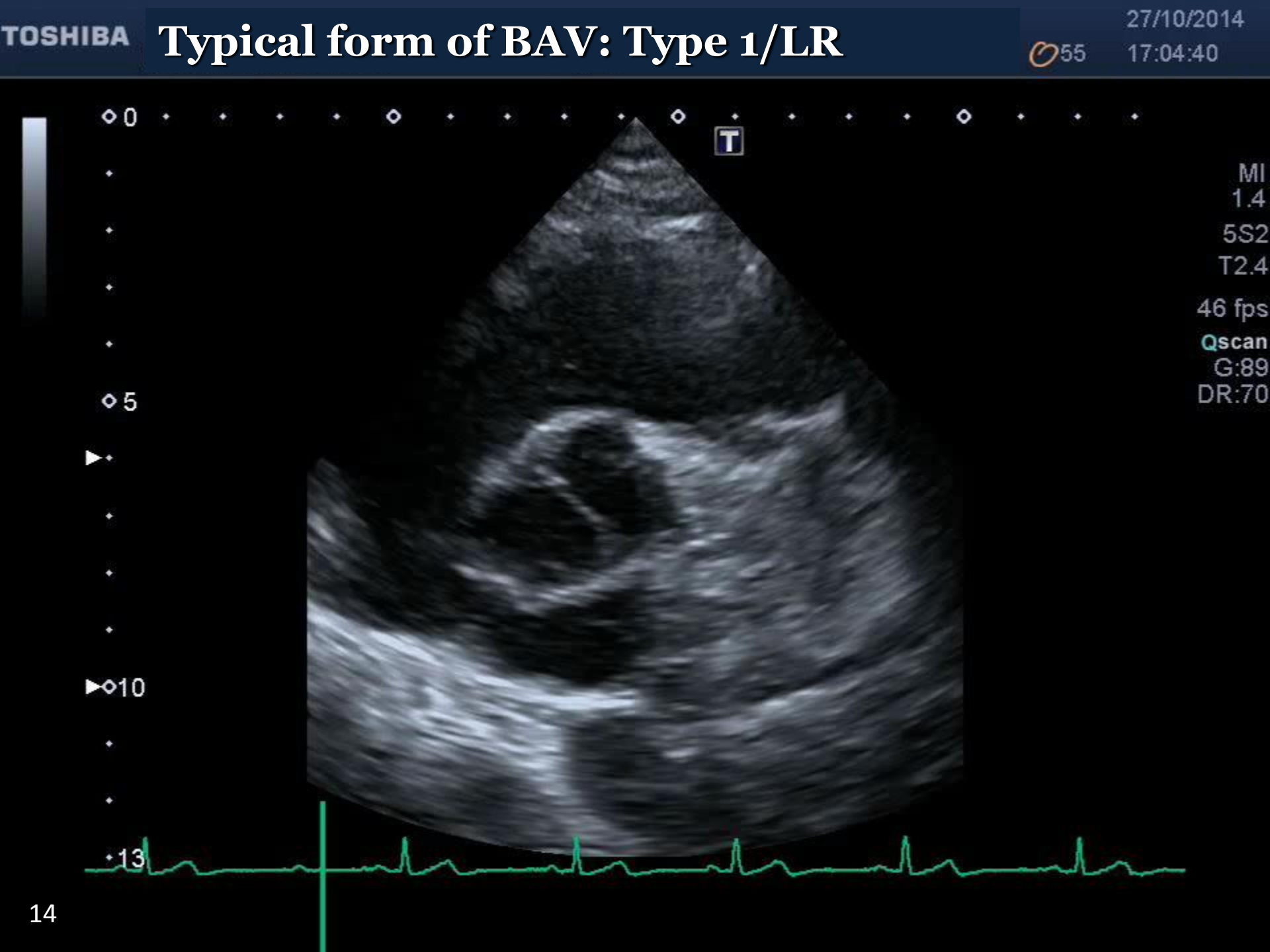
Variable phenotypes of bicuspid aortic valves : classification by CMR

Retrospective study (2003-2007): 105 patients with a bicuspid valve plus 12 with a unicuspid valve, all assessed by CMR



Adapted from Buchner et al. *Heart.* 2010;96:1233e1240

Typical form of BAV: Type 1/LR



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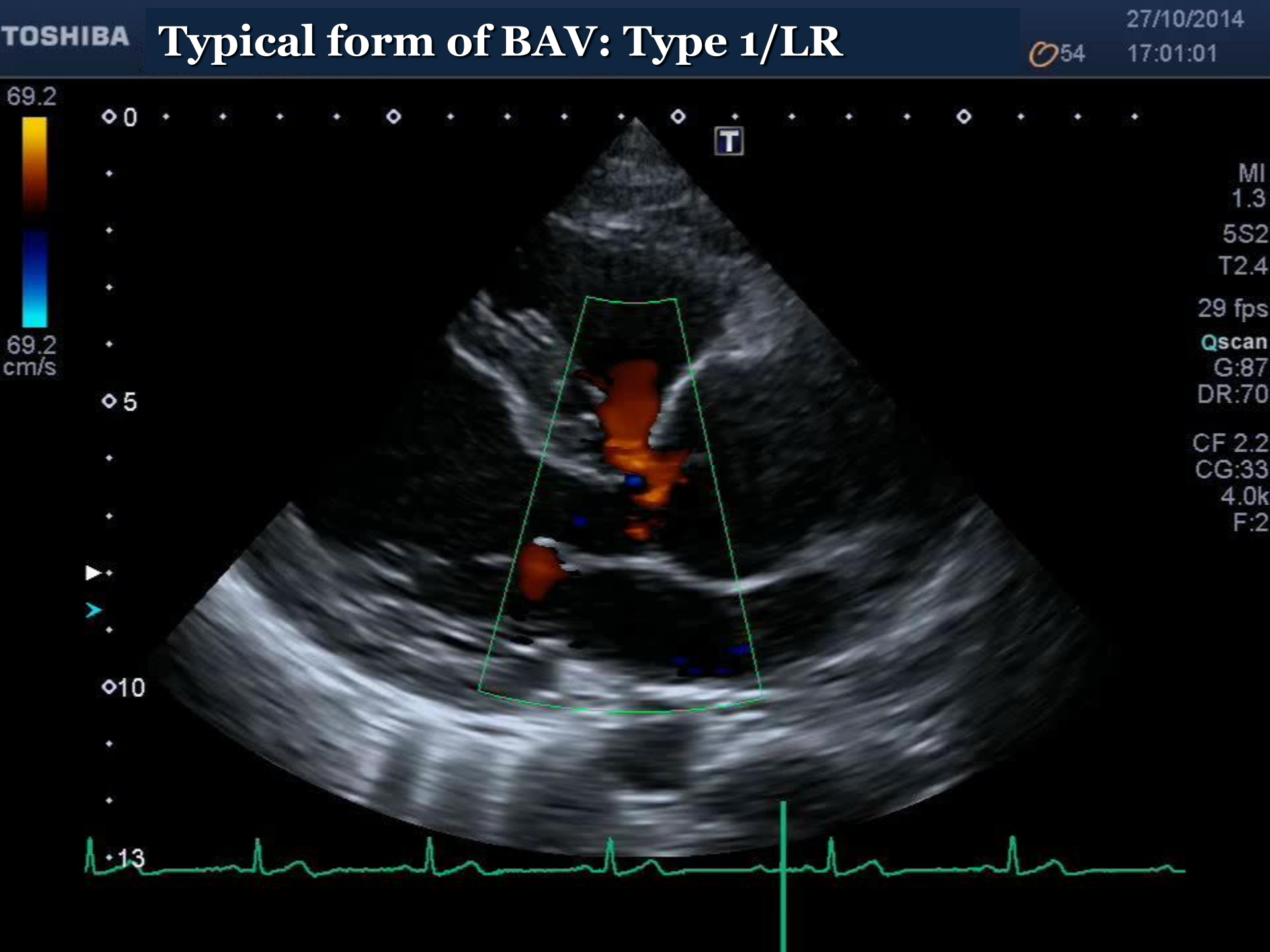
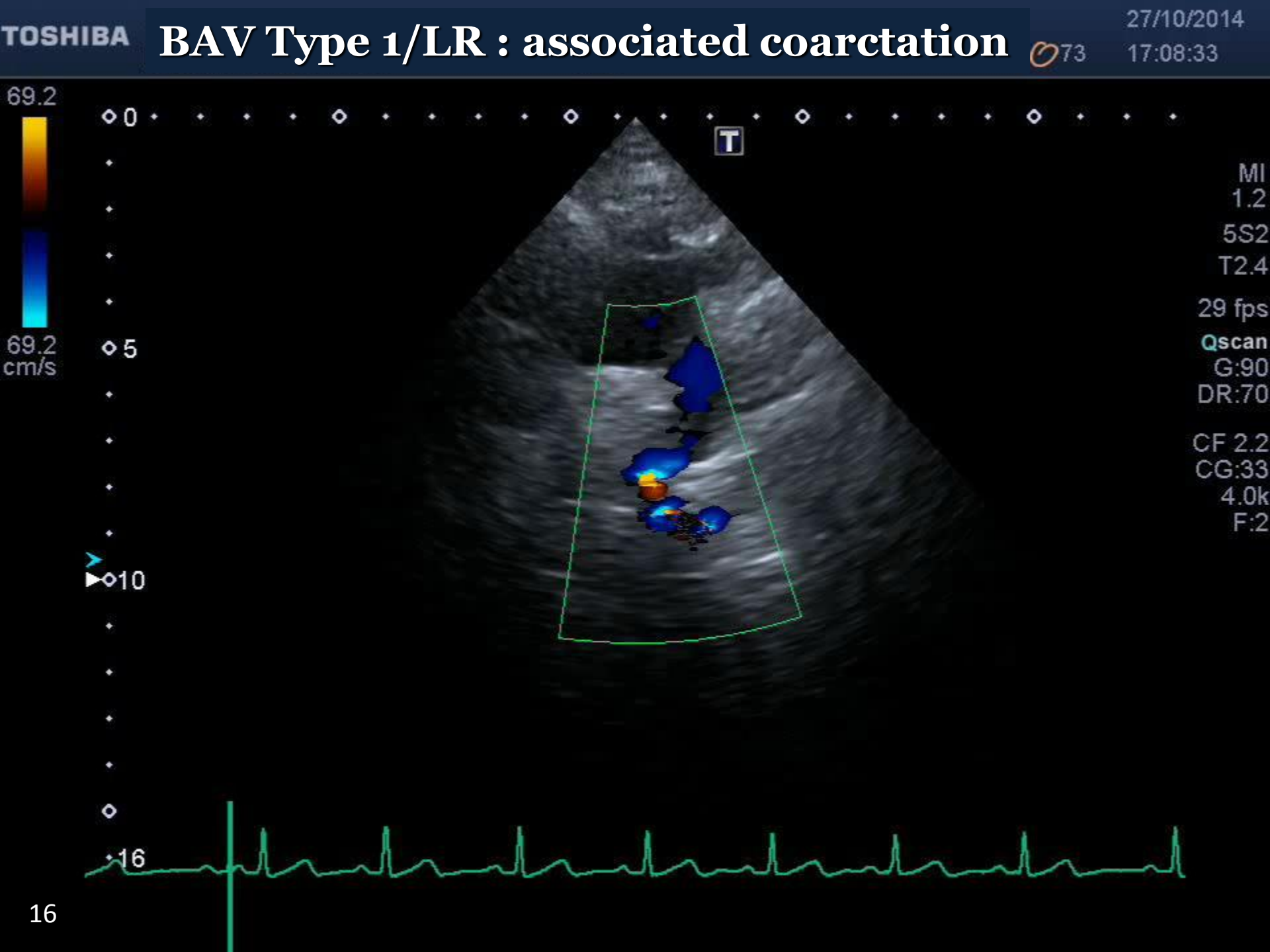
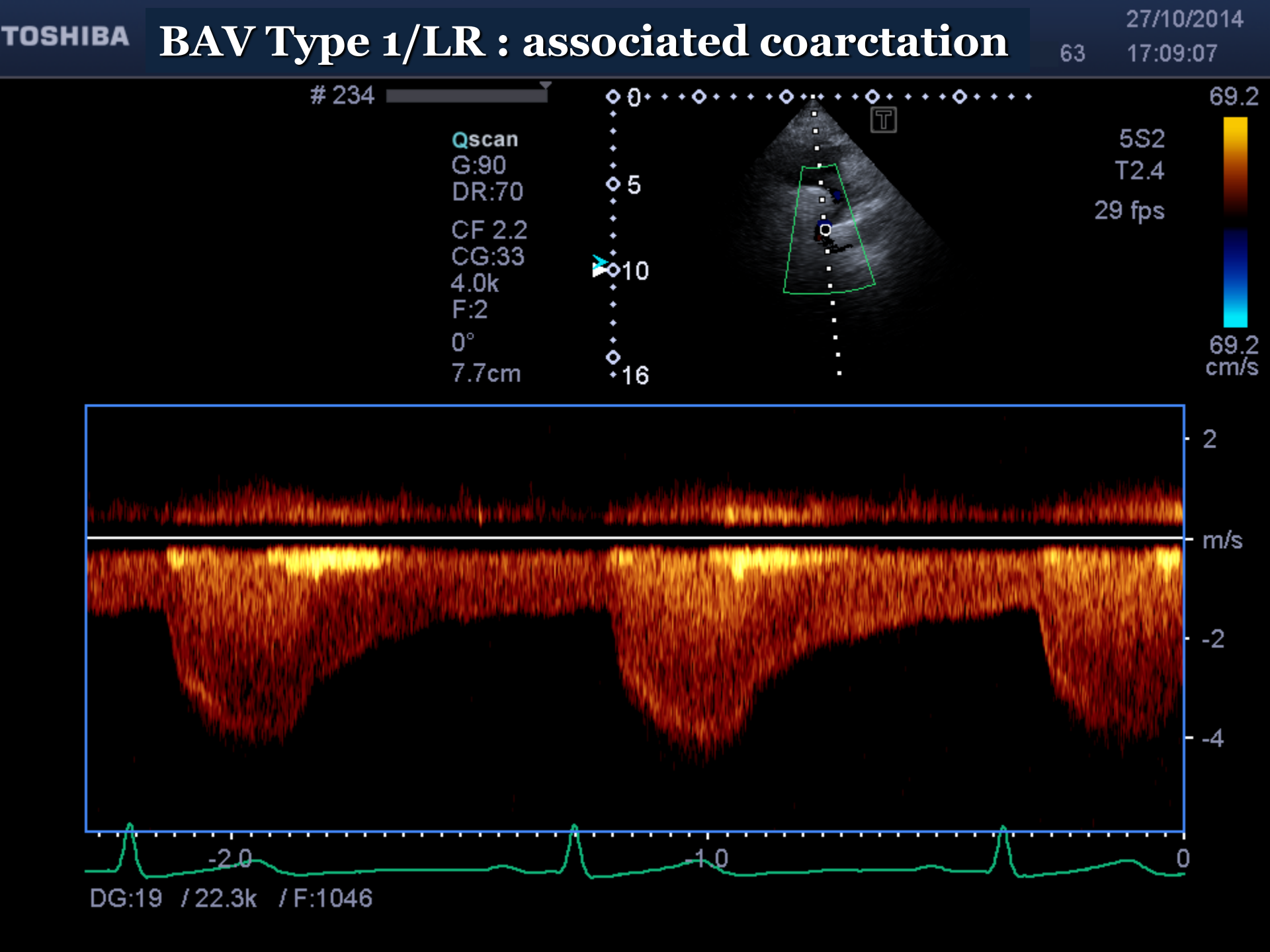


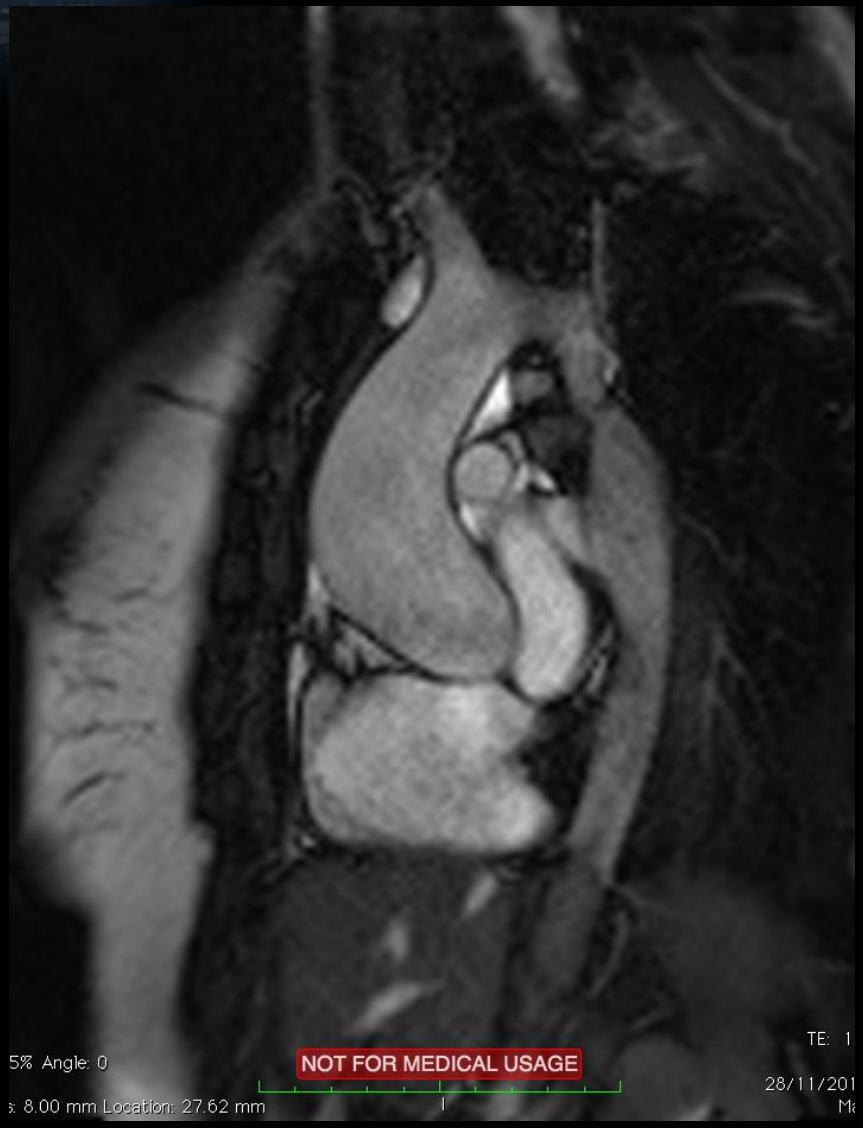
BAV Type 1/LR : associated coarctation

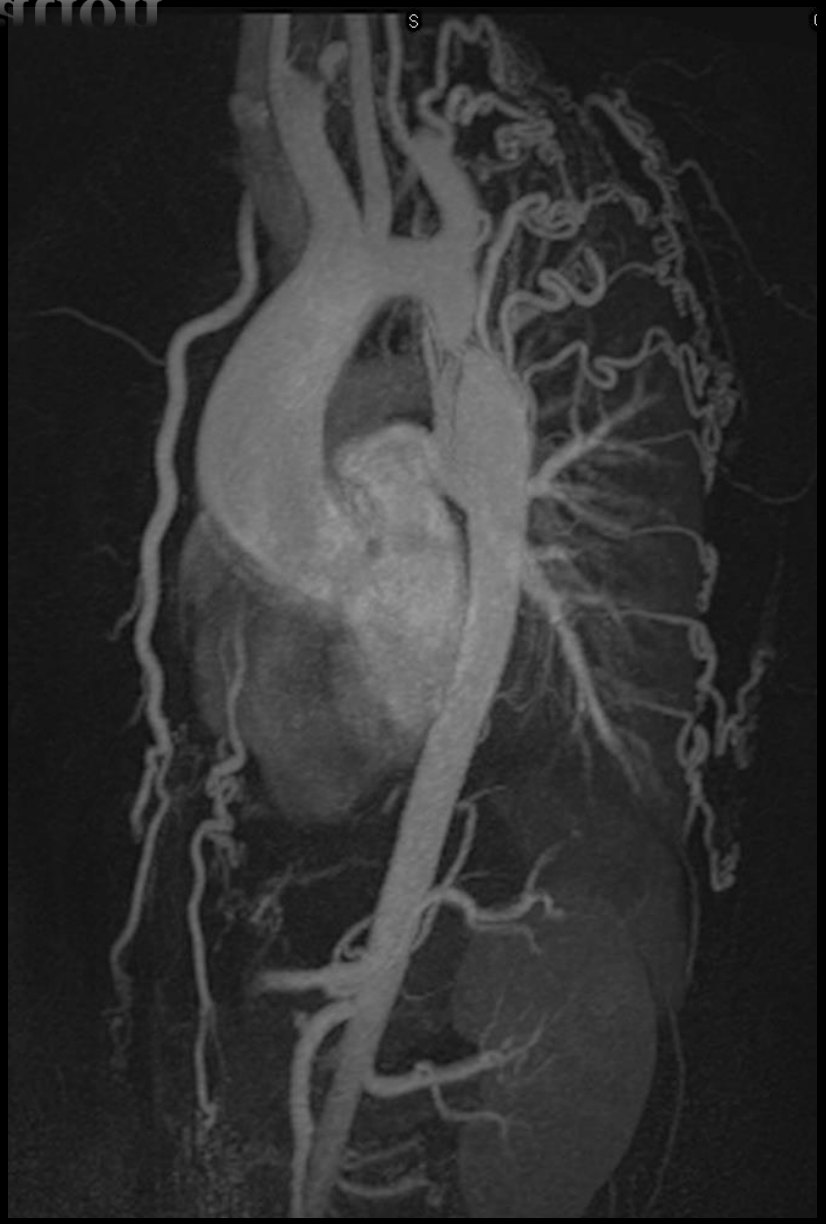


BAV Type 1/LR : associated coarctation



Asymptomatic young woman, 23 years old
BAV type 1/LR + coarctation





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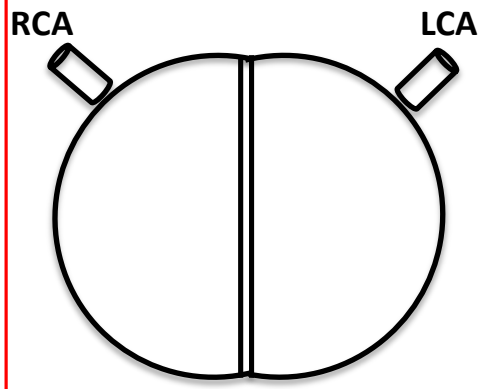
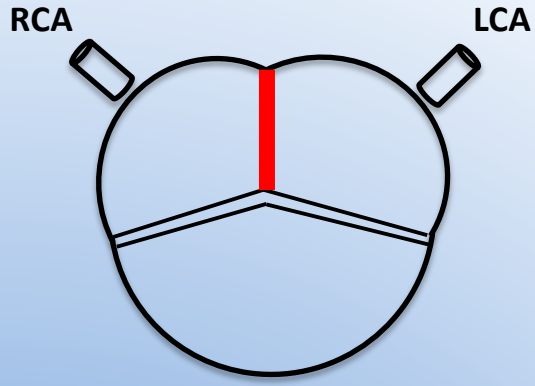
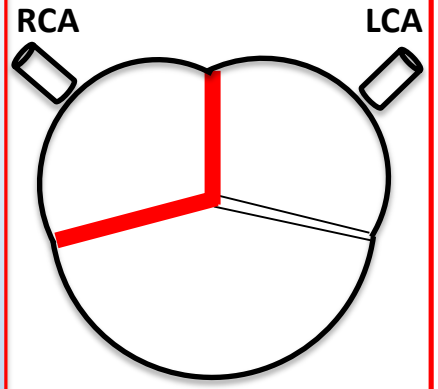
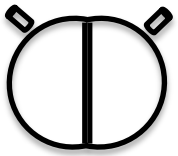
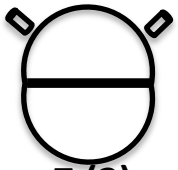
	 21 (7)		 269 (88)			 14 (5)
1. subcategory: spatial position of cusps in Type 0 and raphes in Types 1 and 2	Lat  13 (4)	AP  7 (2)	LR  216 (71)	NR  45 (15)	NL  8 (3)	NR-LR  14 (5)
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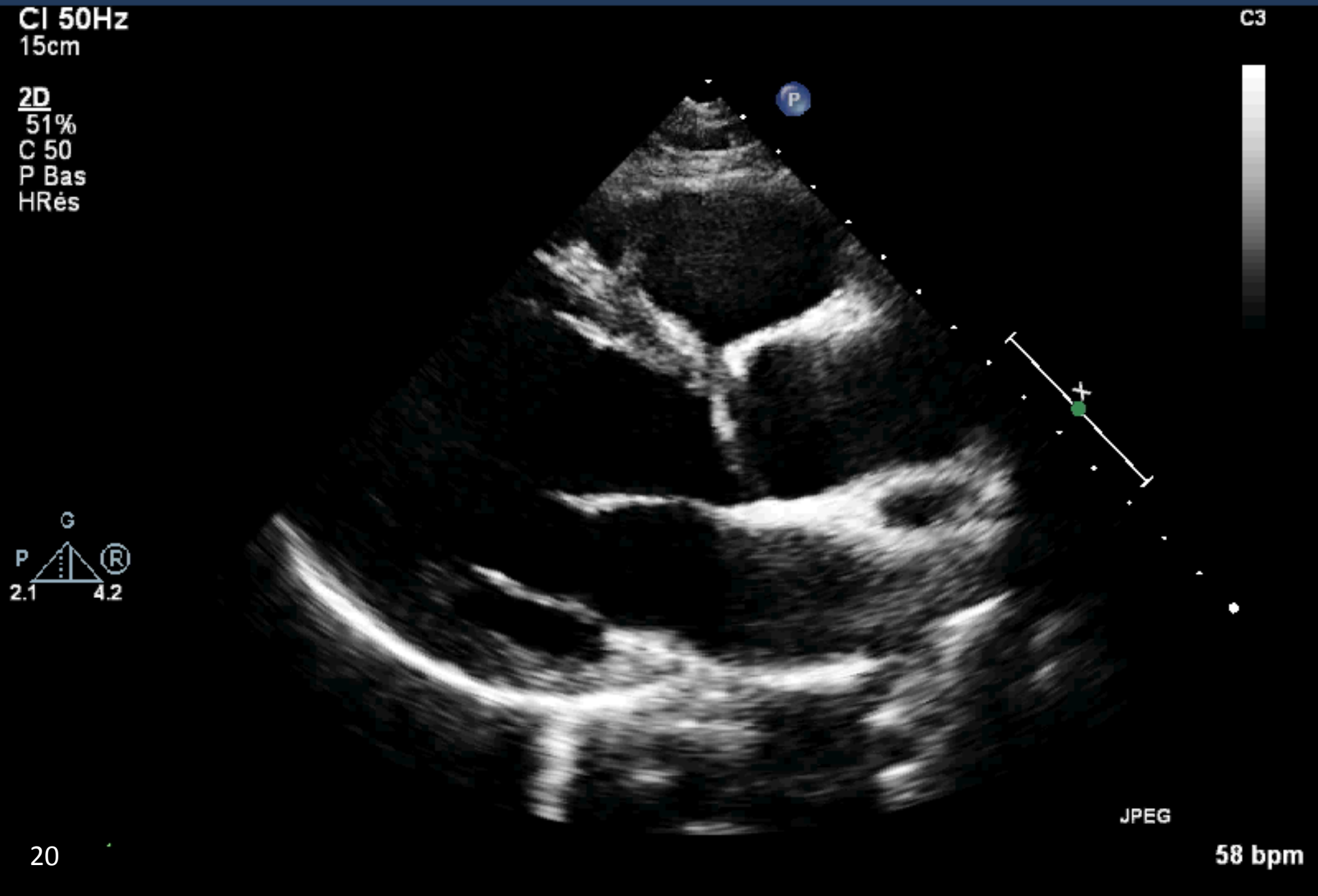
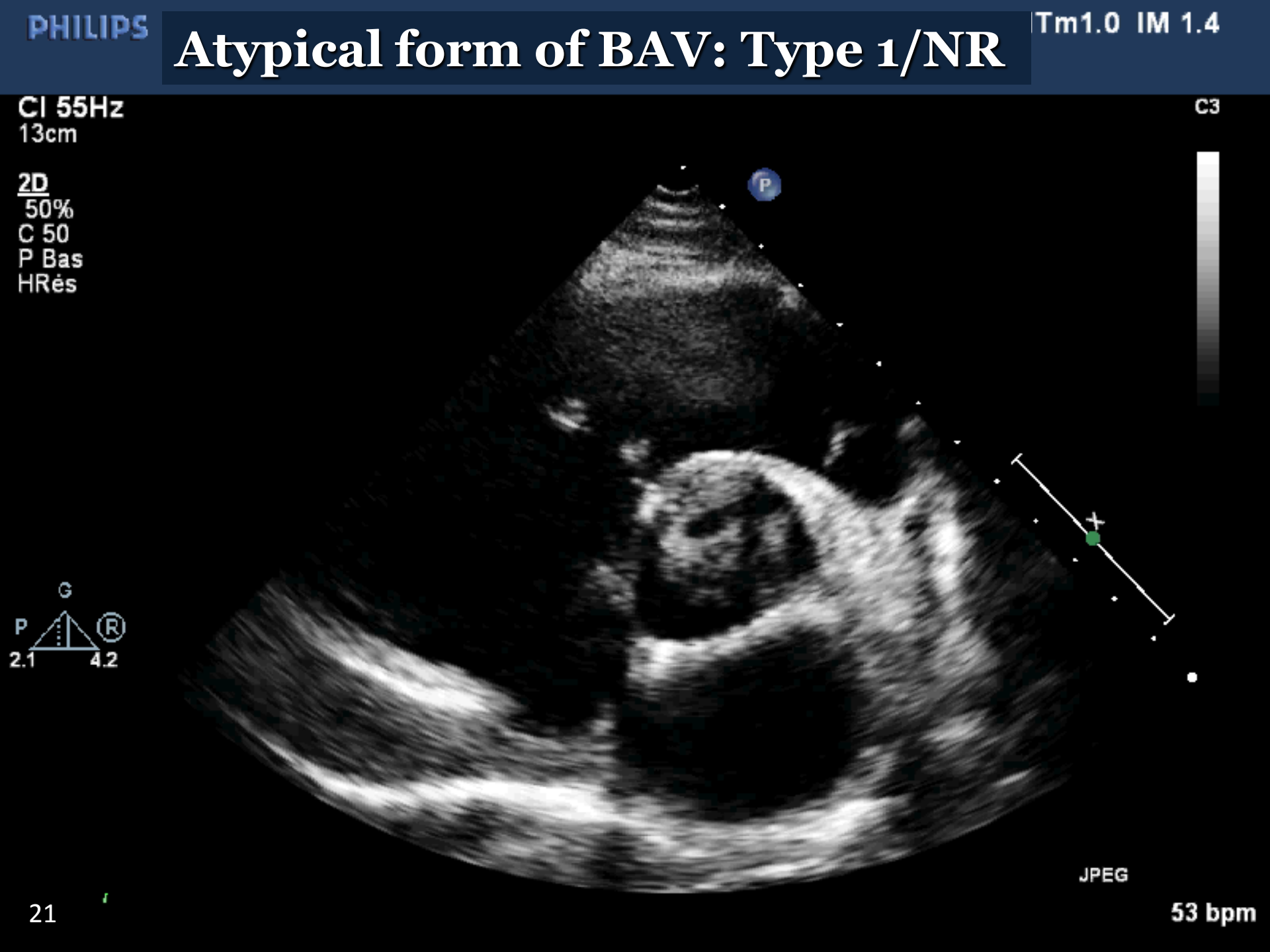
	No			3 (1)	1 (0.3)		

Adapted from Sievers & Schmidtke. *J Thorac Cardiovasc Surg*. 2007; 133:1226-33

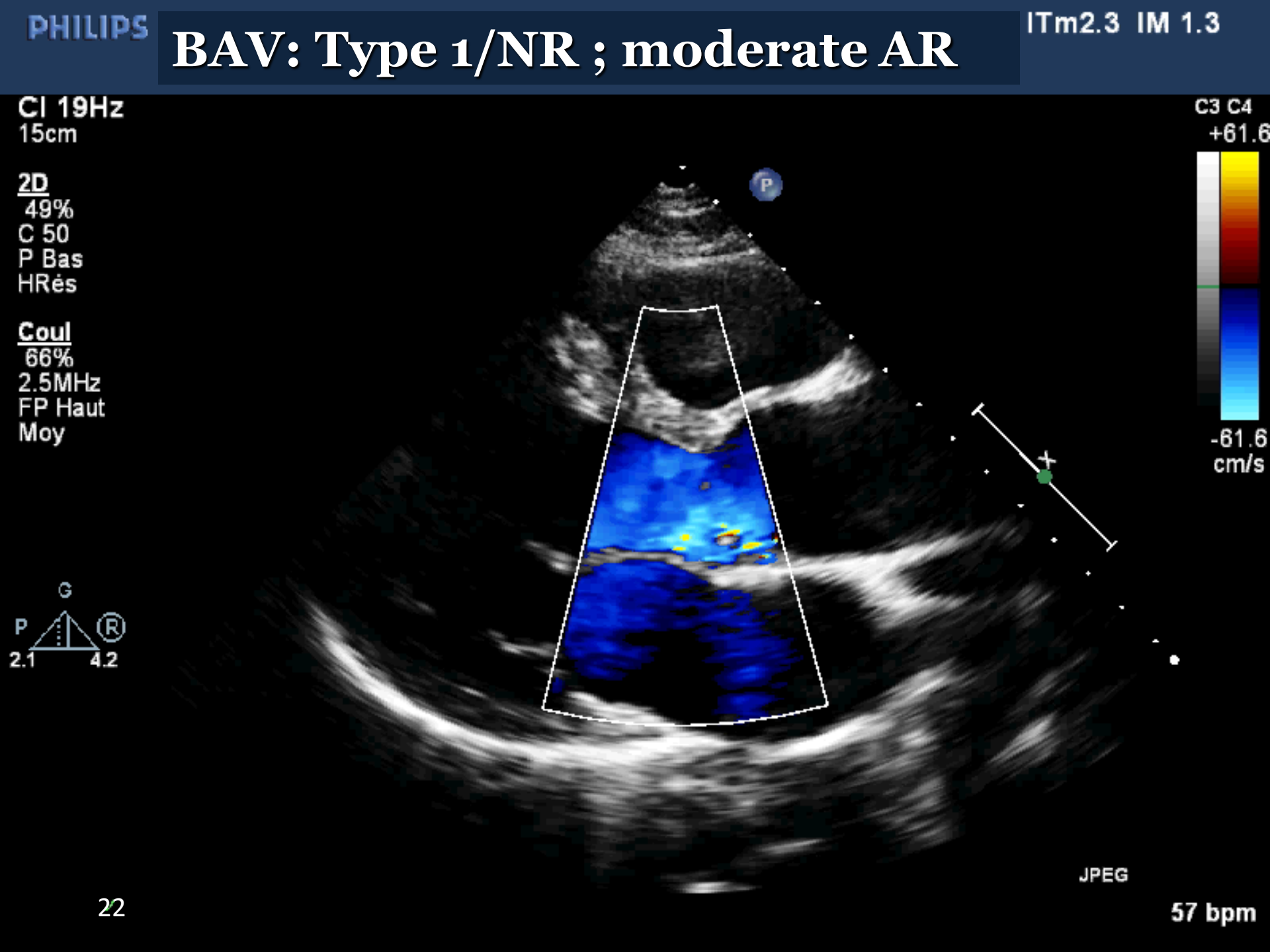
Asymptomatic young male, 34 years old



Atypical form of BAV: Type 1/NR



BAV: Type 1/NR ; moderate AR



Morphology of bicuspid aortic valves in children and adolescents

- Retrospective study: 1 135 patients <18 years (1986-1999) with BAV identified by Echo
- 55% of patients with isolated coarctation had a bicuspid valve
- 40% of patients with a BAV had a coarctation in this study (vs.7-25% in others)

Table 1. Bicuspid Aortic Valve and Associated Congenital Cardiac Malformations

Associated Malformation	Number With Malformation	Number With BAV	Percent With BAV (95% Confidence Interval)
Isolated coarctation	835	459 (55%)	55 (51.5, 58.4)
Complex coarctation	629	111	17.6 (14.7, 20.9)
HLHS or IAA	570	64 (11%)	11.2 (8.8, 14.1)
CAVC defect	1,074	11	1.0 (0.5, 1.8)
Ebstein's anomaly	250	2	0.8 (0.1, 2.8)
TAPVR	247	2	0.8 (0.1, 2.9)
PAPVR	233	2	0.9 (0.1, 3.1)
Tetralogy of Fallot	1,213	7	0.6 (0.2, 1.2)
DORV	773	5	0.6 (0.2, 1.5)
TGA	1,567	1	0.1 (0.0, 0.4)

BAV = bicuspid aortic valve; CAVC = complete atrioventricular canal defect; DORV = double-outlet right ventricle; HLHS = hypoplastic left heart syndrome; IAA = interrupted aortic arch; PAPVR = partial anomalous pulmonary venous return; TAPVR = total anomalous pulmonary venous return; TGA = transposition of the great arteries.

Fernandes et al. *J Am Coll Cardiol.* 2004;44:1648-51

Morphology of bicuspid aortic valves in children and adolescents

- Associated BAV + coarctation: 89% of typical BAV (Type 1/LR, Sievers)
- Aortic coarctation is associated with lesser degrees of AS or AR
- In contrast: Type 1/NR was more often associated with significant AS or AR (OR 2.4, 95% CI: 1.2-4.7; p 0.01)

Table 2. Bicuspid Aortic Valve Morphology

	Total Patients	Valve Morphology, N (%)		
		R-L	R-N	L-N
Isolated BAV	569	335 (58.9)	225 (39.5)	9 (1.6)
Aortic coarctation	295	262 (88.8)	31 (10.5)	2 (0.7)
Left heart defects*	155	123 (79.4)	28 (18.1)	4 (2.6)
Non-left heart defects†	116	79 (68.1)	36 (31.0)	1 (0.9)
All patients	1,135	799 (70.4)	320 (28.2)	16 (1.4)

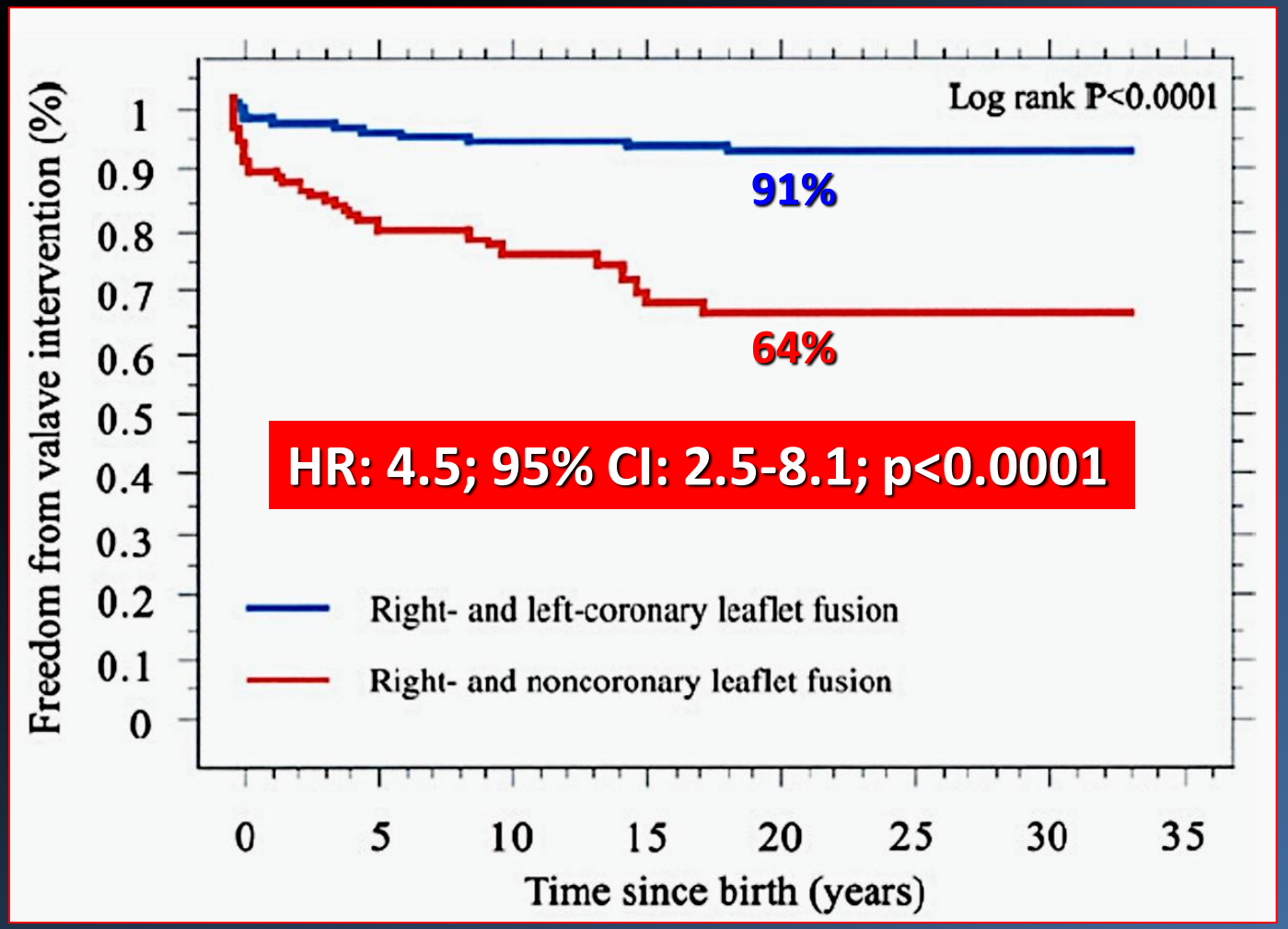
*Includes hypoplastic left heart syndrome, Shone's syndrome, interrupted aortic arch, mitral stenosis, and left ventricular outflow tract obstruction. †Includes atrial and ventricular septal defects, atrioventricular canal defects, anomalous pulmonary venous drainage, right ventricular outflow tract obstruction, Ebstein's malformation, tetralogy of Fallot, and double-outlet right ventricle.

BAV = bicuspid aortic valve; L-N = fusion of left-coronary and noncoronary leaflets; R-L = fusion of right-coronary and left-coronary leaflets; R-N = fusion of right-coronary and non-coronary leaflets.

Fernandes et al. *J Am Coll Cardiol.* 2004;44:1648-51

Type-1 bicuspid valves, LR versus NR : Differences in outcome

310 patients (aged 16 years (6-34), 71% males), BAV: L-R (n=202) vs. N-R (n=108)



Fernandes et al. *J Am Coll Cardiol*. 2007;49:2211-4

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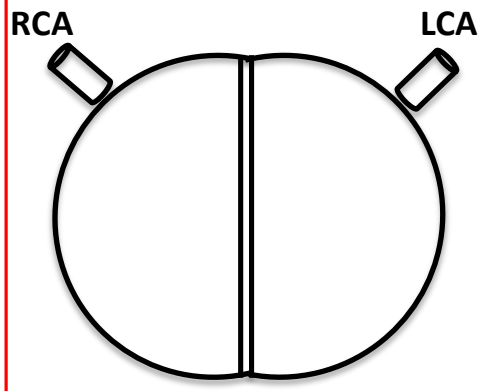
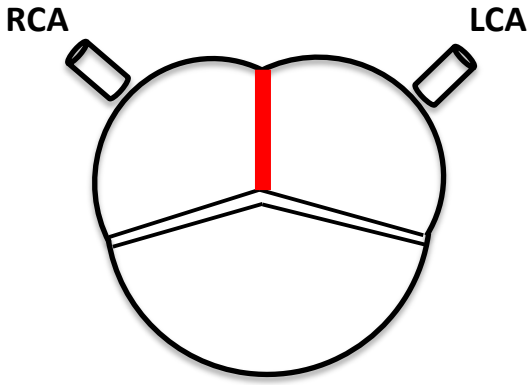
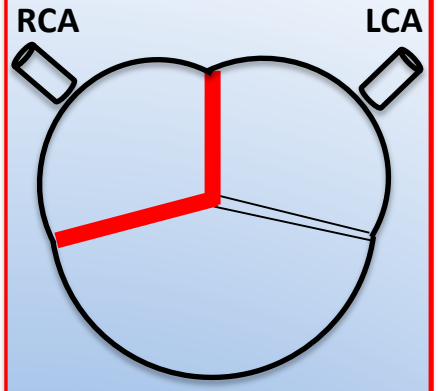
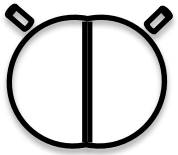
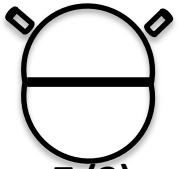
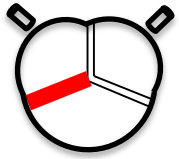
		 21 (7)		 269 (88)			 14 (5)
1. subcategory: spatial position of cusps in Type 0 and raphes in Types 1 and 2		 Lat 13 (4)	 AP 7 (2)	 LR 216 (71)	 NR 45 (15)	 NL 8 (3)	 NR-LR 14 (5)
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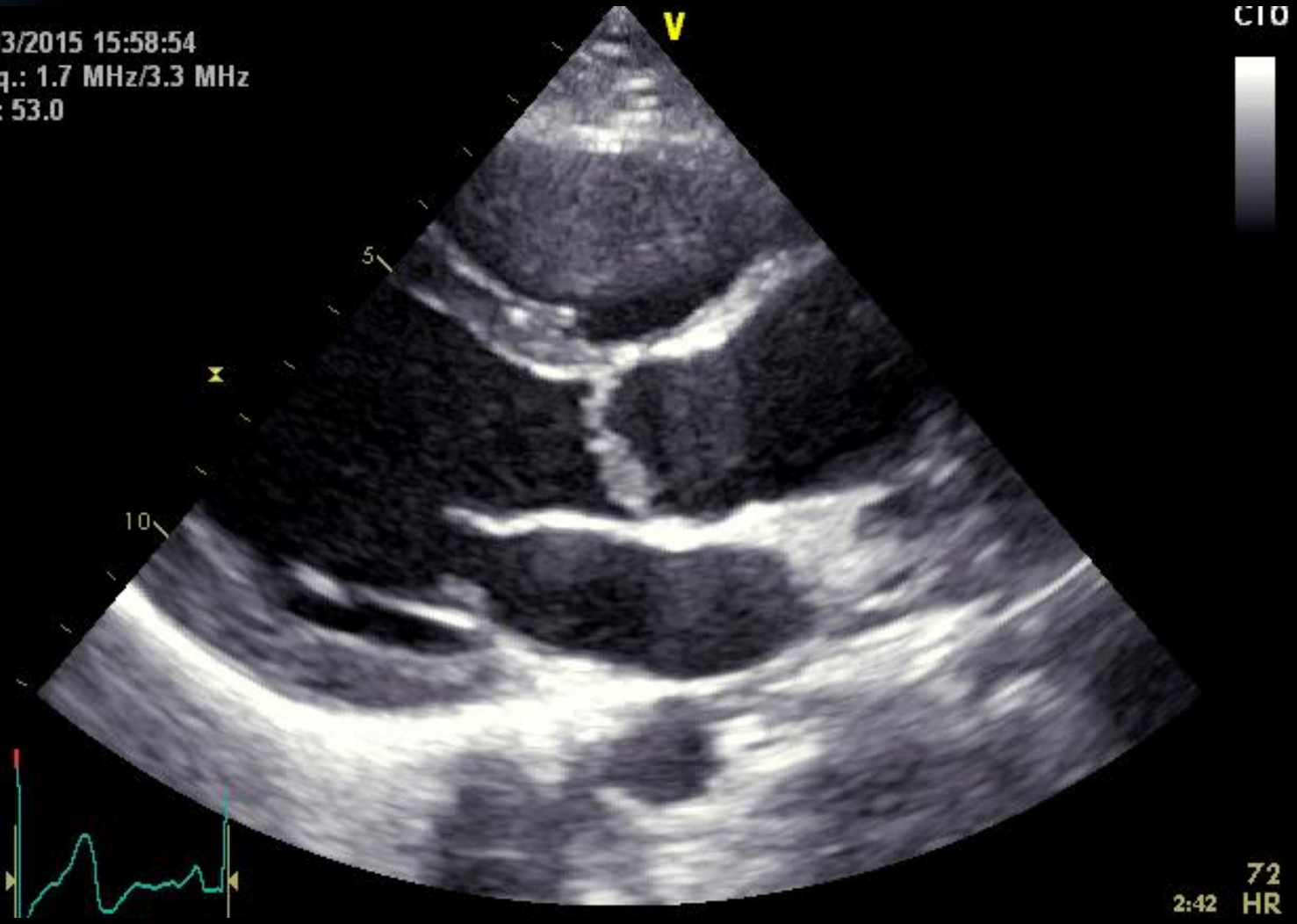
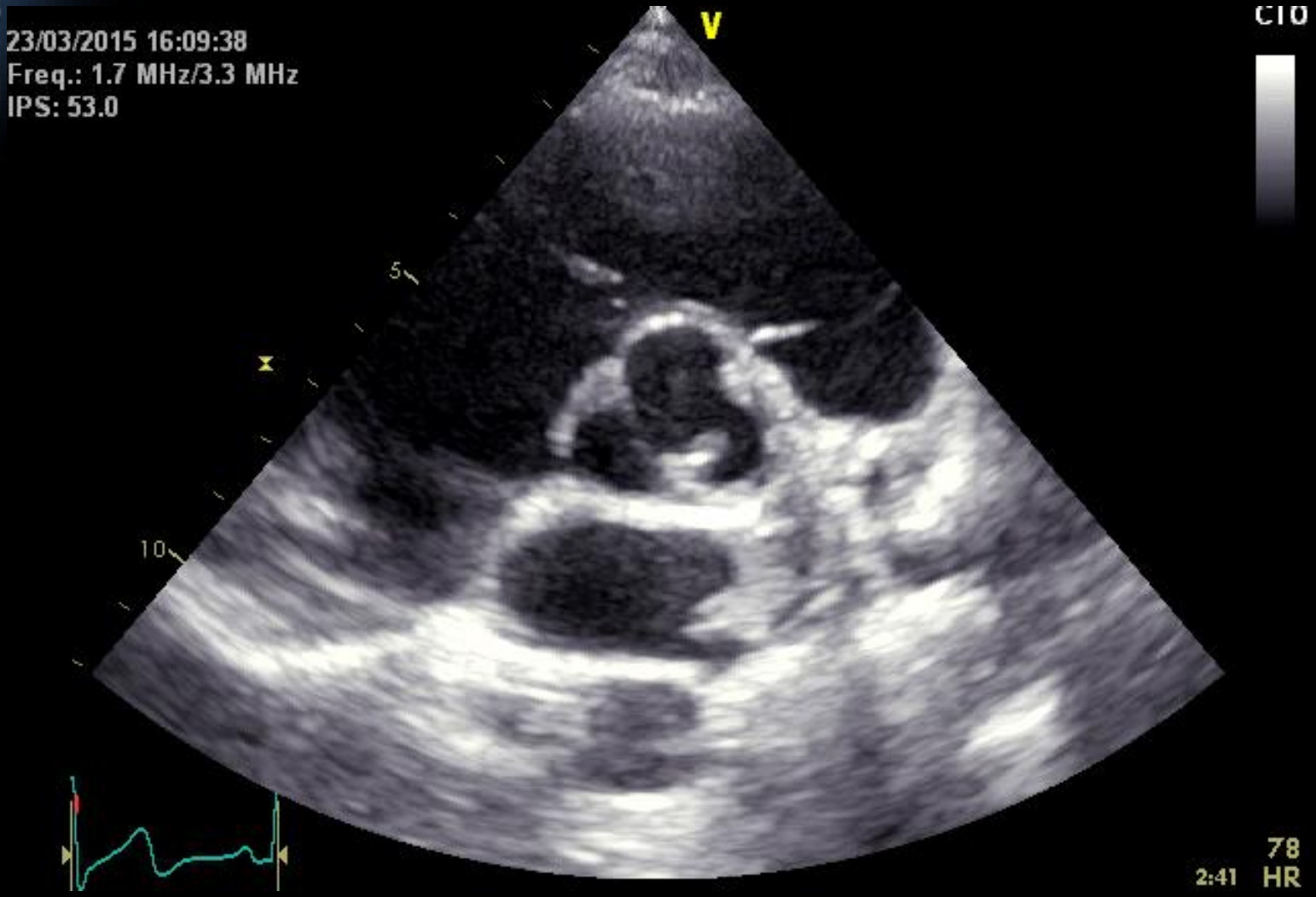
Adapted from Sievers & Schmidtke. *J Thorac Cardiovasc Surg*. 2007; 133:1226-33

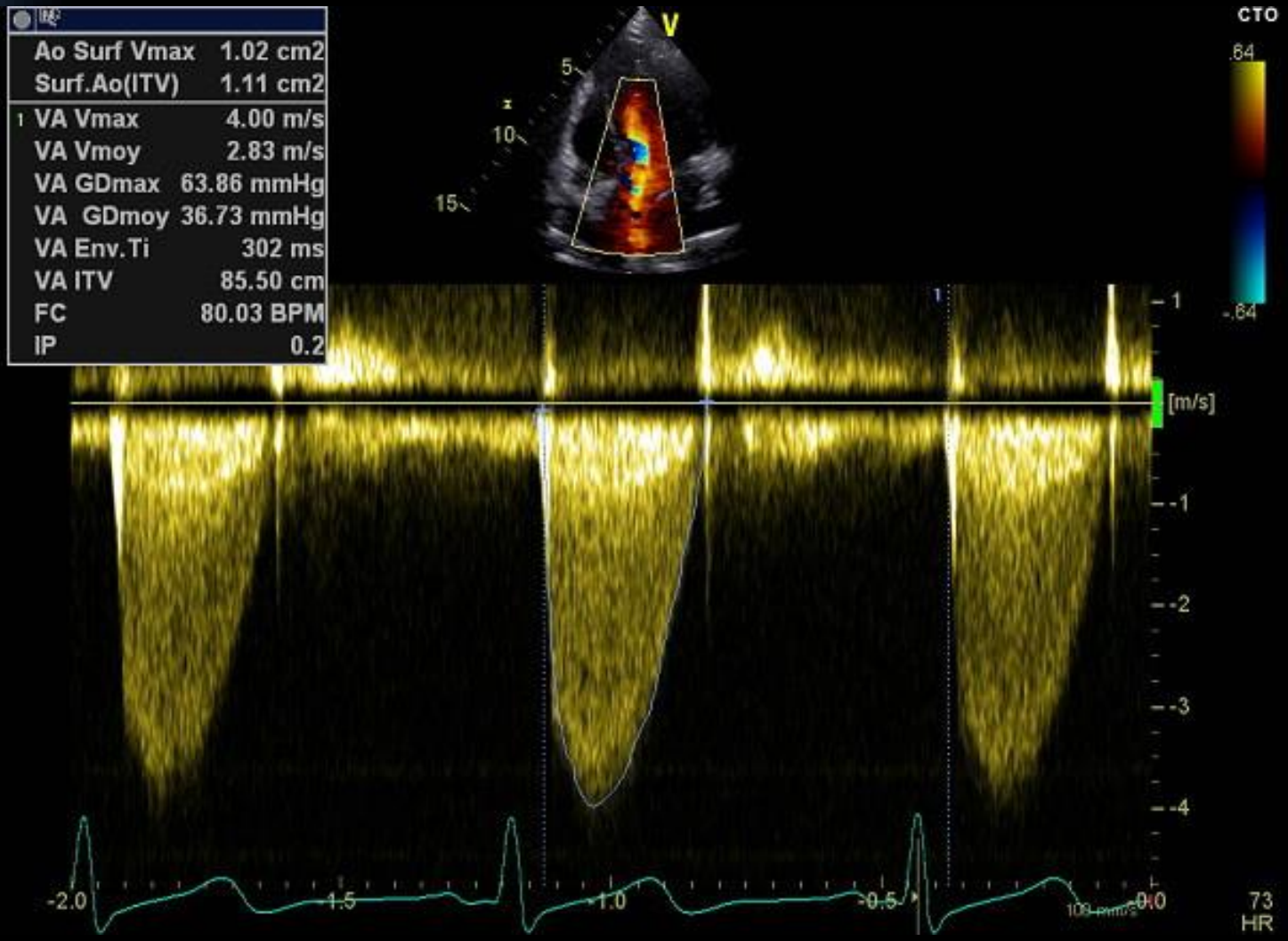
Asymptomatic young woman, 27 years old



BAV type 2 (Unicommisural valve)



Close to severe AS at 27 years old

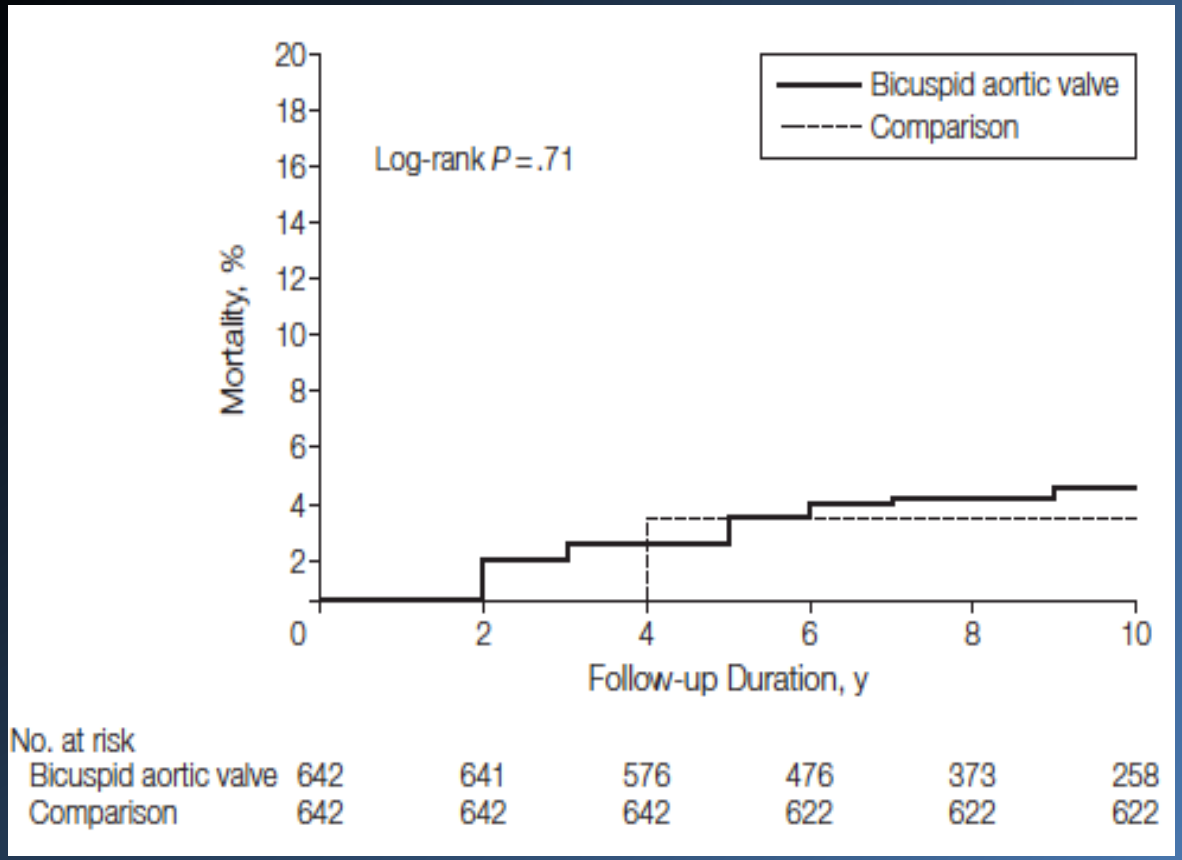


Natural history of BAV: What are the risks?

Outcomes in adults with Bicuspid AV

642 consecutive adults (mean age, 35±16 years; 68% male) with bicuspid AV Toronto Congenital Cardiac center (1994-2001), mean FU: 9±5 years.

- Cardiac mortality rate: 0.3% per patient-year of FU
- Aortic dissection : 0.1% per patient-year of FU
- Overall mortality was not significantly different from age- and sex matched Ontario population estimates (*P=0.71)*



Tzemos et al. *JAMA*. 2008;300: 1317-25

Outcomes in adults with Bicuspid AV

Five patients (1%) had an aortic dissection (3 ascending and 2 descending).
Frequency of dissection: 0.1% per patient-year of follow-up

Indications for intervention:

- **Symptomatic AS (13%)**
- **Symptomatic AR/ LV dysfunction (6%)**
- **Aortic sinus or ascending aorta dilation (2%)**
- **Endocarditis (2%)**

Cardiac Outcomes	No. (%) of Patients (N = 642)
Primary[a]	
Intervention on aortic valve or ascending aorta	142 (22)
Bioprosthetic aortic valve replacement	47
Ascending aortic graft and aortic valve replacement	38
Pulmonary autograft (Ross procedure)	34
Mechanical aortic valve replacement	14
Valve sparing aortic root replacement	5
Aortic valve repair	3
Percutaneous aortic valvotomy	1
Cardiac death	17 (3)
Heart failure	12
Aortic dissection	2
Postoperative after cardiac surgery	3
Hospital admission for heart failure	16 (2)
Aortic complication	11 (2)
Aortic dissection	5
Descending thoracic or abdominal aortic aneurysm	6
Secondary[a]	
Aortic valve endocarditis	13 (2)
Cardiac arrhythmias requiring treatment	30 (5)

[a] Categories are not mutually exclusive.

Tzemos et al. *JAMA*. 2008;300: 1317-25

Incidence of aortic complications in patients with bicuspid aortic valves

Table 1. Baseline Cohort Characteristics by Total Aortic Events[a]

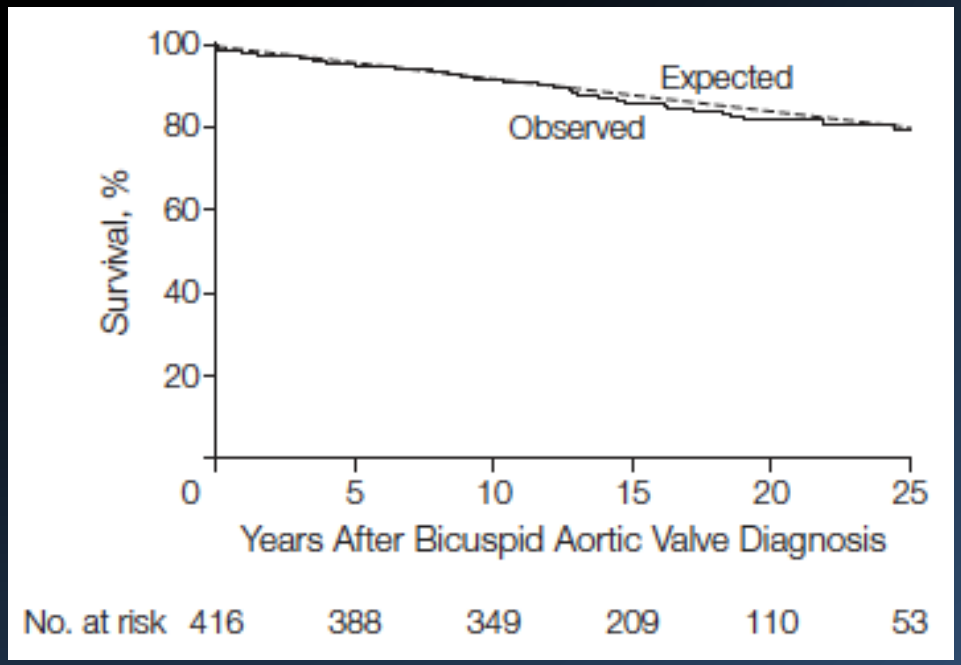
Variable	No. (%) of Patients: Total (N = 416)	Aortic Events (n = 74)	No Aortic Events (n = 342)	*P* Value
Age, mean (SD), y	35 (21)	34 (19)	36 (21)	.45
Men	288 (69)	57 (77)	231 (68)	.12
Hypertension	93 (22)	18 (24)	75 (22)	.64
Smoking	132 (32)	22 (30)	110 (32)	.78
Diabetes	15 (4)	2 (3)	13 (4)	>.99
Atherosclerotic disease[b]	21 (5)	2 (3)	19 (6)	.39
Cardiac symptoms[c]	74 (18)	12 (16)	62 (18)	.86
Coarctation	30 (7)	12 (16)	18 (5)	.009
Ejection fraction %, mean (SD)	62 (7)	62 (6)	63 (7)	.25
Maximum root or ascending aorta diameter, mean (SD), mm	34 (9)	37 (11)	33 (8)	<.001
Typical bicuspid valve	350 (84)	64 (86)	286 (84)	.60
Aortic regurgitation	247 (59)	42 (57)	205 (60)	.71
Aortic stenosis	94 (23)	23 (31)	71 (21)	.06

[a] Aortic events include aneurysm, dissection, and aortic surgery.
[b] History of stroke, transient ischemic attack, or myocardial infarction at baseline.
[c] Any or combinations of cardiac symptoms (syncope, palpitations, dyspnea, typical chest pain) at baseline.

Michelena et al. *JAMA*. 2011; 306: 1104-13

Incidence of aortic complications in patients with bicuspid aortic valves

- Mean FU time of 16±7 years / AORTIC DISSECTION occurred in 2 of 416 patients :
  - Incidence of 3.1 (95% CI, 0.5-9.5) cases per 10 000 patient-years,
  - Age-adjusted relative-risk 8.4 (95% CI, 2.1-33.5; *P*=.003) vs. general population.
- There were no dissections in patients with baseline aortic diameter <45 mm or with normally functioning aortic valves.



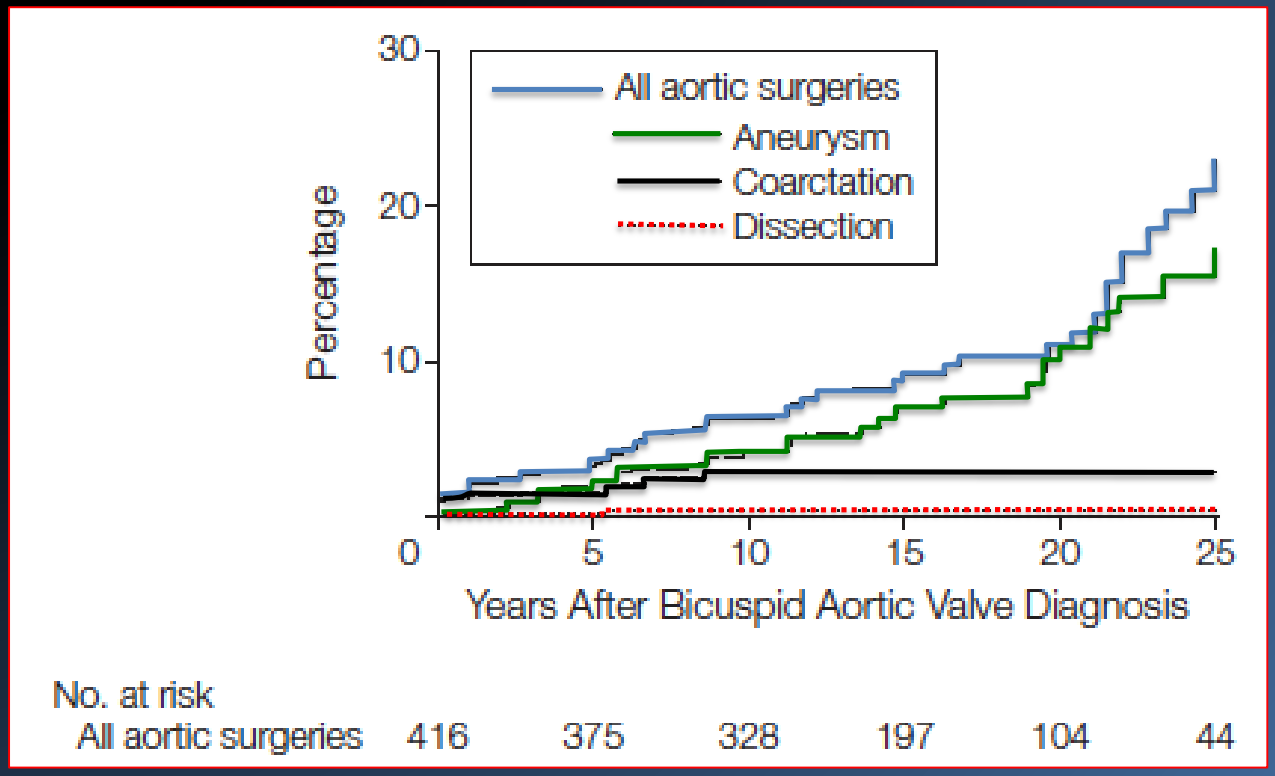
- Survival rate after 25 years was 80% (95% CI 74.2%-85.8%)
- It was identical to the expected survival of the general county population matched for age and sex (*P*=0.98)

Michelena et al. *JAMA*. 2011; 306: 1104-13

Incidence of aortic complications in patients with bicuspid aortic valves

Patients with BAV incur significant morbidity, with 25-year risks of :

- **Aortic valve replacement: 53% (95% CI, 43%-63%)**
- **Surgery of thoracic aorta: 25% (95% CI, 17-33%)**
- **Aneurysm formation: 26% (95% CI, 18-34%)**



Michelena et al. *JAMA*. 2011; 306: 1104-13

Nonsyndromic thoracic aortic aneurysm:
Outcomes versus bicuspid aortopathy and Marfan syndrome

1988-2014: Prospective follow-up of all patients <60 years of age and genetic aortopathy:

1/ Bicuspid aortic valve (BAV), n=228

2/ Marfan syndrome (MFS), n=221

3/ Heritable TAA without obvious physical features = non-syndromic TAA (NS-TAA), n=311

The 687 patients surviving >30 days after presentation were followed for a median of 7 years.

TABLE 2 Clinical Features of Patients Enrolled in Clinical Surveillance

	NS-TAA (n = 253)	MFS (n = 209)	BAV (n = 225)	All Patients (N = 687)
Follow-up time, yrs	6.0 (2.0-12.0)	11.0 (6.0-18.0)*	7.0 (3.0-15.0)	7.0 (3.0-15.0)
Age, yrs	40.7 ± 12.3	27.8 ± 11.7*	38.8 ± 13.7	36.2 ± 13.7
Female	61 (24.1)	90 (43.1)*	39 (17.3)	190 (27.7)
Family history of aneurysm	154 (60.9)	136 (65.1)	35 (15.6)†	325 (47.3)
Family history of dissection	80 (31.6)	43 (20.6)*	5 (2.2)	128 (18.6)
Initial aortic diameter, mm‡	45.6 ± 10.5	41.9 ± 9.2§	44.1 ± 8.4‖	44.0 ± 9.5
Systolic BP, mm Hg	127.3 ± 15.5	117.6 ± 14.5*	128.0 ± 12.4	124.5 ± 15.0
Diastolic BP, mm Hg	77.4 ± 8.6	71.3 ± 9.0*	76.8 ± 8.7	75.3 ± 9.1

Values are median (interquartile range), mean ± SD, or n (%). *p < 0.001 versus NS-TAA and BAV. †p < 0.001 versus NS-TAA and MFS. ‡Available for n = 643 (93.6%). §p < 0.001 versus NS-TAA. ‖p < 0.05 versus MFS.
Abbreviations as in Table 1.

Sherrah et al. *J Am Coll Cardiol.* 2016;67:618–26

Nonsyndromic thoracic aortic aneurysm:
Outcomes versus bicuspid aortopathy and Marfan syndrome

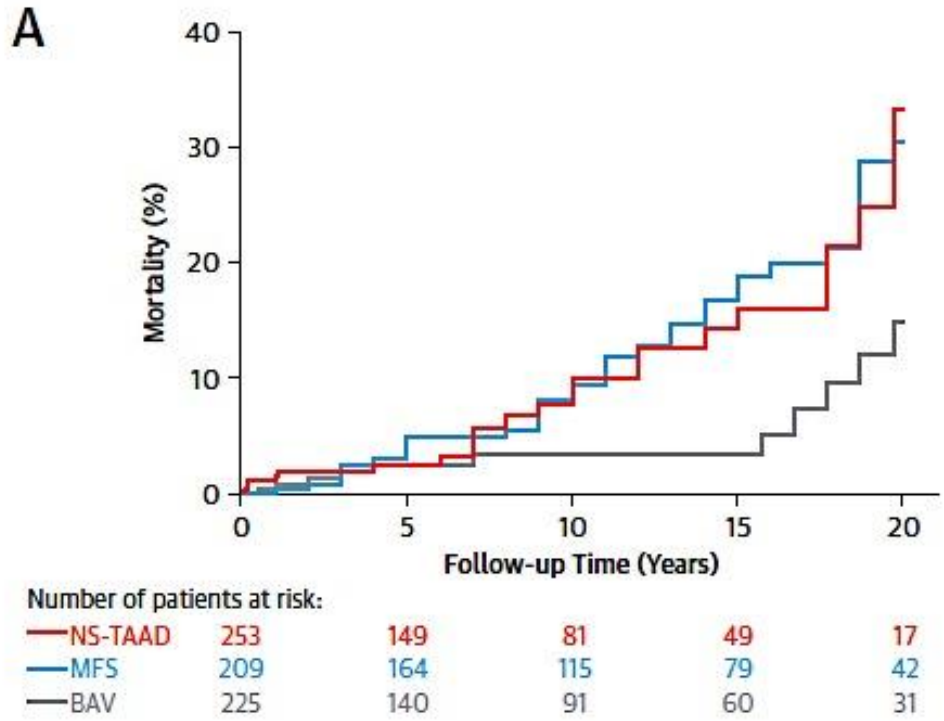
10-year mortality was 3.5% for BAV, 7.8% for NS-TAA and 8.7% for MFS: p <0.05 for BAV vs. NS-TAA and MFS

Factors associated with all-cause mortality:

- Marfan syndrome (p < 0.04)
- Age at presentation
- Family history of dissection (doubling the mortality risk)

Aorta size = NS

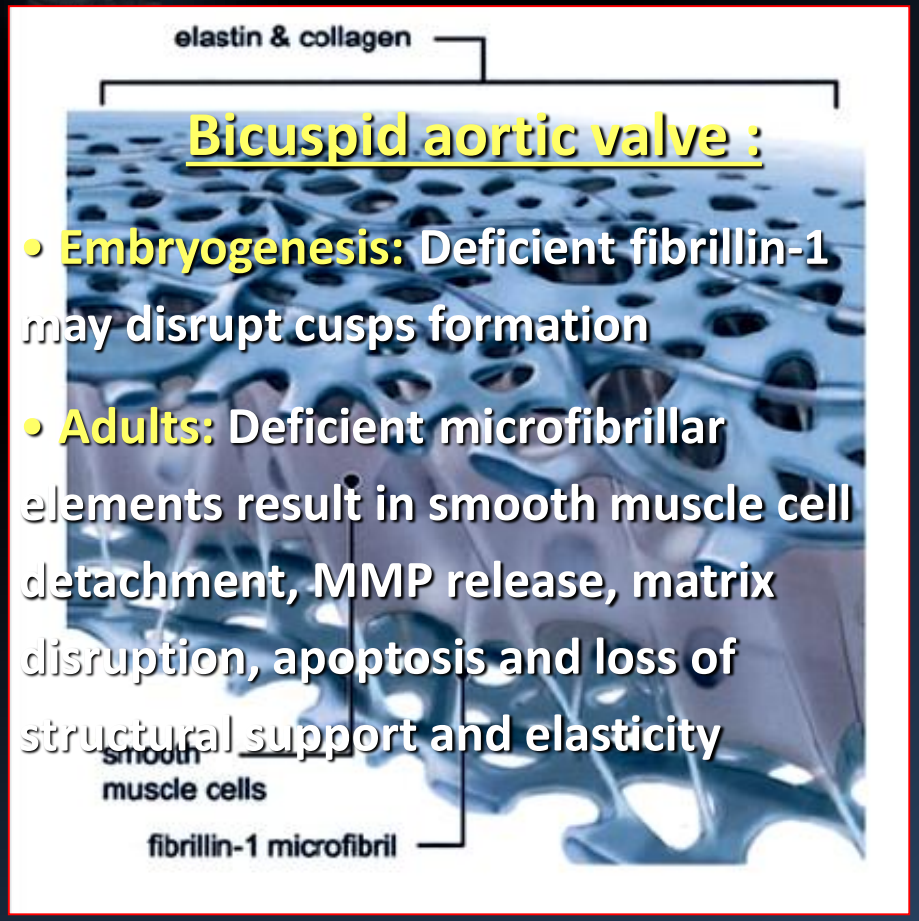
FIGURE 1 Mortality in Clinical Surveillance Group



Sherrah et al. *J Am Coll Cardiol.* 2016;67:618–26

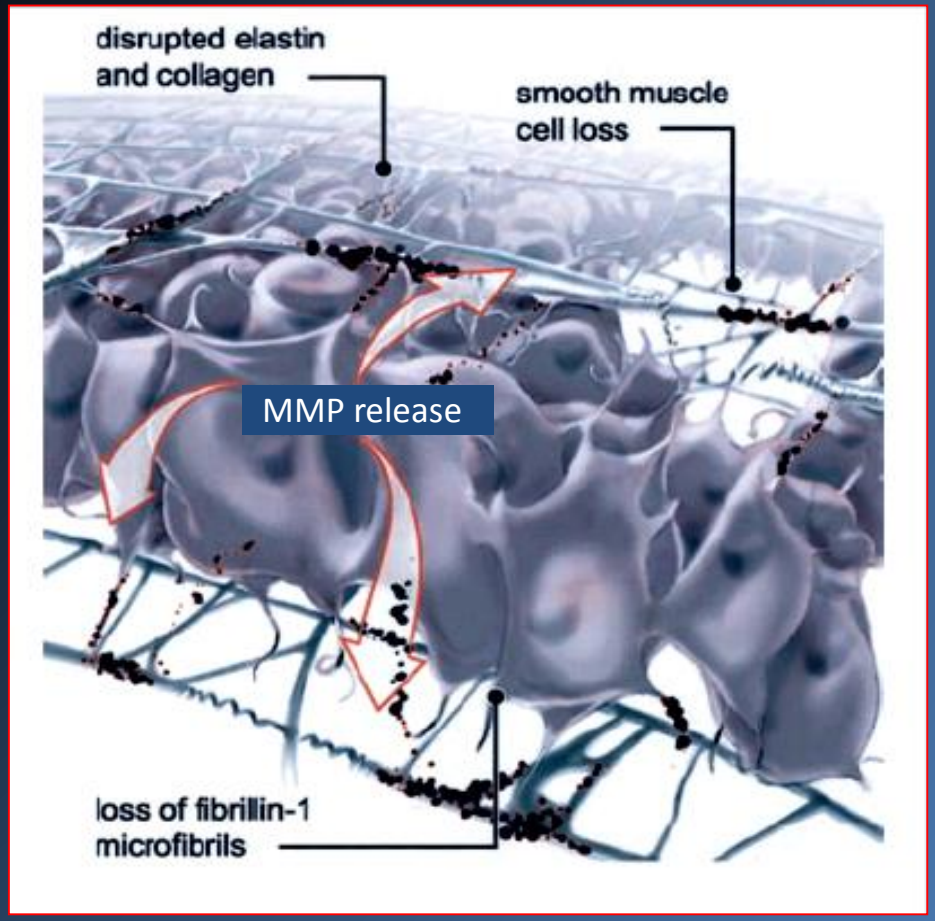
Bicuspid Aortopathy: Genetics or hemodynamics?

Bicuspid Aortopathy: Pathophysiological Features



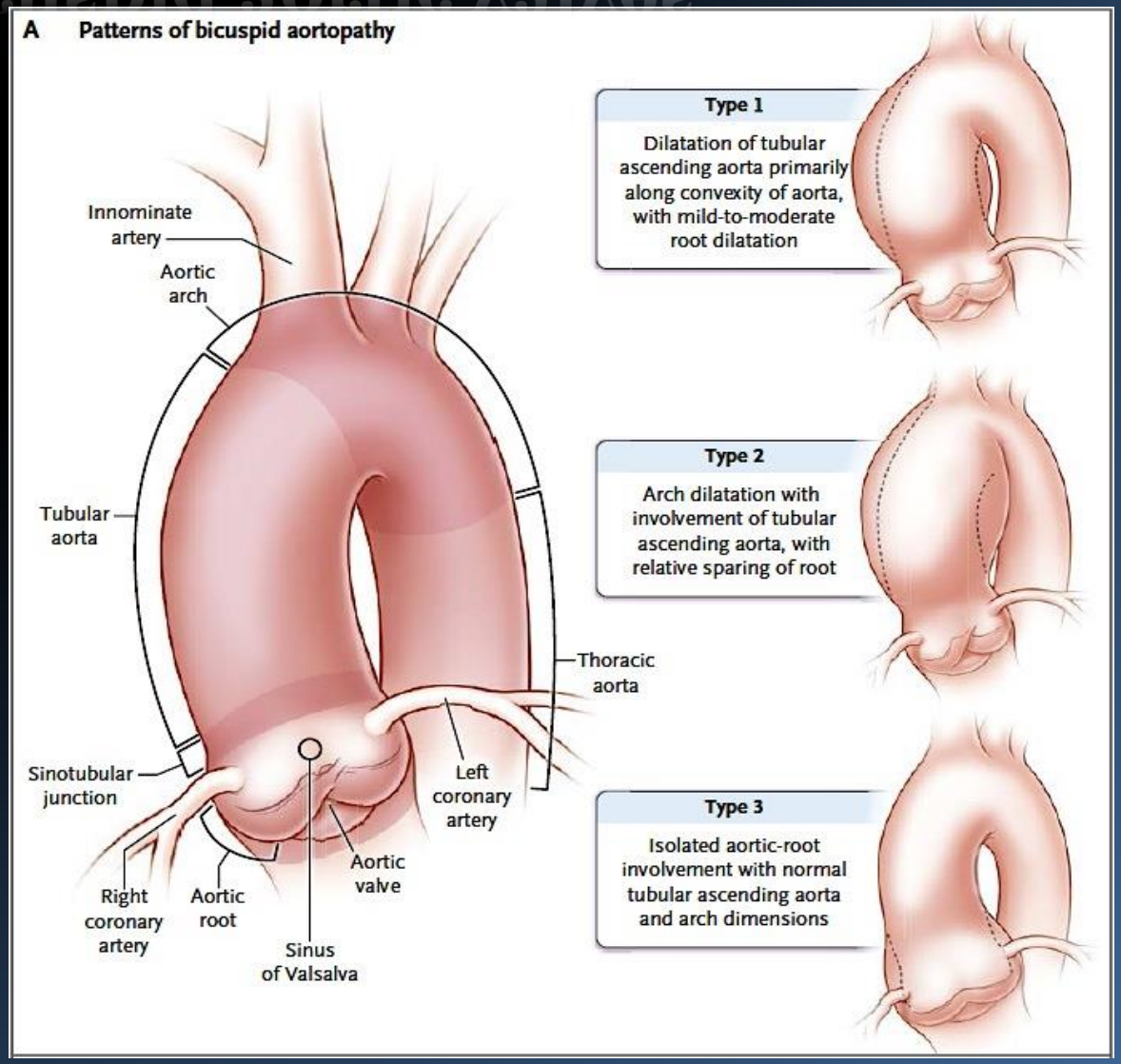
Bicuspid aortic valve :

- **Embryogenesis:** Deficient fibrillin-1 may disrupt cusps formation
- **Adults:** Deficient microfibrillar elements result in smooth muscle cell detachment, MMP release, matrix disruption, apoptosis and loss of structural support and elasticity



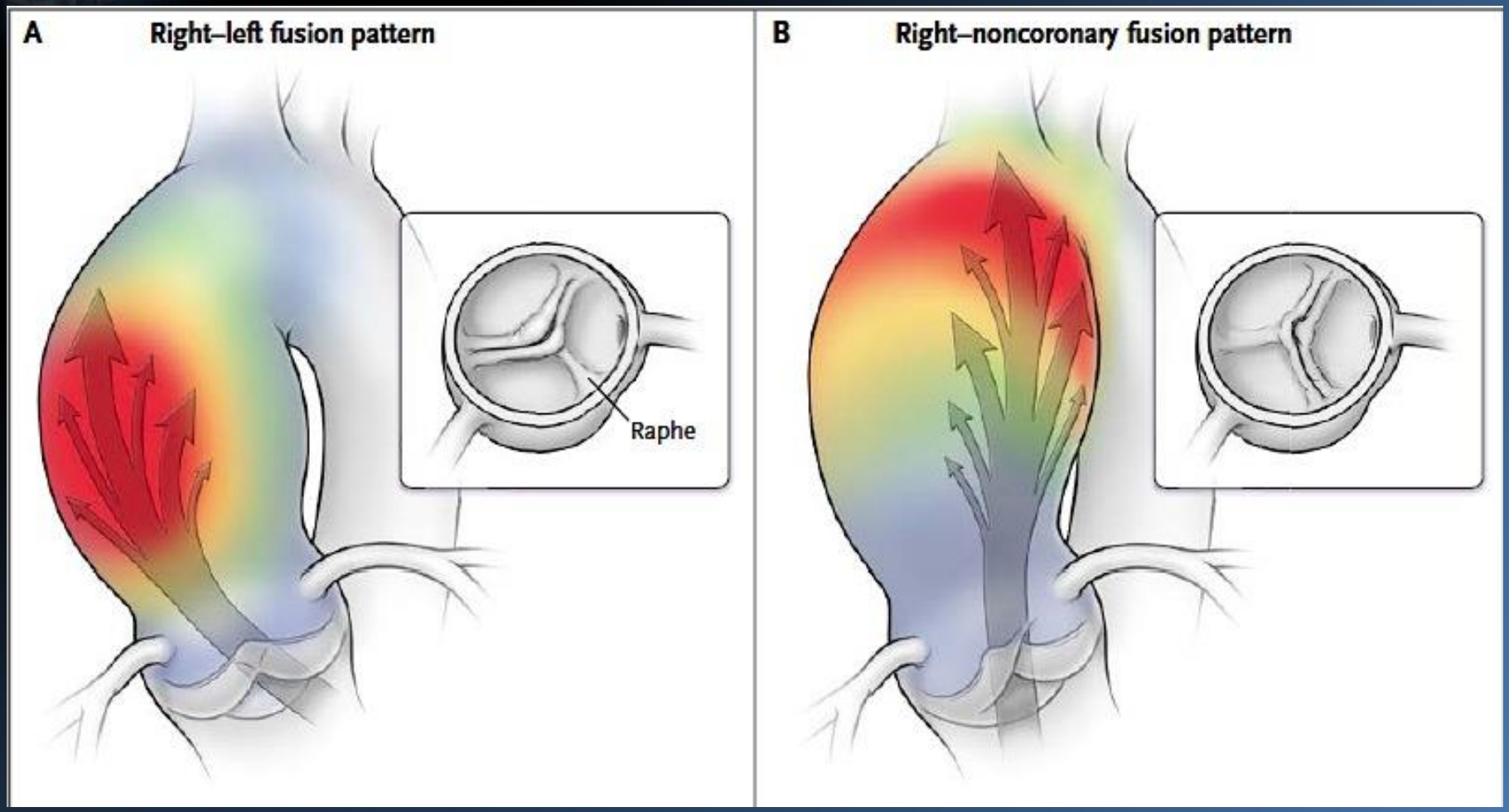
Fedak et al. *Circulation.* 2002; 106:900-4

Aortic dilatation in patients with bicuspid aortic valves



Verma & Siu. *N Engl J Med.* 2014; 370: 1920-9

Aortic dilatation in patients with bicuspid aortic valve

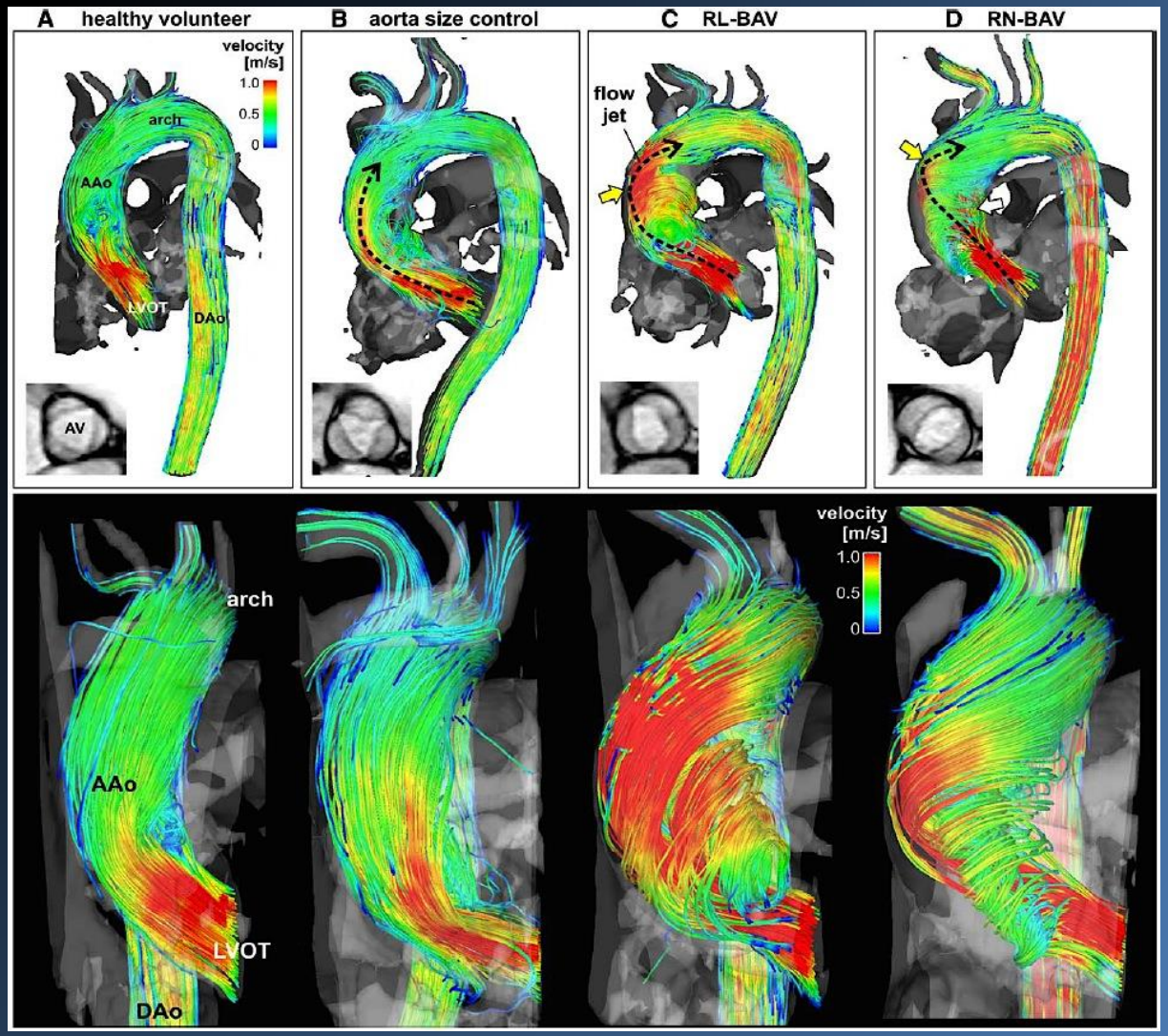


Verma & Siu. *N Engl J Med.* 2014; 370: 1920-9

Bicuspid aortic cusp fusion alters aorta flow patterns (3D-MRI)

Four-dimensional flow-MRI can measure in vivo 3-D blood flow in the aorta

The type of BAV fusion is associated with regional wall shear stress distribution, systolic flow eccentricity and expression of BAV aortopathy



Mahadevia et al. *Circulation*. 2014; 129: 673-82

Henri Mondor

Valve-related hemodynamics mediate human bicuspid aortopathy

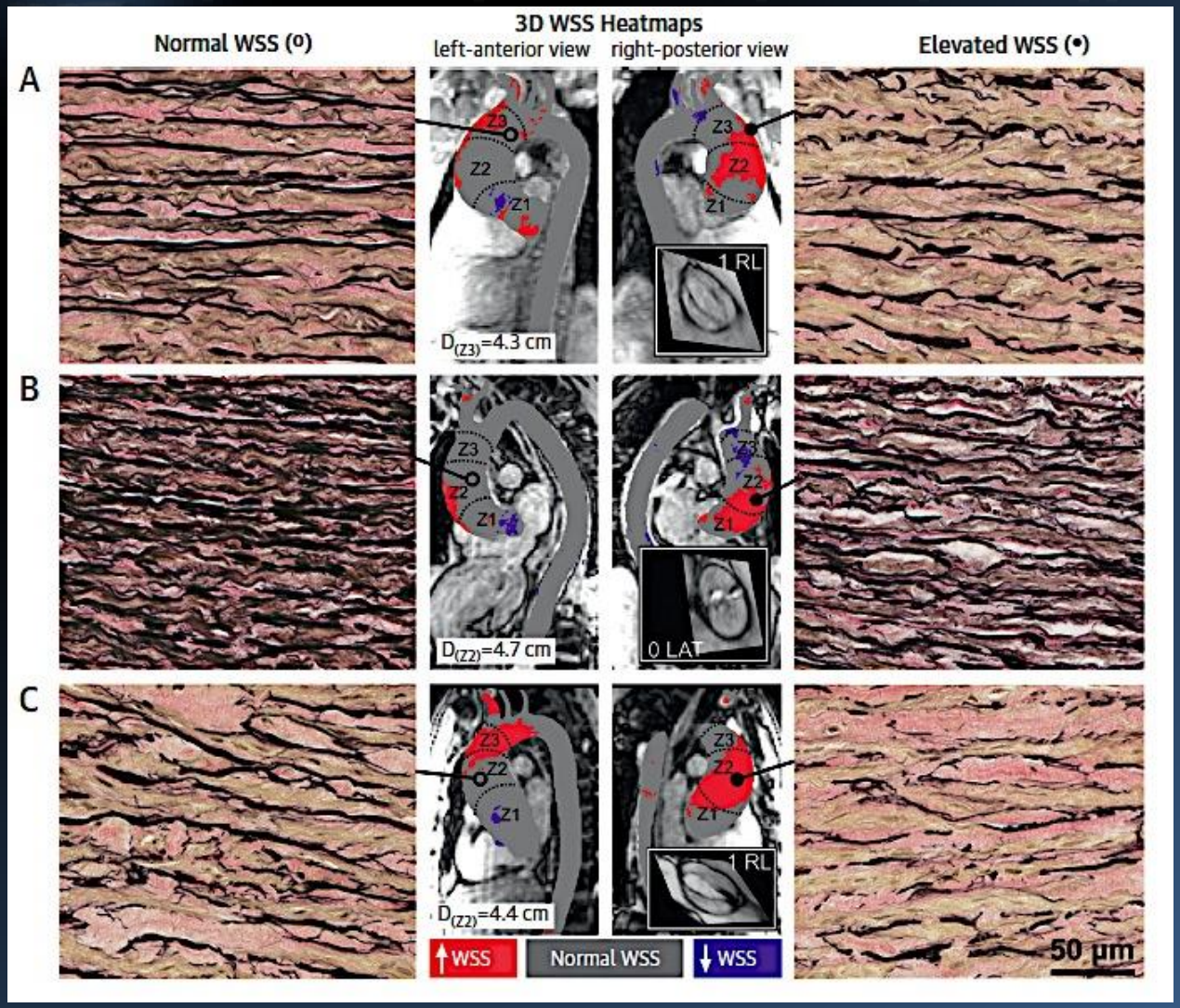
TABLE 1 Patient Characteristics in BAV Study Population (N = 20)

Age, yrs	48 ± 15
Female	2 (10)
BAV classification	
Type 0, lateral	2 (10)
Type 1, RN	1 (5)
Type 1, RL	12 (60)
Type 2, RL/RN	5 (25)
Aortic diameter, cm	
Sinus of Valsalva	4.4 ± 0.5 (range 3.7–5.7)
Mid ascending aorta	4.7 ± 0.6 (range 3.6–6.3)
Aortic valve function	
No AS, moderate/severe AR	5 (25)
Mild AS, moderate/severe AR	1 (5)
Moderate/severe AS, no AR	5 (25)
Moderate/severe AS, mild AR	3 (15)
Moderate/severe AS, moderate/severe AR	6 (30)
Hypertension	7 (35)
Surgical procedure: aortic valve	
Repair	1 (5)
Replacement	19 (95)
AVR	4 (20)
Bentall	14 (70)
Ross	1 (5)
Surgical procedure: AsAo	
AsAo replacement	20 (100)
Root replacement	16 (80)
Hemi-arch	8 (40)

- BAV patients (n = 20) undergoing ascending aortic resection underwent pre-operative 4D-flow CMR to regionally map aortic wall sheer stress (WSS).
- Paired aortic wall samples (within-patient samples obtained from regions of elevated vs. normal WSS) were collected and compared for medial elastin degeneration by histology and extra-cellular matrix (ECM) regulation by protein expression (mainly MMP and TGFβ).

Guzzardi et al. *J. Am Coll Cardiol.* 2015; 66: 892–900

Valve-related hemodynamics mediate human bicuspid aortopathy



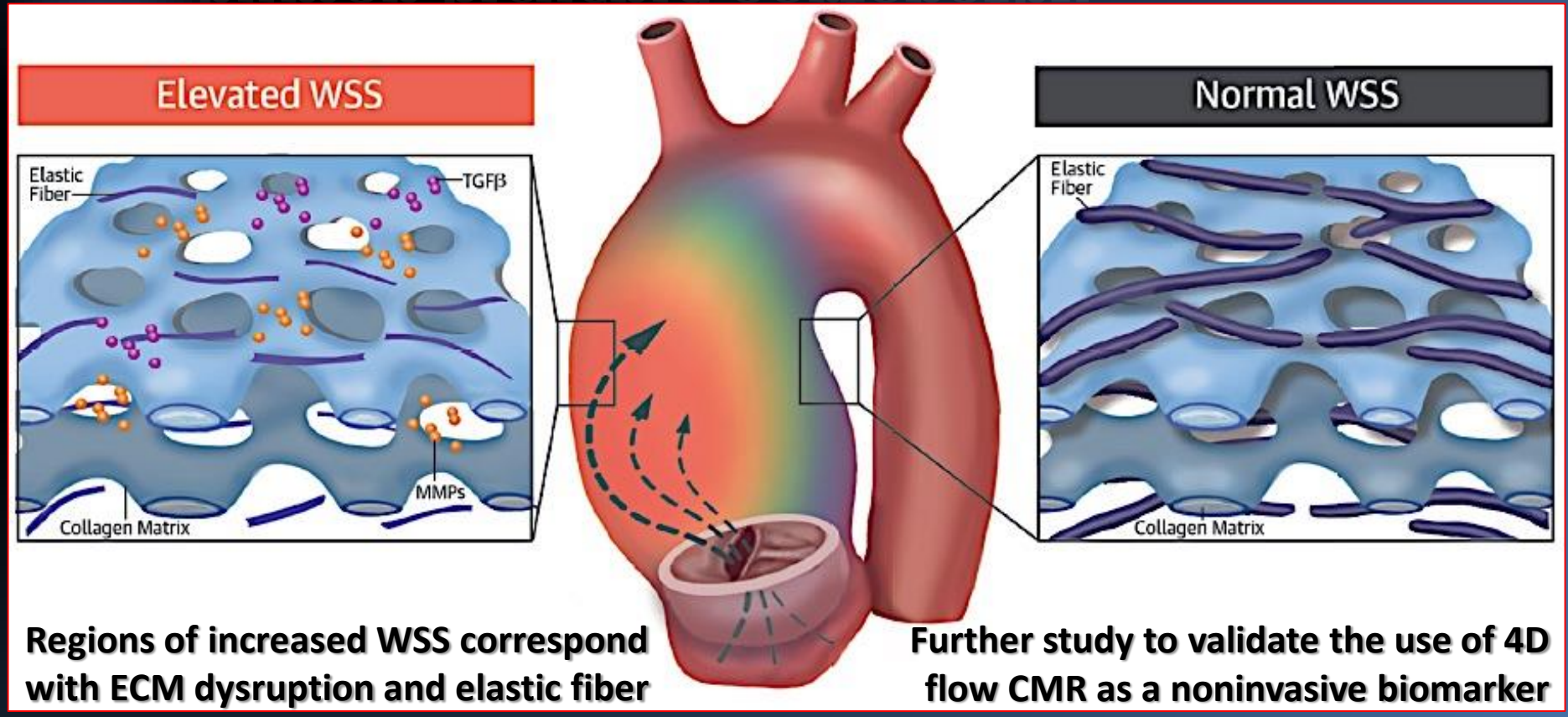
Regions of high wall shear stress (WSS) had:

1. Fewer elastin fibers (black)
2. Thinner Elastin fibers
3. farther apart

as compared with regions with normal WSS

Guzzardi et al. *J. Am Coll Cardiol.* 2015; 66: 892–900

Valve-related hemodynamics mediate human bicuspid aortopathy



Regions of increased WSS correspond with ECM dysruption and elastic fiber degeneration in the ascending aorta of BAV patients, implicating valve-related hemodynamics as a contributing factor in the development of aortopathy.

Further study to validate the use of 4D flow CMR as a noninvasive biomarker of disease progression and its ability to individualize resection strategies is warranted

Guzzardi et al. *J. Am Coll Cardiol.* 2015; 66: 892–900

FOLLOW UP?

Bicuspid aortic valves/ Aortopathy

• When echocardiography does not provide adequate images of the ascending aorta to a distance ≥4.0 cm from the valve plane, additional imaging is needed.

Class I : CMR or MDCT is indicated in patients with a bicuspid valve when morphology of the proximal aorta cannot be assessed accurately or fully by echocardiography. *(Level of Evidence: C)*

• Cardiac Magnetic Resonance imaging is preferred over CT Scan, when possible, because of the absence of ionizing radiation exposure in patients who likely will have multiple imaging studies over their lifetime.

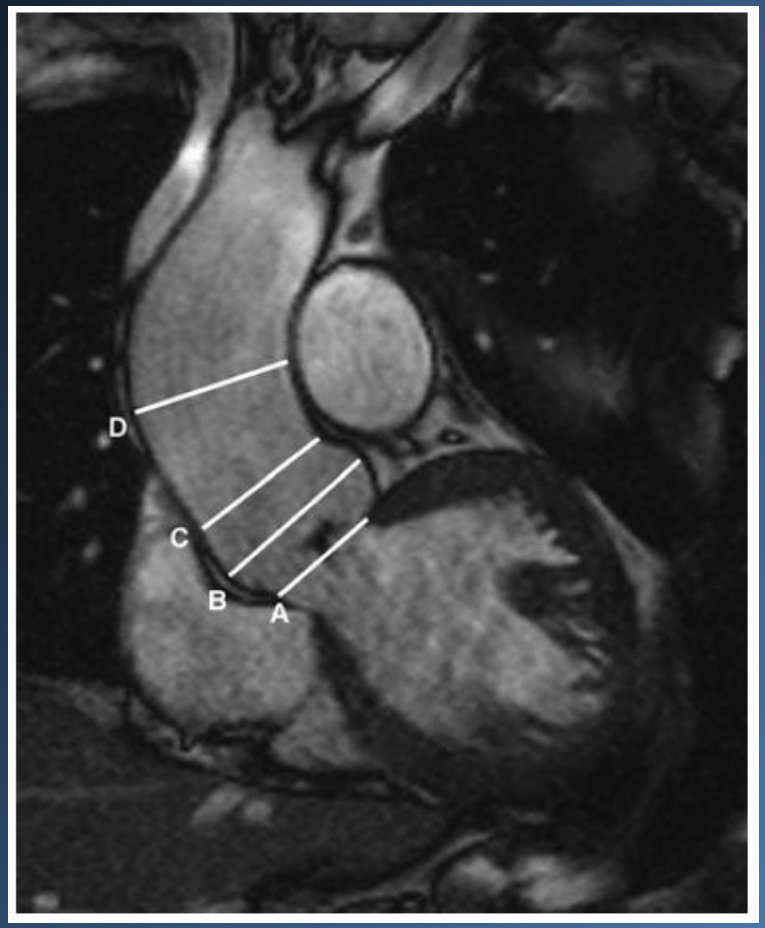
AHA/ACC Guidelines on VHD. *Circulation*. 2014; 129: e521-e643

Bicuspid aortic valves/ Aortopathy

Class I : Serial assessment of the proximal aorta by TTE, CMR (MDCT) is recommended in patients with a bicuspid valve and aortic diameter >40 mm.

Examination interval is determined by the rate of progression of aortic dilation and by family history.

In patients with an aortic diameter >45 mm, evaluation should be performed annually. ***(Level of Evidence: C)***



AHA/ACC Guidelines on VHD. *Circulation*. 2014; 129: e521-e643

Bicuspid AV/ Ascending Aorta : What about sports?

- Patients without aortic root dilatation (<40 mm or <21 mm/m²) and no significant AS or AR may participate in all competitive sports

- Patients with dilated aortic roots between 40 and 45 mm may participate in low/moderate static or dynamic competitive sports, but should avoid any sports involving the potential for bodily collision or trauma

- Patients with dilated aortic roots >45 mm can participate in only low-intensity competitive sports

Bonow et al. 36th Bethesda Conference/ Task Force 3: *J Am Coll Cardiol.* 2005;14:1334–40.

Henri Mondor

SURGERY ?

Bicuspid aortic valves/ Aortopathy

Class I : Surgery to repair the aortic sinuses or replace the ascending aorta is indicated in patients with a bicuspid aortic valve if any diameter of the proximal aorta is >55 mm.
(Level of Evidence: B)

Class IIa : Surgery to repair the aortic sinuses or replace the ascending aorta is reasonable in patients with a bicuspid aortic valve if the diameter of the proximal aorta is >50 mm with additional risk factor for dissection. (family history of aortic dissection/ increase in diameter ≥5 mm per year).
(Level of Evidence: C)

AHA/ACC Guidelines on VHD. *Circulation*. 2014; 129: e521-e643

Bicuspid aortic valves/ Aortopathy

Class IIa : Replacement of the ascending aorta is reasonable in patients with a bicuspid aortic valve who are undergoing aortic valve surgery because of severe AS or AR if the diameter of the ascending aorta is >45 mm. *(Level of Evidence: C)*

Replacement of the sinuses of Valsalva is not necessary in all cases and should be individualized based on the displacement of the coronary ostia, because progressive dilation of the sinus segment after separate valve and graft repair is uncommon.

AHA/ACC Guidelines on VHD. *Circulation*. 2014; 129: e521-e643

What is a significant increase in size? Should we index to body size?

- Inter- and intra-observer variability of CT for AAA are around 5 and 3 mm respectively.
- Thus, any change of ≥5 mm on serial CT can be considered a significant change, smaller changes are difficult to interpret.

RECOMMENDATIONS	COR	LOE
Allergy to contrast media, pregnancy and renal function should be systematically assessed to select the best imaging modality with minimal radiation exposure, except for emergency cases	I	C
The risk of radiation exposure should be assessed, especially in young adults and those undergoing serial imaging	IIa	B
Aortic diameters may be indexed to BSA, specially for patients with small body size	IIb	B

ESC Guidelines on aortic diseases. *Eur Heart J*. 2014; 35: 2873-926

Moderate aortic enlargement and bicuspid aortic valve are associated with aortic dissection in Turner syndrome

- Data from 20 individuals with acute AD (Type A in 17 cases, 5%) from the International Turner Syndrome Aortic Dissection Registry

18 of 19 patients (95%) with AAD had an associated cardiac malformation that included a bicuspid aortic valve.

For those with type A dissections, the mean ascending aorta size index (ASI) was 2.7±0.6 cm/m²

Patients with Turner syndrome aged >18 years with ASI >2.5 cm/m² should be considered for an aortic operation to prevent aortic dissection

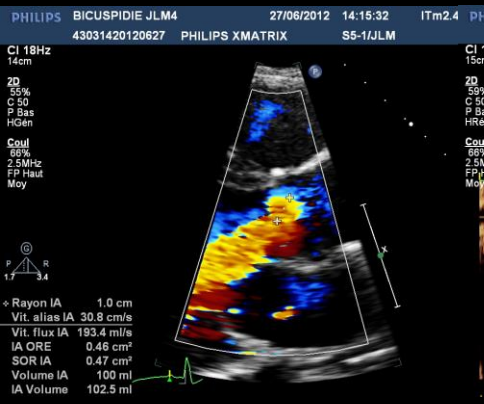
Patient No.	Age at Dissection, y	Cardiac Diagnosis (Comment)	Hypertension (Y/N)	Location of Dissection	Symptom Duration	Outcome
	18	BAV	N	Type A	>24 hr	Death
	18	BAV, coarctation	Y	Type A	>24 hr	Death
	21	BAV	N	Type A	>24 hr	Alive
	23	BAV, h/o IAA	N	Type A	>24 hr	Death
	24	BAV, coarctation	N	Type A	>24 hr	Death
	27	BAV	N	Type A	>24 hr	Alive
	28	BAV	N	Type A	<24 hr	Alive
	28	BAV	Y	Type A	>24 hr	Death
	28	BAV, unrepaired coarctation	N	Type A	<24 hr	Death
	29	BAV	Y	Type A	>24 hr	Death
	29	BAV	N	Type B	<24 hr	Death
	30	BAV	Y	Type A	>24 hr	Death
	34	BAV	N	Type A	<24 hr	Death
	35	coarctation (dissection during stent)	Y	Type B	<24 hr	Alive
	37	BAV, VSD	Y	Type A	>24 hr	Death
	40	None	N	Type A	<24 hr	Death
	41	BAV	N	Type A	>24 hr	Alive
	44	BAV	N	Type A	>24 hr	Alive
	48	BAV/severe AS	Unknown	Type A	<24 hr	Alive
20*	48	BAV, abberrant RSA	Y	Type B	>24 hr	Alive
Mean, SD, med, 1st quart, 3rd quart (for type A dissections only)	30.5, 8.7, 28, 23.5, 38.5					

*Not previously reported, reference is given for known prior reports others may or may not have been reported.
AS indicates aortic stenosis; BAV, bicuspid aortic valve; IAA, interrupted aortic arch; VSD, ventricular septal defect; RSA, right subclavian artery.

Carlson et al. *Circulation.* 2012; 126: 2220-26

Take-Home Message

- BAV is the most frequent congenital heart defect
- It should be detected by TTE in young patients with cardiac murmurs, aortic insufficiency or dilatation, first relatives of patients with BAV and women with Turner syndrome
- The primary risk of BAV is severe AS (or AR) requiring surgery between 45-65 years of age (50% risk)
- Lower risk of surgery for aortic aneurysm: 25% around the same age
- Although significantly higher than the general population, the risk of aortic dissection is significantly lower than in Marfan syndrome or other forms of genetic aneurysms
- Contemporary favorable outcomes are at the price of regular follow-up (clinical and imaging) and proactive guideline-based elective aortic surgery for ascending aortic aneurysms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Vit. flux IA 193.4 ml/s
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SOR IA 0.47 cm²
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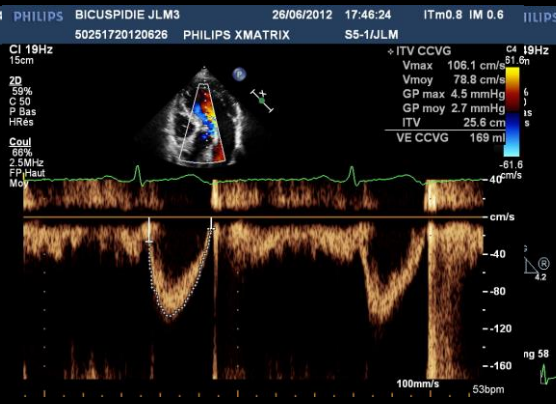
IA Volume 102.5 ml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ITV CCVG
Vmax 106.1 cm/s
Vmoy 78.8 cm/s
GP max 4.5 mmHg
GP moy 2.7 mmHg
ITV 25.6 cm
VE CCVG 169 ml
cm/s
-40
-80
-120
-160
100mm/s
53bpm


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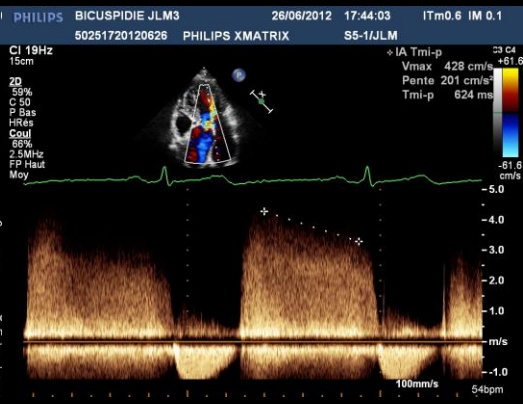
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Coul
66%
2.5MHz
FP Haut
Moy
IA Tmi-p
Vmax 428 cm/s
Pente 201 cm/s²
Tmi-p 624 ms
-5.0
-4.0
-3.0
-2.0
-1.0
m/s
100mm/s
54bpm

Aortic Dilatation and Dissection in Turner Syndrome

- **166 adult volunteers with Turner Syndrome (aged 36±11 years)** not selected for CV disease and 26 healthy female control subjects.
- Ascending and descending aortic diameters measured by MRI at the right pulmonary artery.

Table 1. Aortic Dimensions in Women With TS and Female Control Subjects

	TS (n=166)	NV (n=26)	NV 95th Percentile	*P*
Age, y	36.2/11.3	35.3/9.5	...	0.725
Height, cm	147.0/7.8	164.2/5.5	...	<0.0001
BSA, m^2	1.54/0.2	1.71/0.1	...	<0.0001
AD, cm	2.86/0.48	2.91/0.30	3.40	0.647
DD, cm	1.94/0.34	2.18/0.23	2.56	0.0007
AD/DD	1.49/0.25	1.34/0.10	1.50	0.002
AD/BSA (ASI), cm/m^2	1.89/0.34	1.70/0.16	1.96	0.008
DD/BSA, cm/m^2	1.28/0.26	1.27/0.11	1.45	0.855

NV indicates normal volunteer. Data are mean/SD. Mean values compared by ANOVA.

Matura et al. *Circulation.* 2007; 116: 1663-70

Aortic Dilatation and Dissection in Turner Syndrome

- Only 9.5% of women with Turner Syndrome (TS) exceeded the 95th percentile for absolute ascending aorta diameter
- 24% and 45% of women with TS exceeded the 95th percentile for Aorta Size Index (ASI) and Ascending/ Descending Aorta ratio (AD/ DD) respectively.
- The presence of a bicuspid aortic valve and/or elongation of the transverse aortic arch (ETA) was associated with greater ASI.

Table 2. Factors Associated With Aortic Dilatation in TS

	AD/M^2	*P*	DD/M^2	*P*	AD/DD	*P*
BAV (n=28/150)	2.0/0.4	...	1.2/0.2	...	1.7/0.3	...
TAV (n=122/150)	1.8/0.3	0.001	1.3/0.2	0.053	1.5/0.2	<0.0001
Web neck (n=49/152)	1.9/0.4	...	1.3/0.3	...	1.5/0.3	...
No web neck (n=103/152)	1.9/0.3	0.725	1.2/0.2	0.573	1.5/0.3	0.989
ETA (n=58/109)	1.9/0.4	...	1.3/0.3	...	1.5/0.2	...
No ETA (n=51/109)	1.8/0.3	0.017	1.2/0.2	0.016	1.5/0.2	0.814
X^M (n=81/113)	1.9/0.3	...	1.3/0.2	...	1.5/0.2	...
X^P (n=32/113)	1.9/0.4	0.835	1.2/0.2	0.469	1.5/0.3	0.292

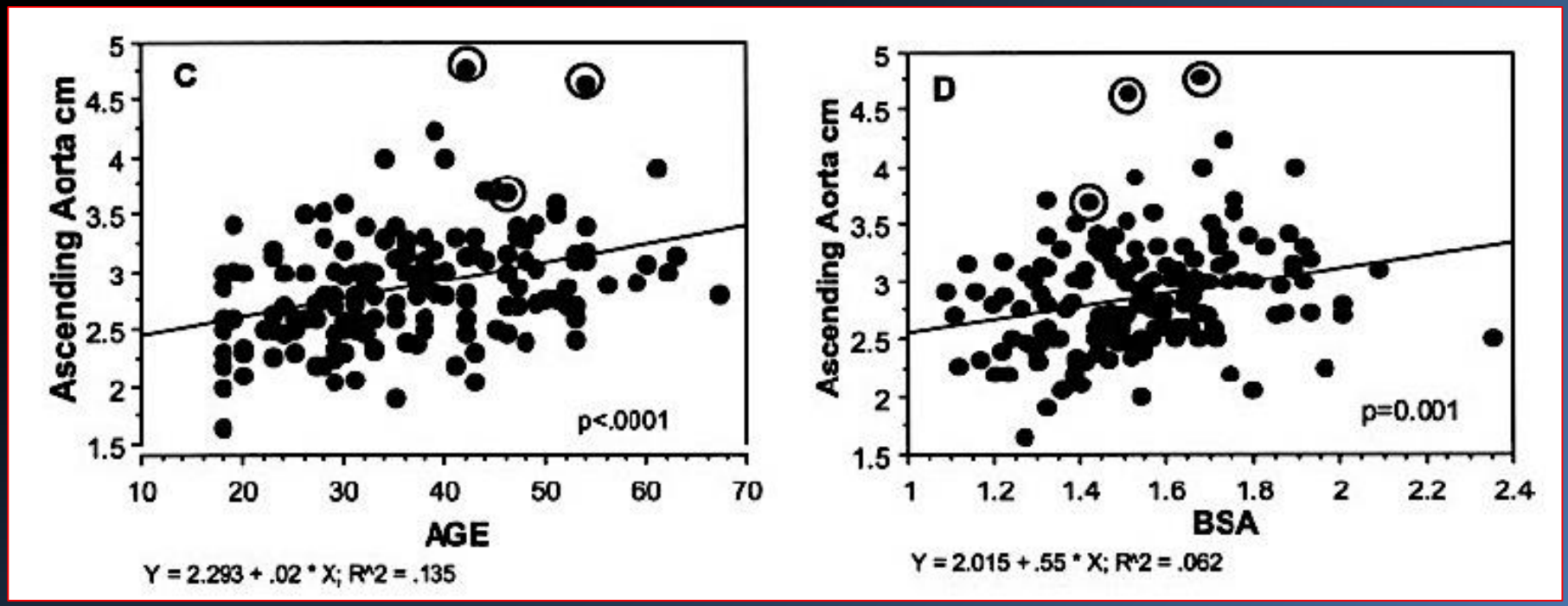
BAV indicates bicuspid aortic valve; TAV, tricuspid aortic valve; and ETA, elongated transverse arch of the aorta. Data are from TS subjects. Mean values compared by ANOVA, with age as the covariate. Cardiac measurements are normalized to BSA.

Matura et al. *Circulation.* 2007; 116: 1663-70

Henri Mondor

Aortic Dilatation and Dissection in Turner Syndrome

- After 2.9 years of FU: 3 cases of aortic dissection (aged 44, 47, and 57 years)
- Two had a bicuspid aortic valve, and all 3 had elongated transverse arches.
- All 3 women had ADs >3.5 cm and ASI >2.5 cm/m^2.



Matura et al. *Circulation.* 2007; 116: 1663-70

Type-A aortic dissection in patients with bicuspid valves: comparison with tricuspid aortic valves

- Observational study of all patients with confirmed BAV and AD from 1980–2010 (n=47), compared with a consecutive group with TAV and AD (n= 53, 2005-2010)

RESULTS: Patients with AD and BAV :

- Were younger
- Had less hypertension, more aortic valve stenosis and previous AVR
- Larger aortic dimension
- Worse aortic medial degeneration
- High prevalence of aortic coarctation
- 50% of patients with BAV had known aortic dilatation prior to AD

Table 1 Aortic dissection patient characteristics

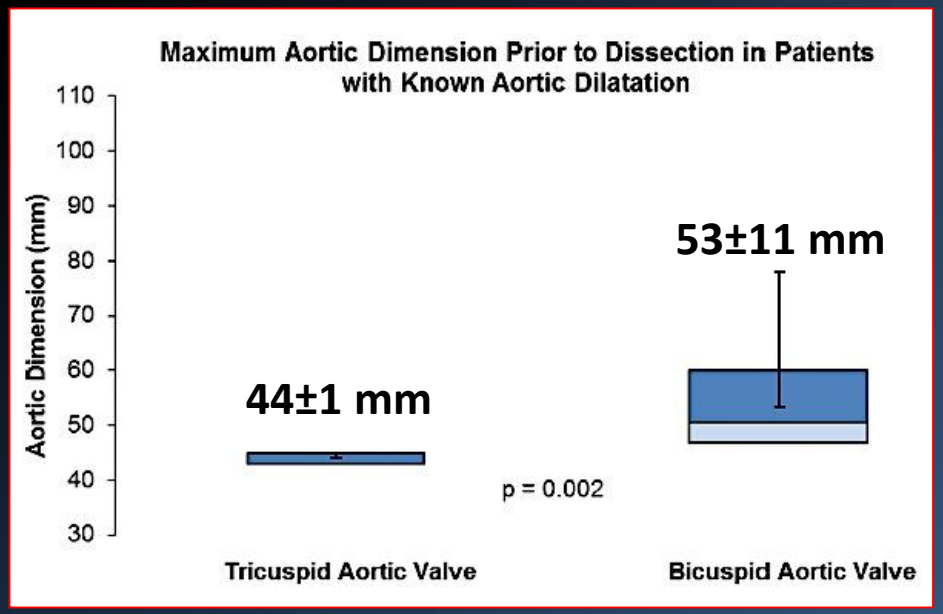
	Bicuspid aortic valve (N=47)	Tricuspid aortic valve (N=53)	p Value
Age	58±14	66±13	0.007
Male sex	36 (77%)	41 (77%)	1.0
Chronic dissection	16 (34%)	18 (34%)	1.0
Stanford type A	47 (100%)	53 (100%)	1.0
Max ascending aorta dimension (mm)	66±15	56±11	0.0004
Previous cardiac surgery	23 (49%)	23 (43%)	0.69
Previous aortic valve replacement	11 (23%)	3 (6%)	0.02
Previous coronary artery bypass grafting	12 (26%)	16 (30%)	0.67
Previous aortic dissection surgery	2 (4%)	5 (9%)	0.44
Hypertension	32 (68%)	46 (87%)	0.03
Active smoking	11 (23%)	7 (13%)	0.20
Previous smoking	20 (43%)	25 (47%)	0.69
Hyperlipidaemia	23 (49%)	26 (49%)	1.0
Diabetes mellitus	4 (9%)	6 (11%)	0.75
Coronary artery disease	18 (38%)	20 (38%)	1.0
Atrial fibrillation	1 (2%)	9 (17%)	0.02

Table compares baseline clinical characteristics between patients with BAV and patients with TAV presenting with aortic dissection.
BAV, bicuspid aortic valve; Max, maximum; TAV, tricuspid aortic valve.

Eleid et al. *Heart.* 2013; 99: 1668–74

Type-A aortic dissection in patients with bicuspid valves: comparison with tricuspid aortic valves

- Of the 23 patients with BAV with known aortic dilatation, maximal diameters were ≥55 mm in 5 patients (22%), 50–54 mm in 6 (26%)
- 7 of 11 patients had significant aorta dilatation at the time of previous AVR (diameter ≥45 mm and/or ≥moderate aortic dilatation by direct surgical inspection)



CONCLUSION: Implementation of current guidelines could have theoretically prevented aortic dissection in several patients with BAV

Eleid et al. *Heart.* 2013; 99: 1668–74