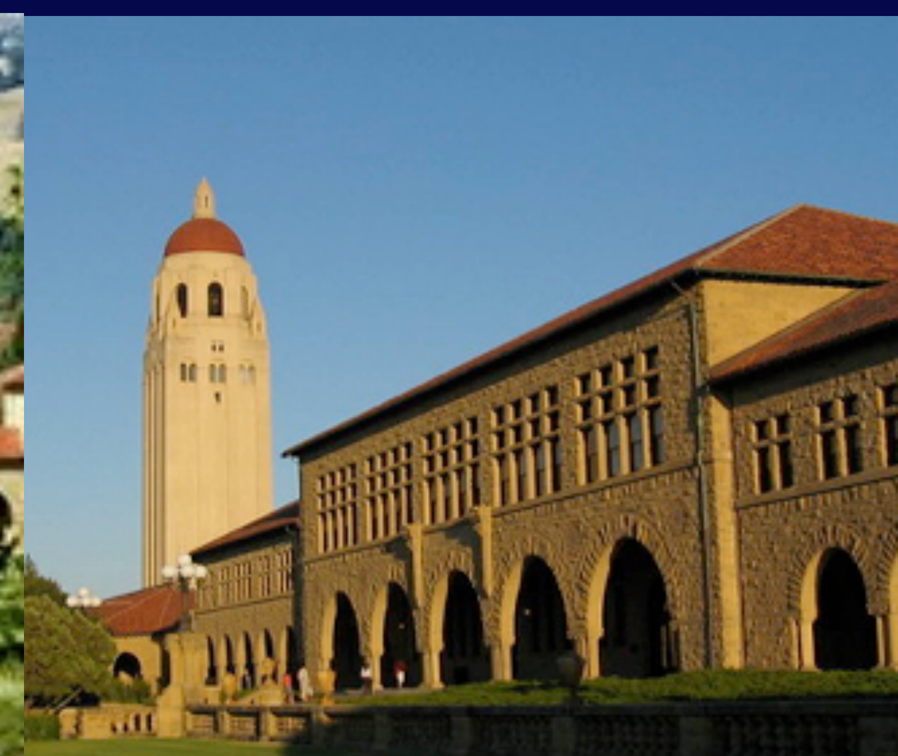
Echocardiography of the Aortic Valve





Norman H Silverman MD. D Sc (Med), FACC, FASE, FAHA.

Professor of Pediatrics (Cardiology) U.C. San Francisco,

Professor Emeritus, Stanford University

Clinical Significance Of BAV

- A bicuspid aortic valve is found in most (~50-60%) patients with aortic stenosis (Edwards WD, Mayo Clin Proc 1987, Davies et al, Heart, 1996)
- Aortic valve disease is the third most common cause of life threatening cardiovascular conditions in the western world (after systemic hypertension and atherosclerosis)
- Aortic valve replacement is the second most common cardiac surgery (after coronary bypass)

BAV & Dilatation of the Ascending Aorta

In 1928, Abbot theorized that bicuspid aortic valve, coarctation of the aorta, aortic wall thinning, which led to rupture were related to a common developmental abnormality

Abbot, Am Heart J, 1928

Aortic Dissection

- The incidence of a bicuspid valve is 28% in patients with aortic dissection younger than 40 years of age!

Gore, Arch Pathol, 1953

Clinical Significance of BAV

- A bicuspid aortic valve is found in most (~50-60%) patients with aortic stenosis (Edwards WD, Mayo Clin Proc 1987, Davies et al, Heart, 1996)
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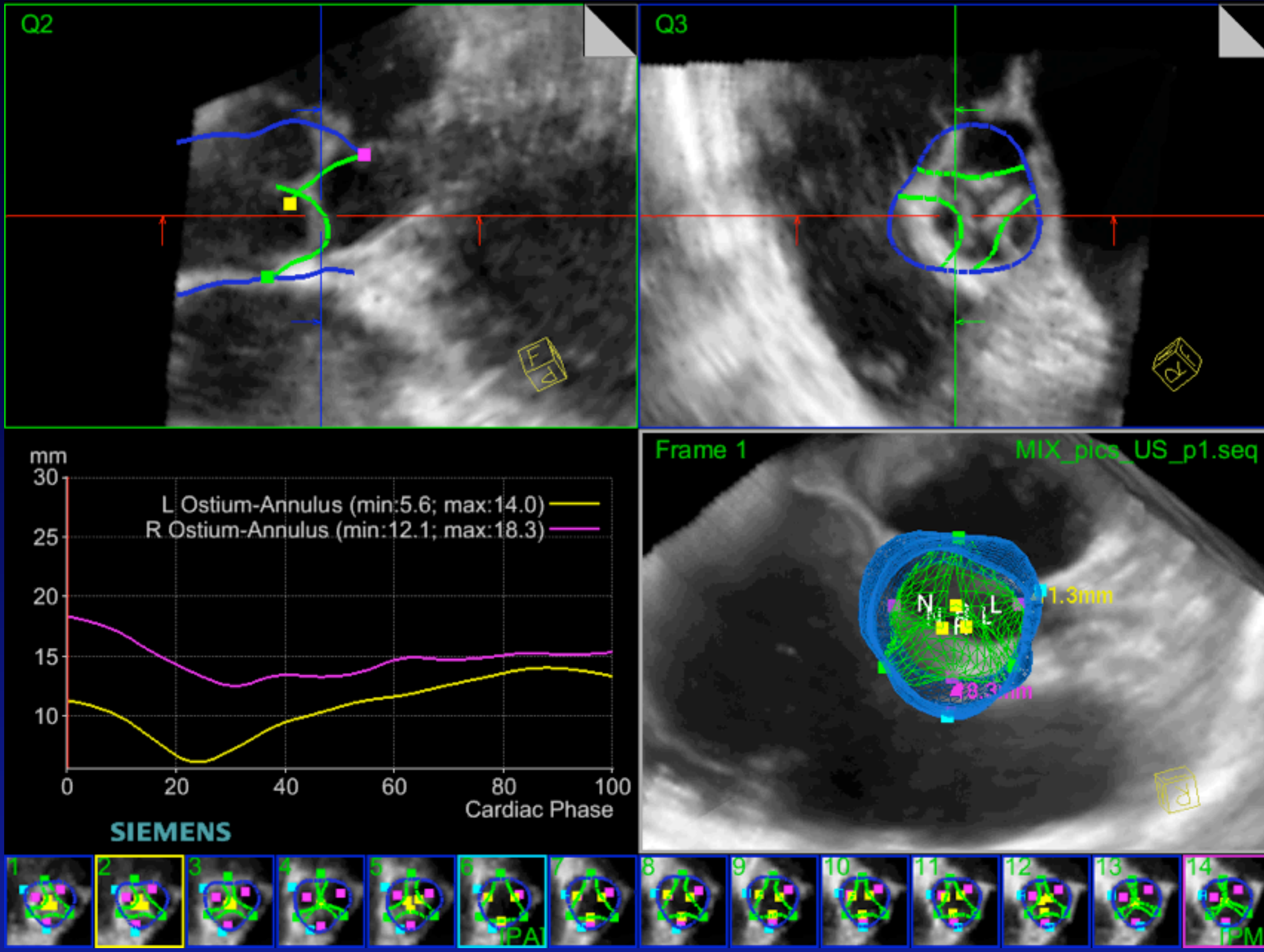
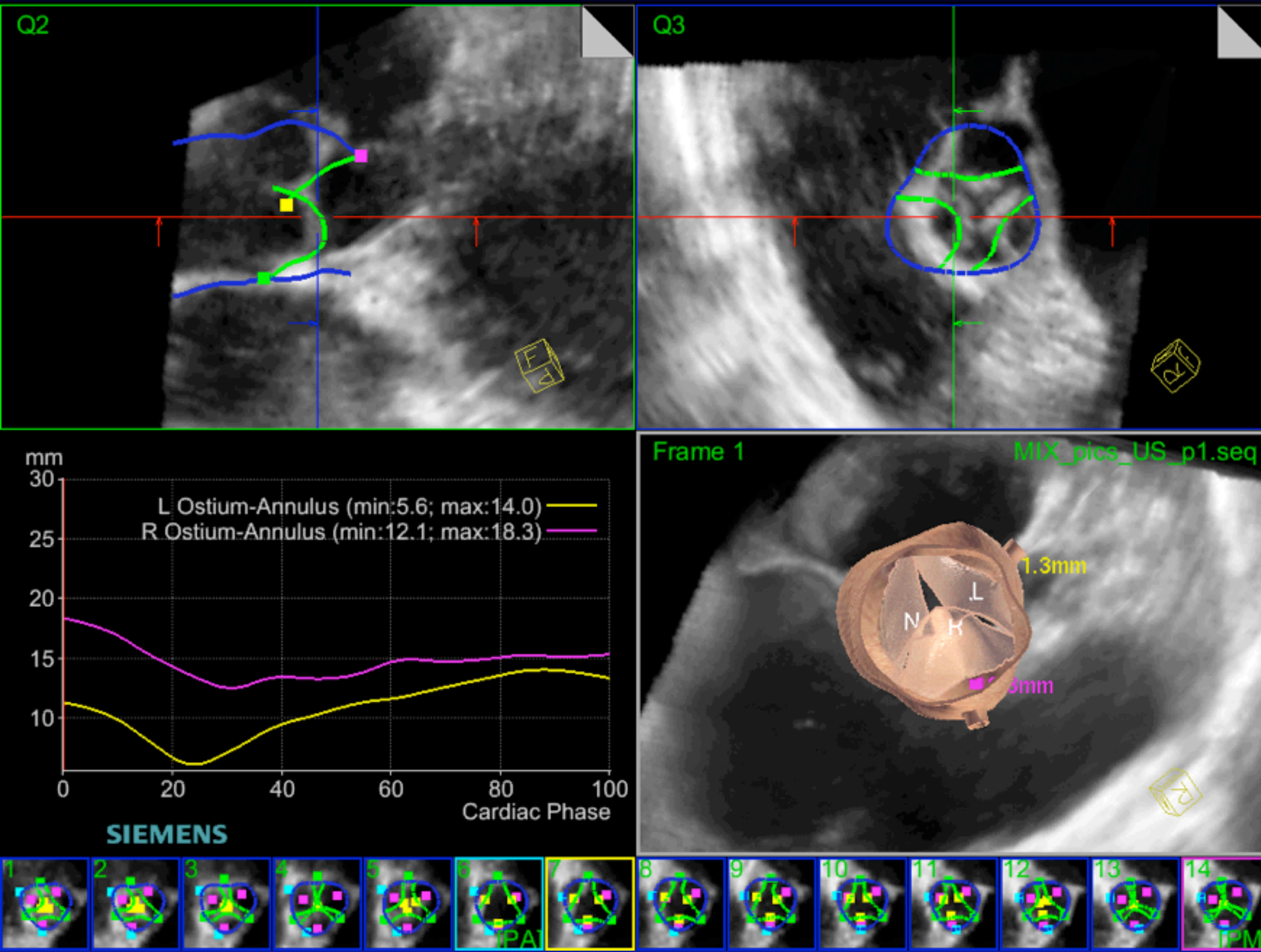
New Real-Time Valvar Functional Reconstruction



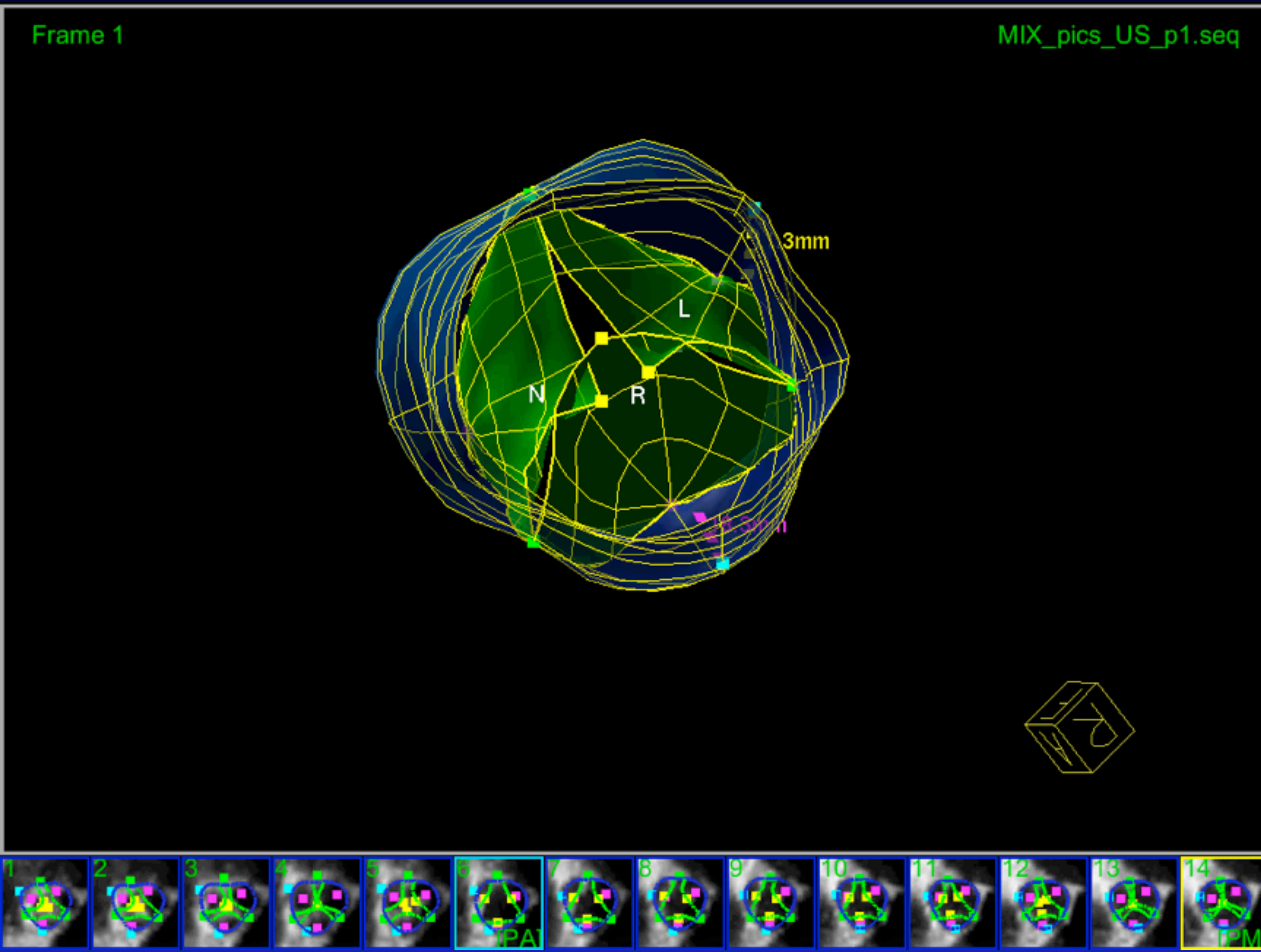
Courtesy of David Roberson

New Real-Time Valvar Functional Reconstruction



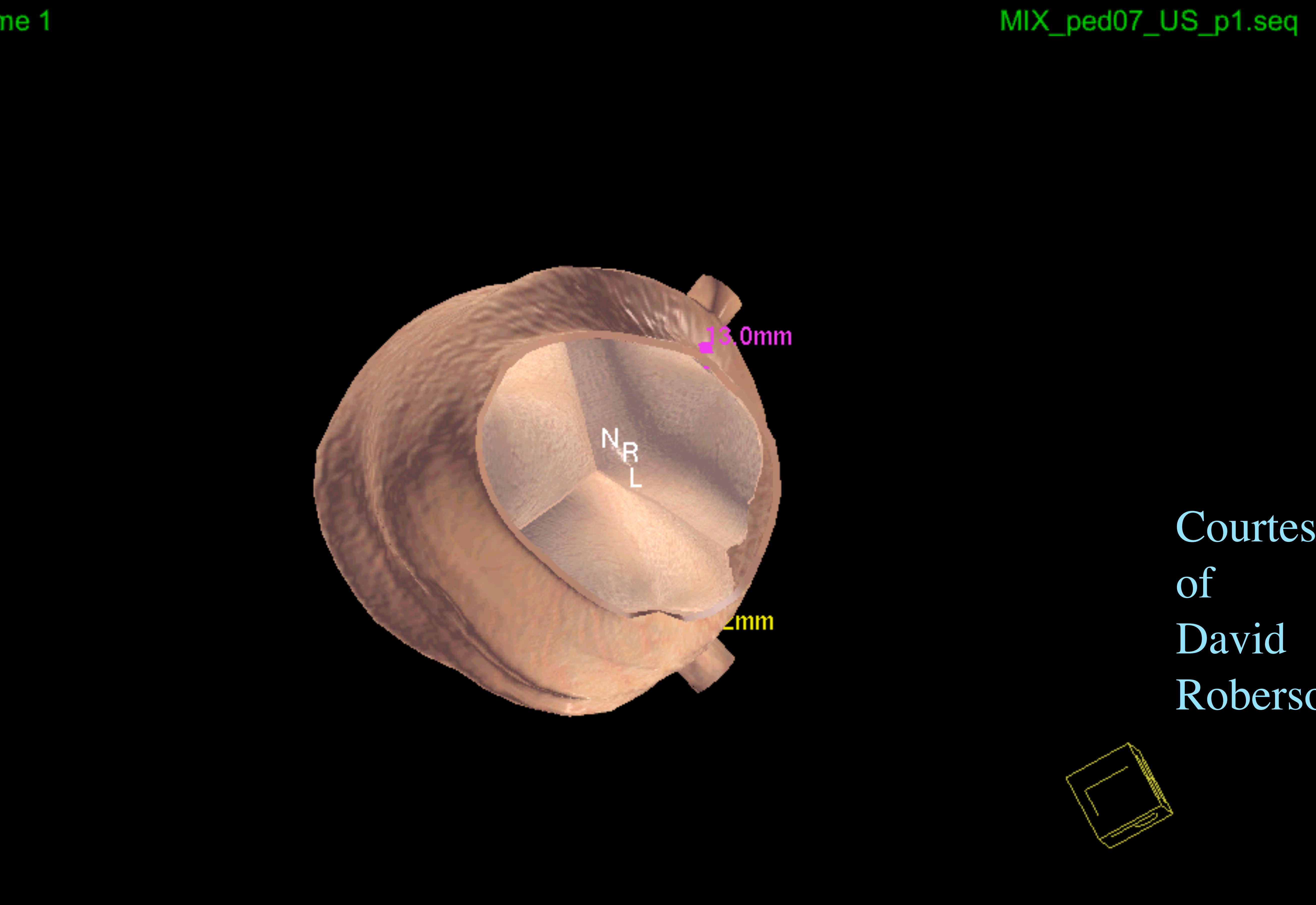
Courtesy of David Roberson

New Real-Time Valvar Functional Reconstruction



Courtesy of David Roberson

New Real-Time Valvar Functional Reconstruction



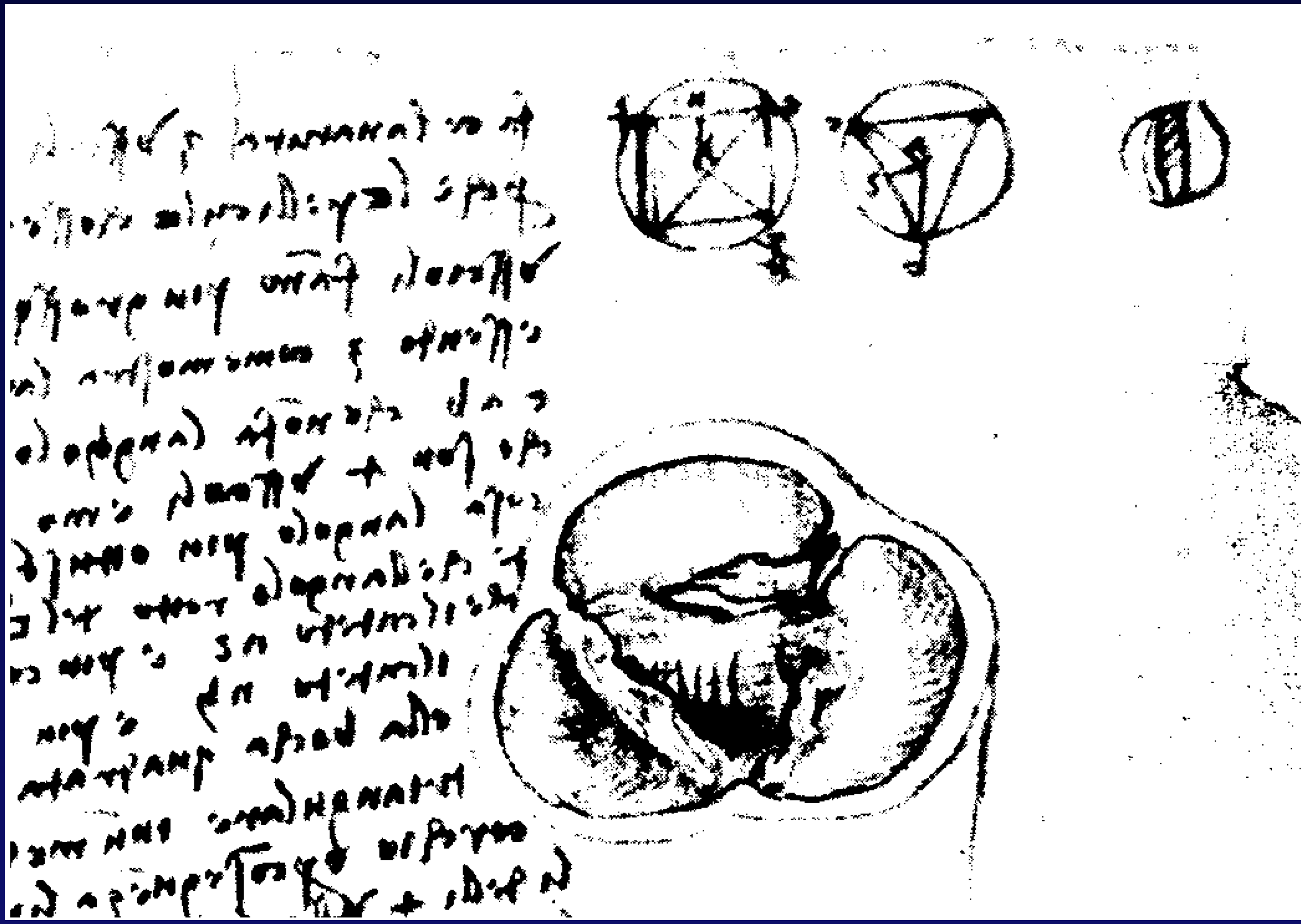
Courtesy of David Roberson

Bicuspid Aortic Valve

- AORTIC COARCTATION
- AORTIC ROOT DILATATION
- AORTIC DISSECTION
- CERVICOCEPHALIC ARTERIAL DISSECTION
- CYSTIC MEDIAL NECROSIS
- MITRAL AND OTHER DISEASES

Bicuspid Aortic Valve:Historical Perspectives

~1500: Leonardo da Vinci sketched aortic valves with 2, 3 and 4 cusps. Concluded 3-cusp valve had optimal function



1844: Paget described BAV as especially liable to disease

1868: Peacock described aortic stenosis in BAV

1886: Osler reported BAV as particular liability to develop infective endocarditis.

Mills P. Br Heart J 1978;40:951-7
Roberts WC. Am J Cardiol 1970;20:70-83

Genetics of the Bicuspid Aortic Valve

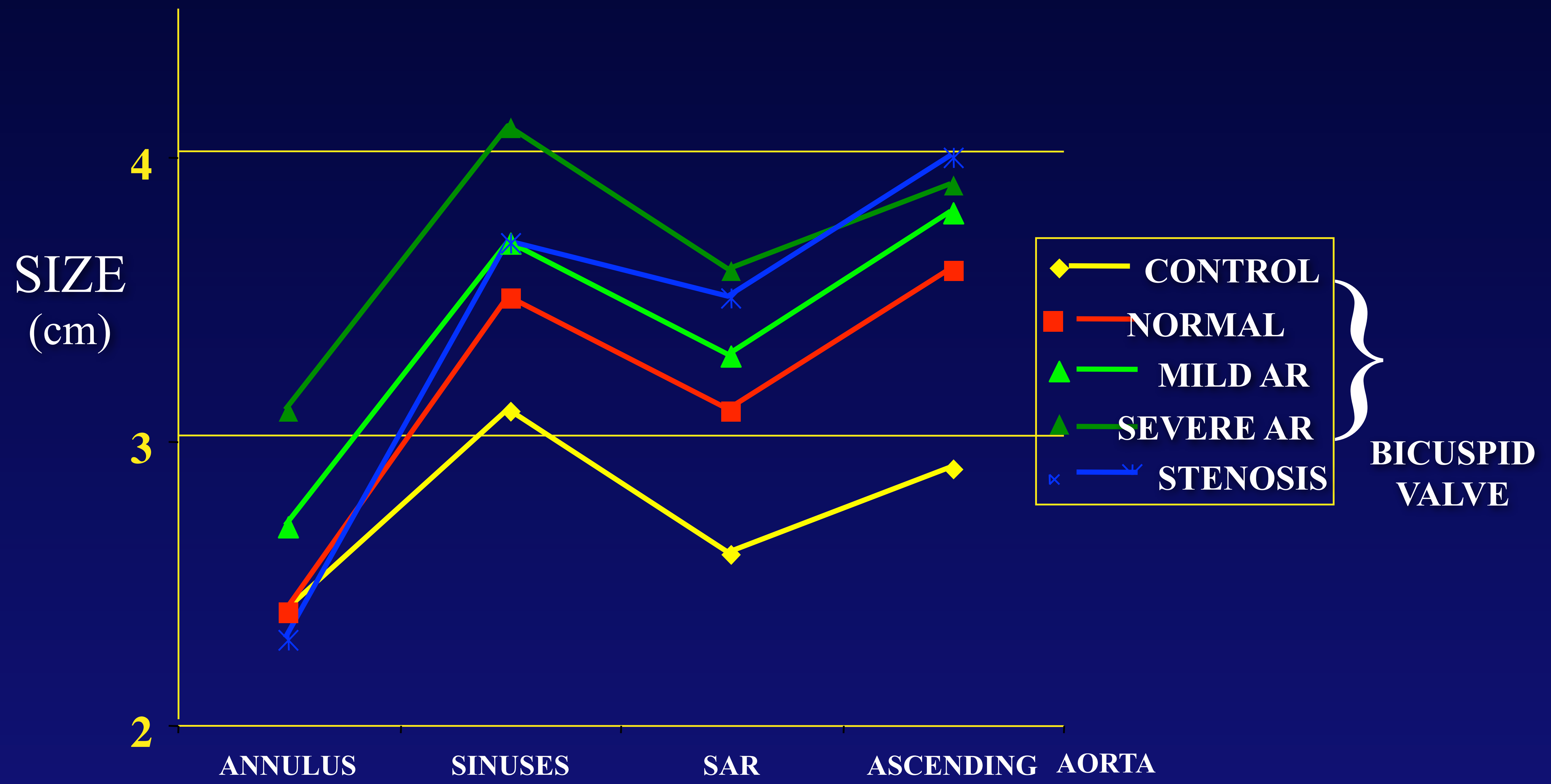
- ♥ The specific gene loci or products responsible for the development of BAVs, whether structural proteins or ones with vital roles in cardiac development, have yet to be discovered.
- ♥ Animal models with BAV have provided insight into potential pathogenetic mechanisms, including abnormalities in eNOS, NKX2.5 and *NOTCH1* signaling.
- ♥ Human studies have demonstrated the genetic influences on left-sided outflow lesions including hypoplastic left heart and BAV.
- ♥ Animal models with BAV have provided insight into potential pathogenetic, including abnormalities in eNOS, NKX2.5 and *NOTCH1* signaling.

Bicuspid Aortic Valves With Different Spatial Orientations of the Leaflets Are Distinct Etiological Entities

(J Am Coll Cardiol 2009;54:2312–8)

Borja Fernández, PHD,* Ana C. Durán, PHD,* Teresa Fernández-Gallego, BSC,*
M. Carmen Fernández, BSC,* Miguel Such, MD,† Josep M. Arqué, MD,‡ Valentín Sans-Coma, PHD*
Málaga, Spain

Aortic Root Dimensions In Patients With Bicuspid AoV

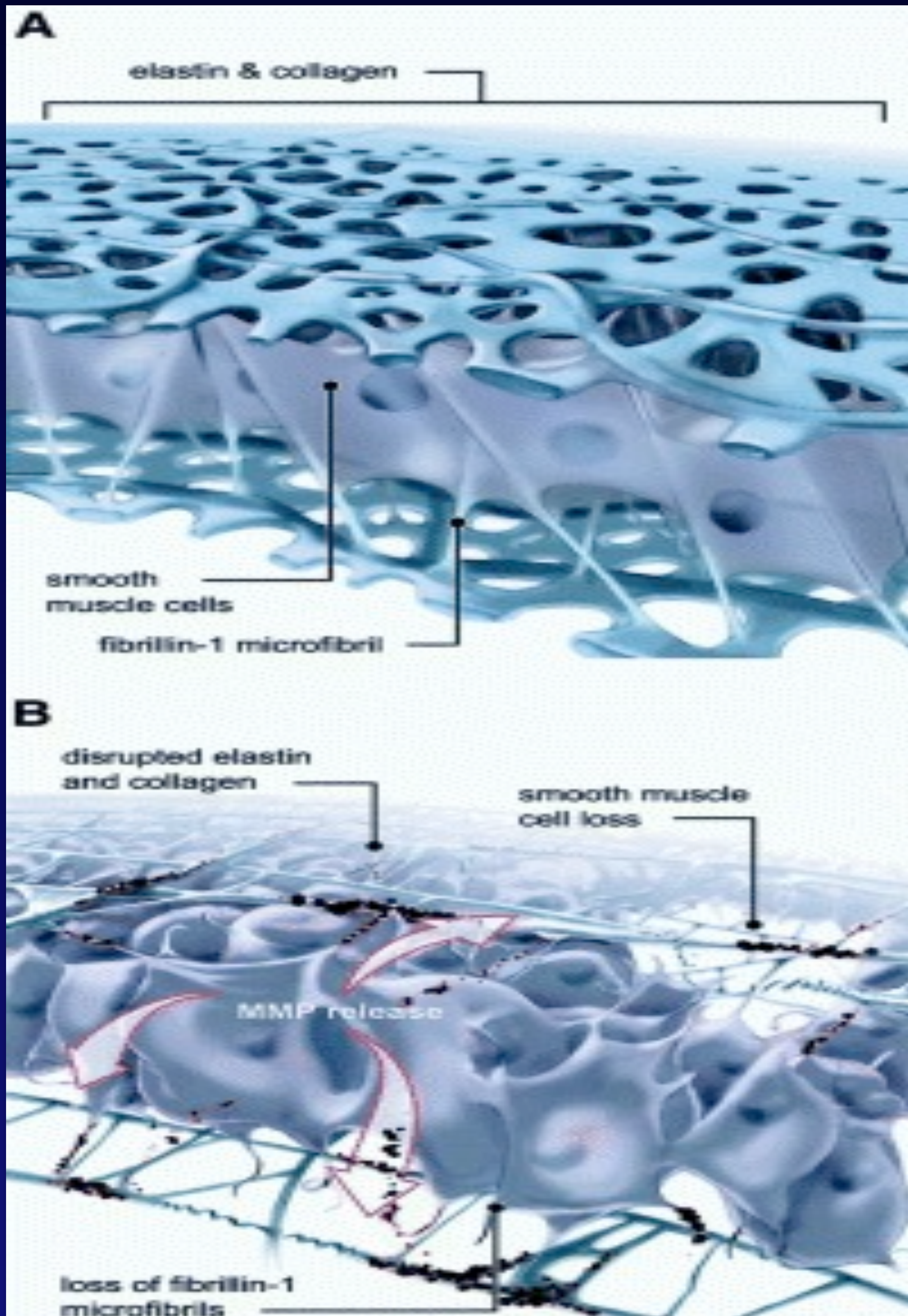


Adapted from: Hahn et al, JACC, 199

Pathogenesis of Bicuspid Aortic Valve: Genetic Etiology

TAV

BAV



- Differentiation of cushion mesenchymal cells into mature valve cells correlates with the expression of the microfibrillar proteins, fibrillin and fibulin.
- Microfibrillar elements within the aortic matrix may be deficient resulting in SMC detachment, MMP release, matrix disruption, cell death and loss of structural support and elasticity.
- Defects in the genes or transcriptional elements that encode protein production may be defective in BAV pts.

Four Dimensional Echocardiography

Courtesy of Philips Ultrasound

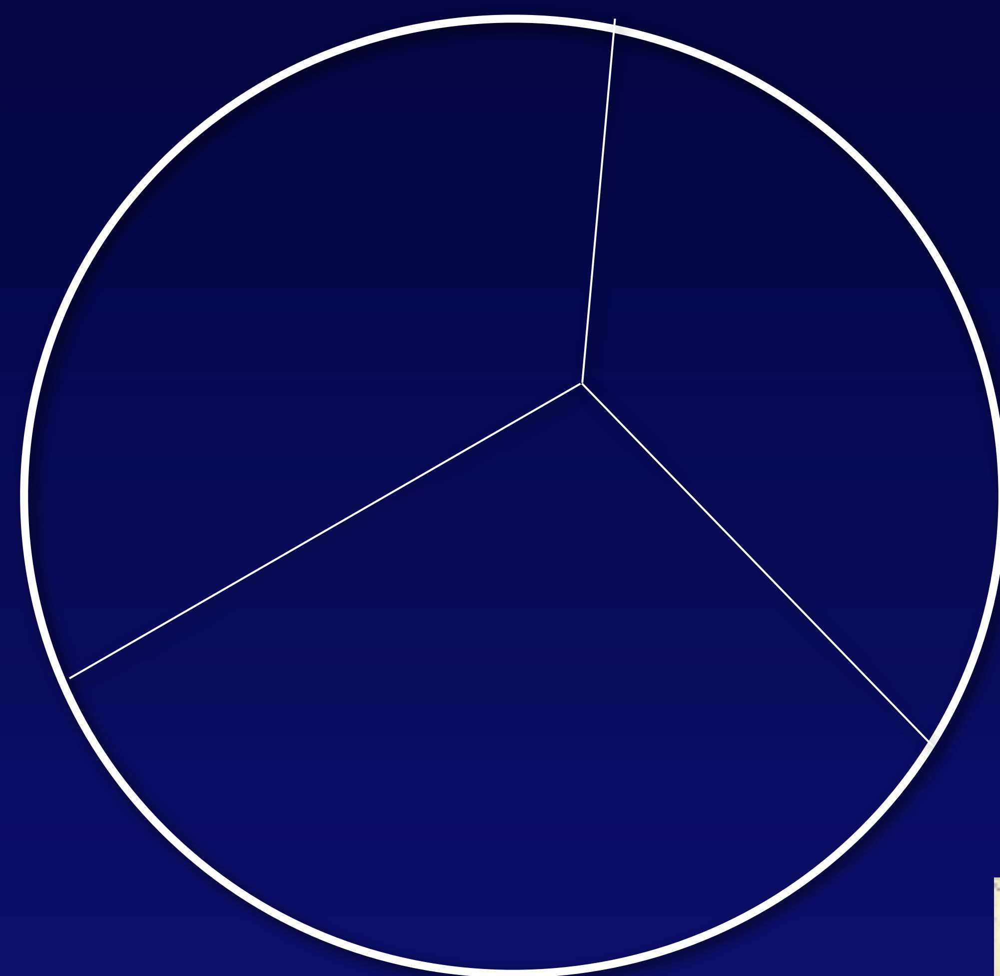
Morphology and Function of the Bicuspid Aortic Valve With and Without Coarctation of the Aorta in the Young

Giovanna R. Ciotti, MD[a,*], Antonios P. Vlahos, MD[b], and Norman H. Silverman, MD, DSc (Med)[c]

(Am J Cardiol 2006;98:1096–1102)

The Area of the Normal Aortic Valvar Cusps

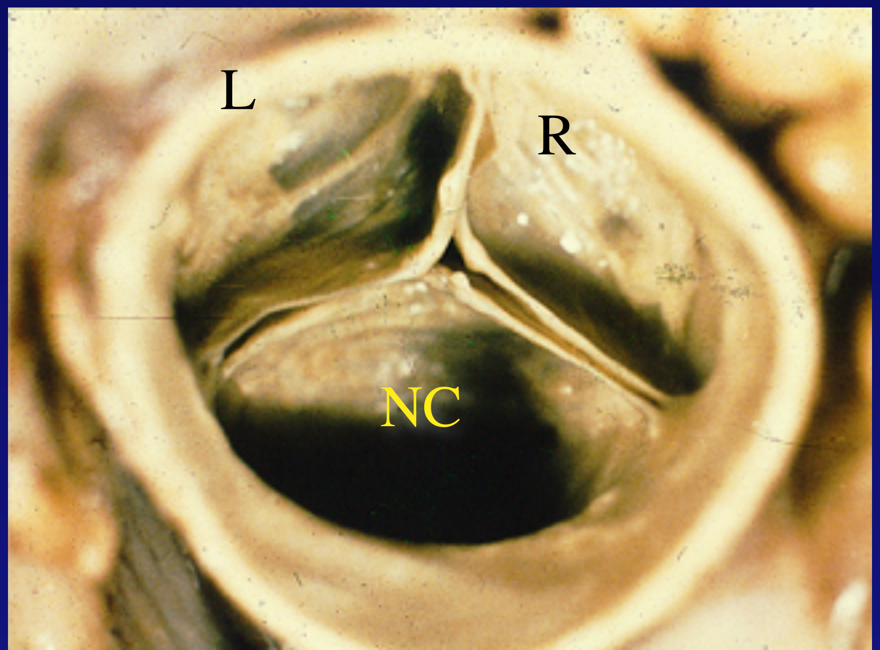
Right Coronary
32 ± 0.69%



Left Coronary
28 ± 0.46%

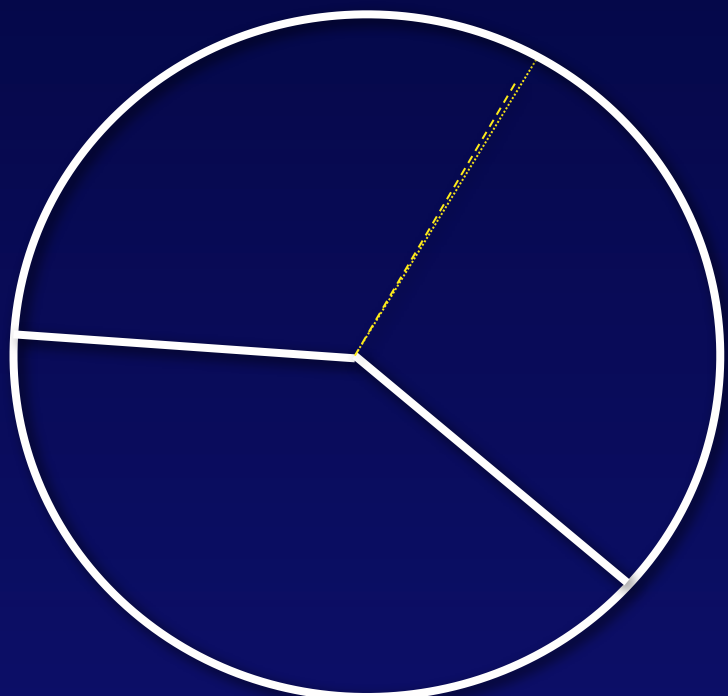
Non-Coronary
39 ± 11%

This is Not a Mercendes Benz



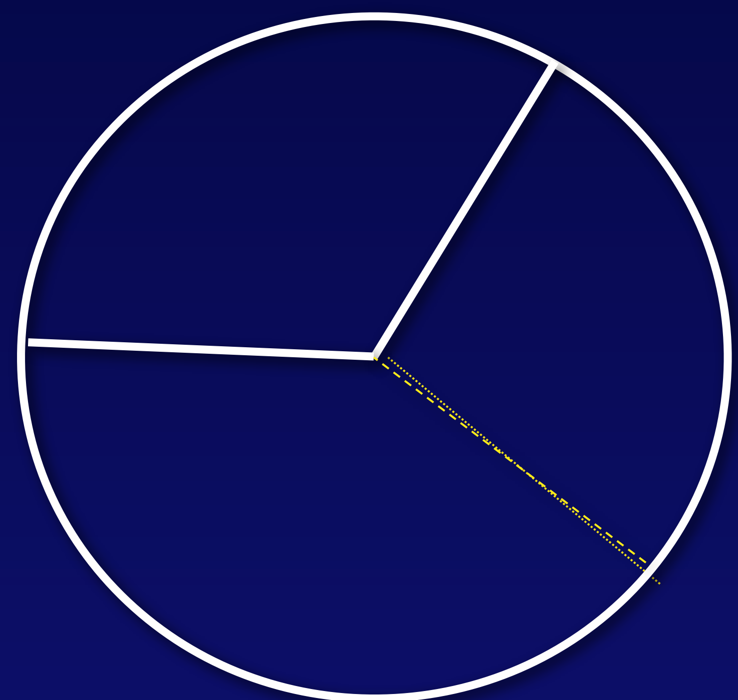
Types of Fusion of Aortic Valve Leaflets

Left- Right Fusion



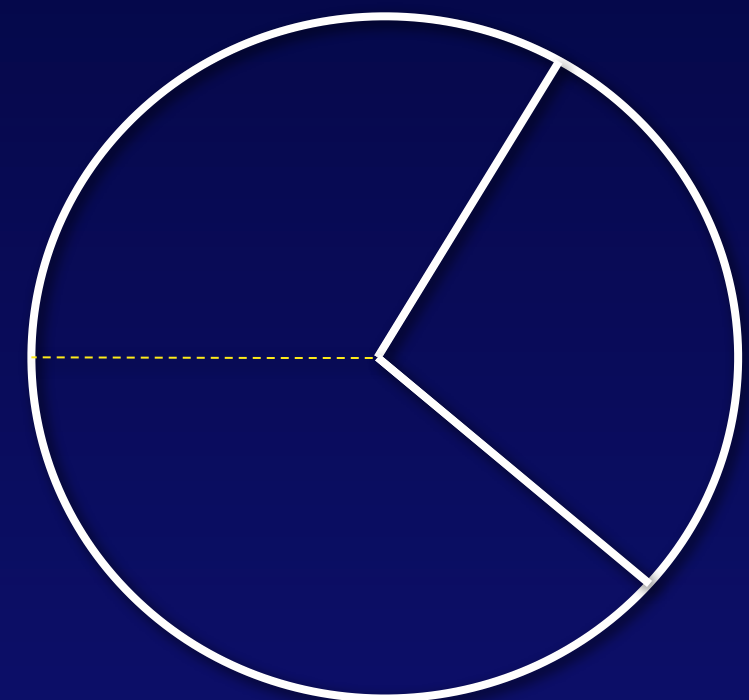
59%

Left- Non Fusion



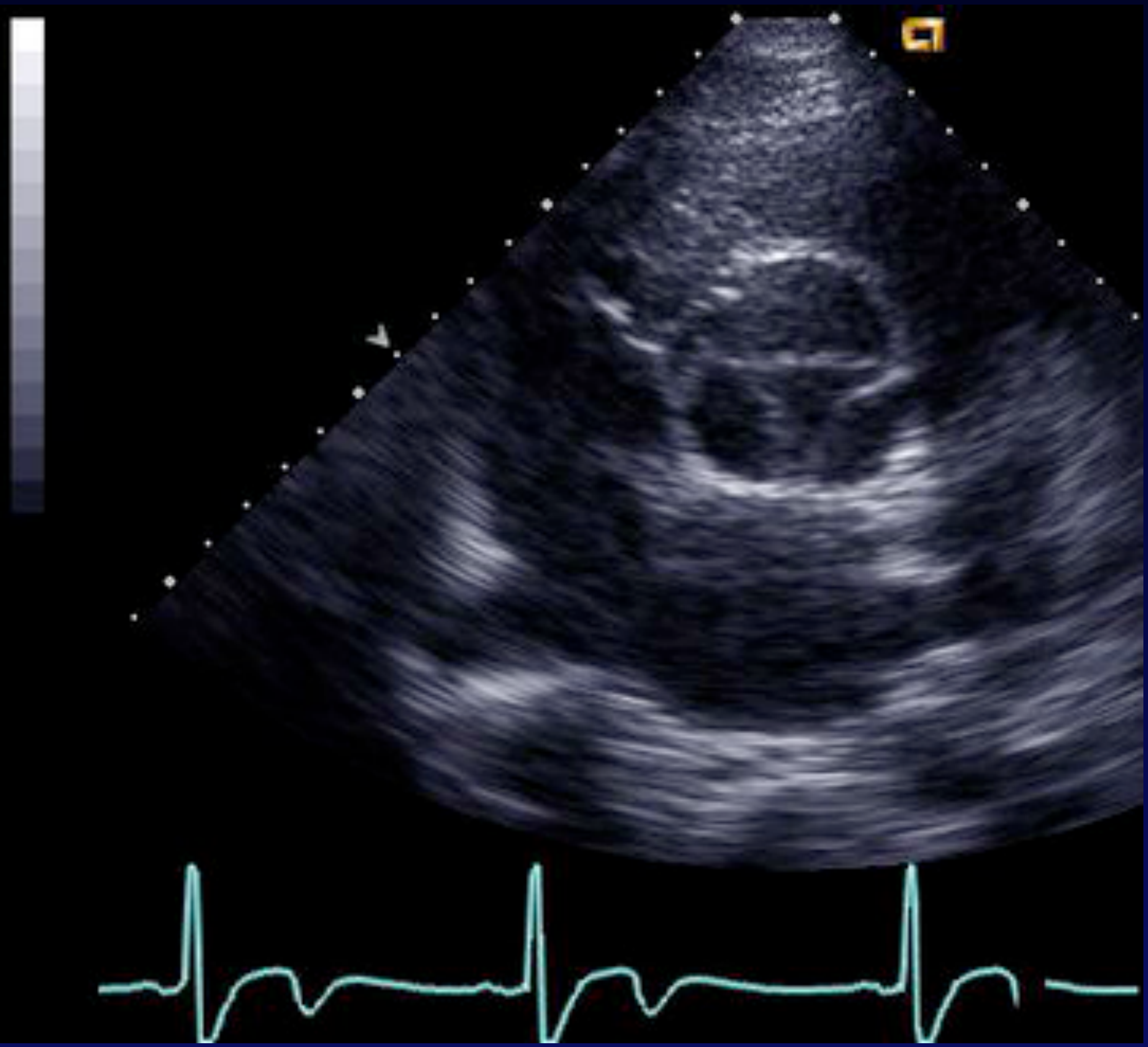
4%

Right - Non Fusion



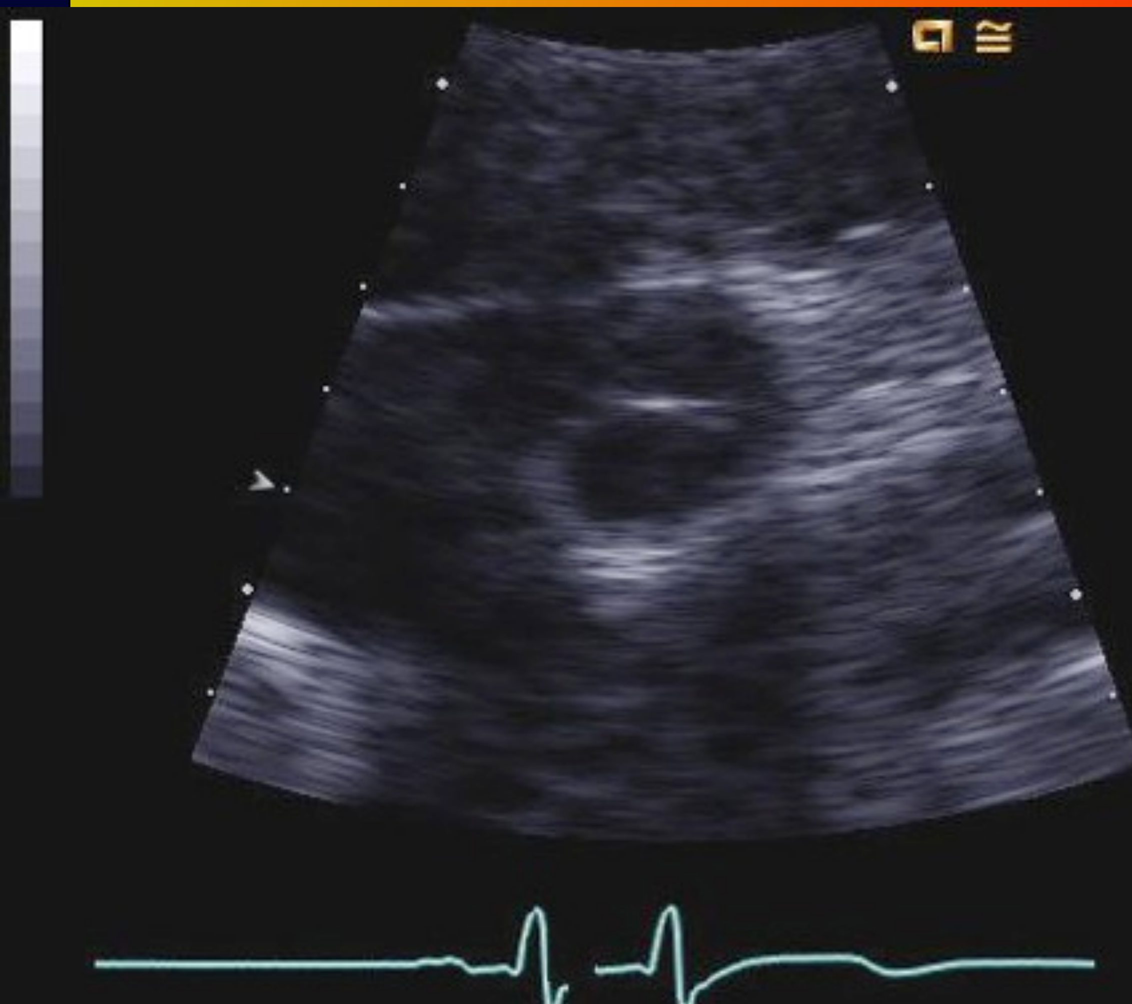
37%

Normal Aortic Valve
Parasternal Short Axis/ TEE



NHS.

Bicuspid Aortic Valve



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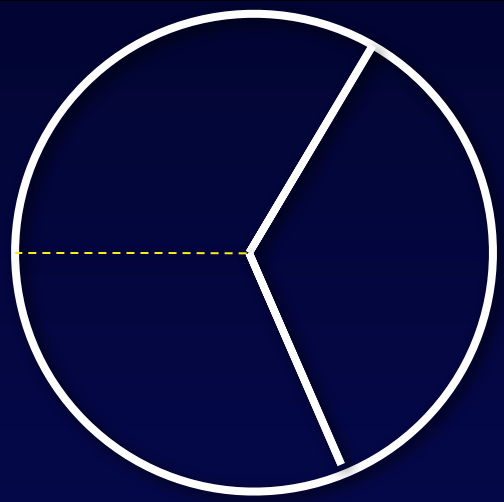
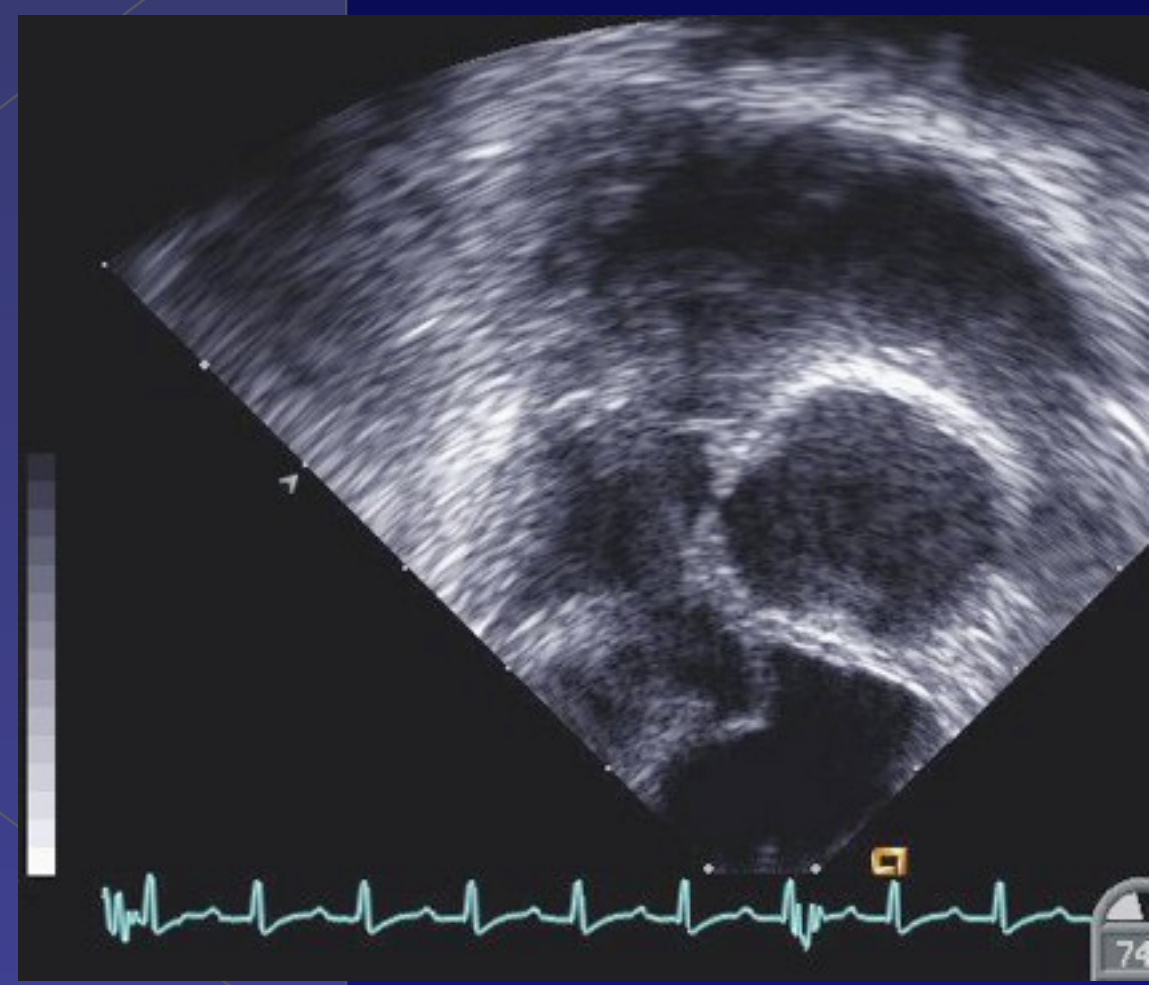
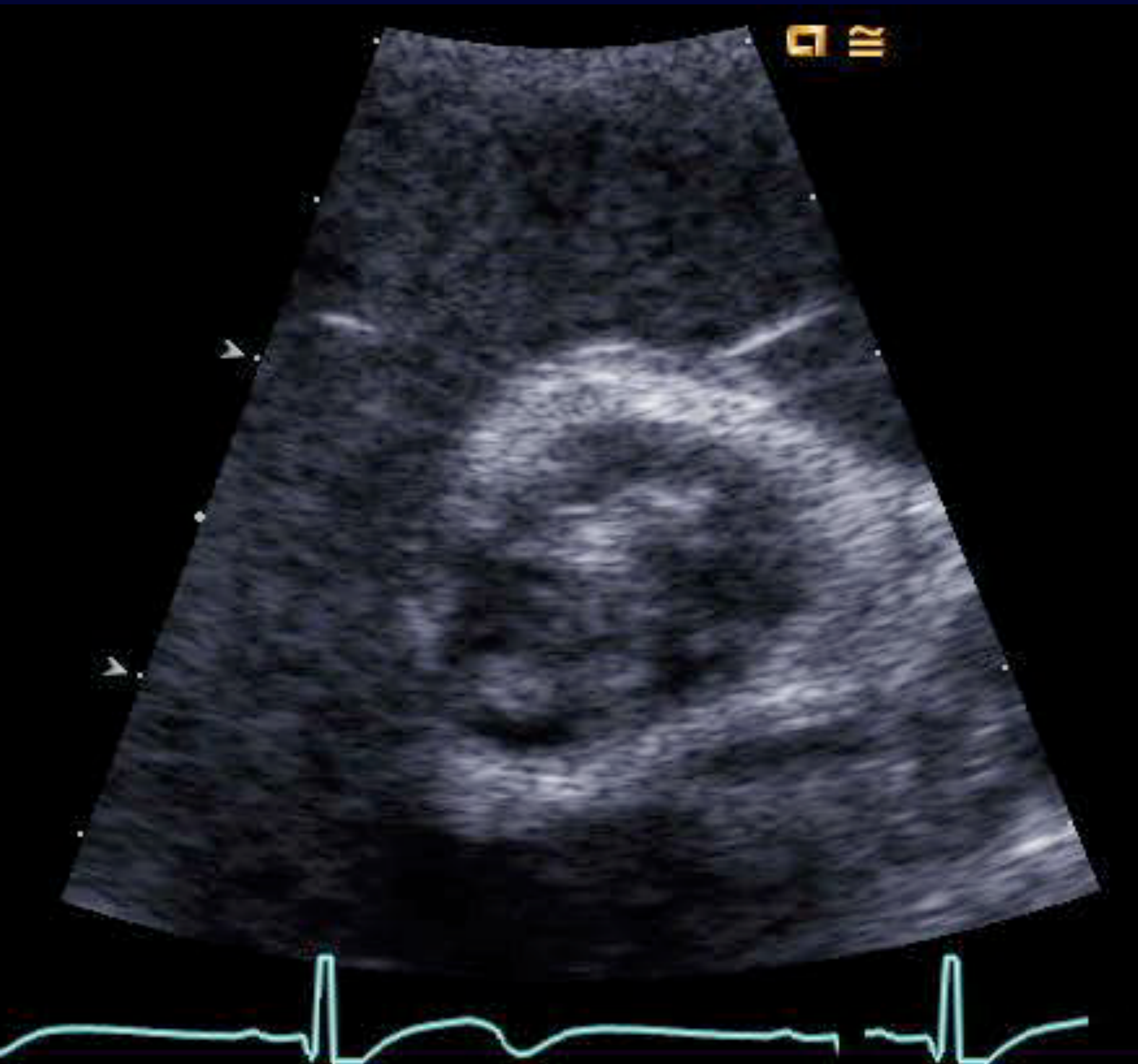
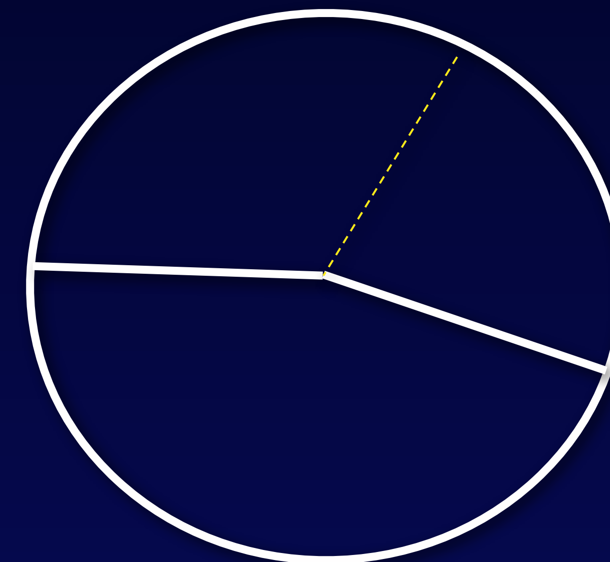
Right- Non Coronary Leaflet Fusion.

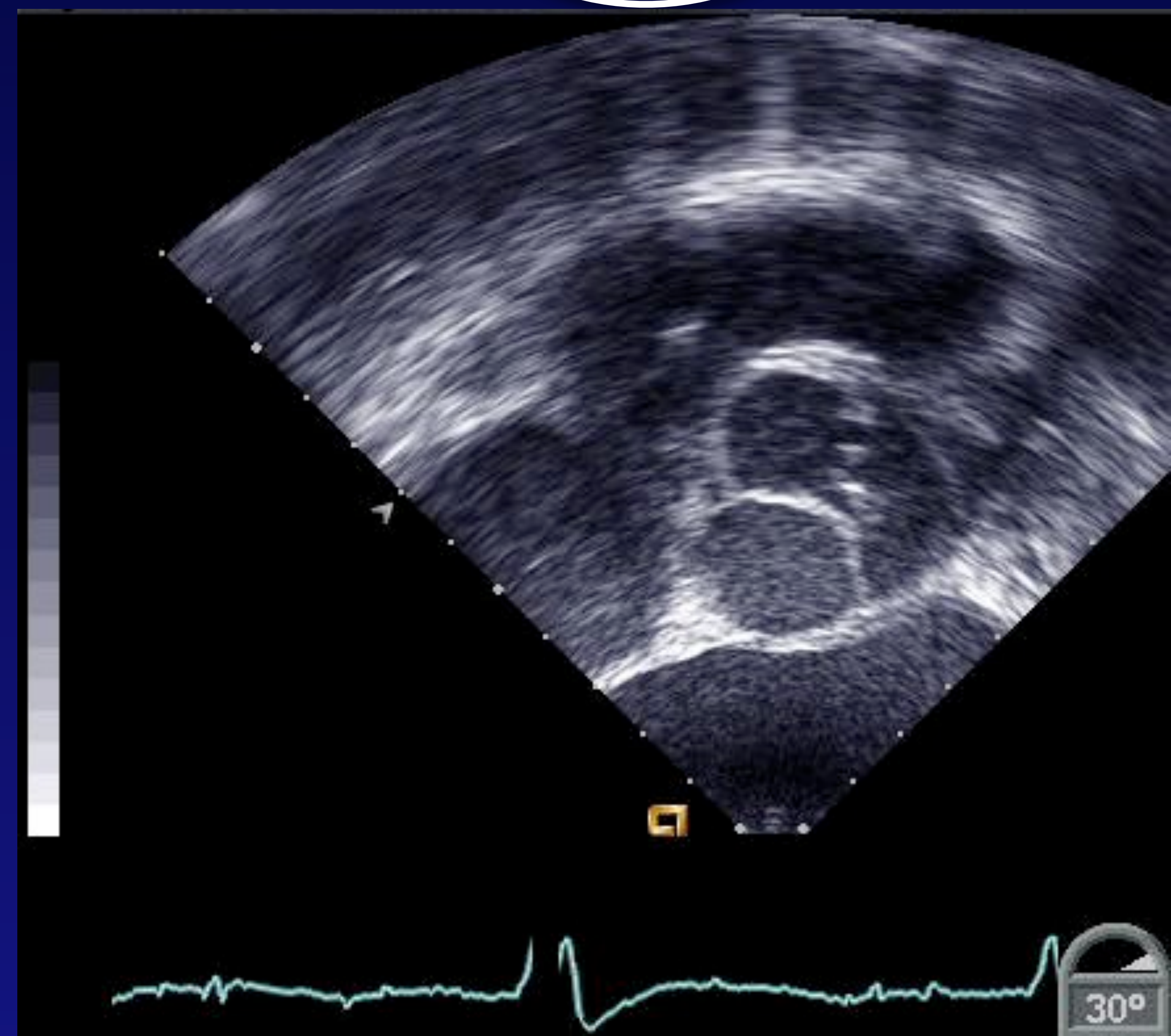




Left Right Cusp Fusion

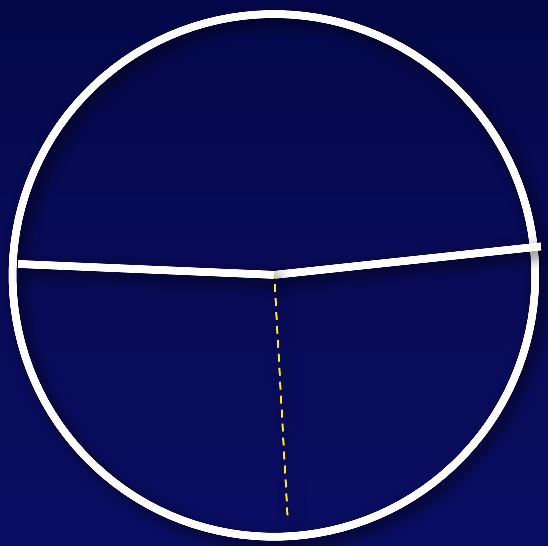


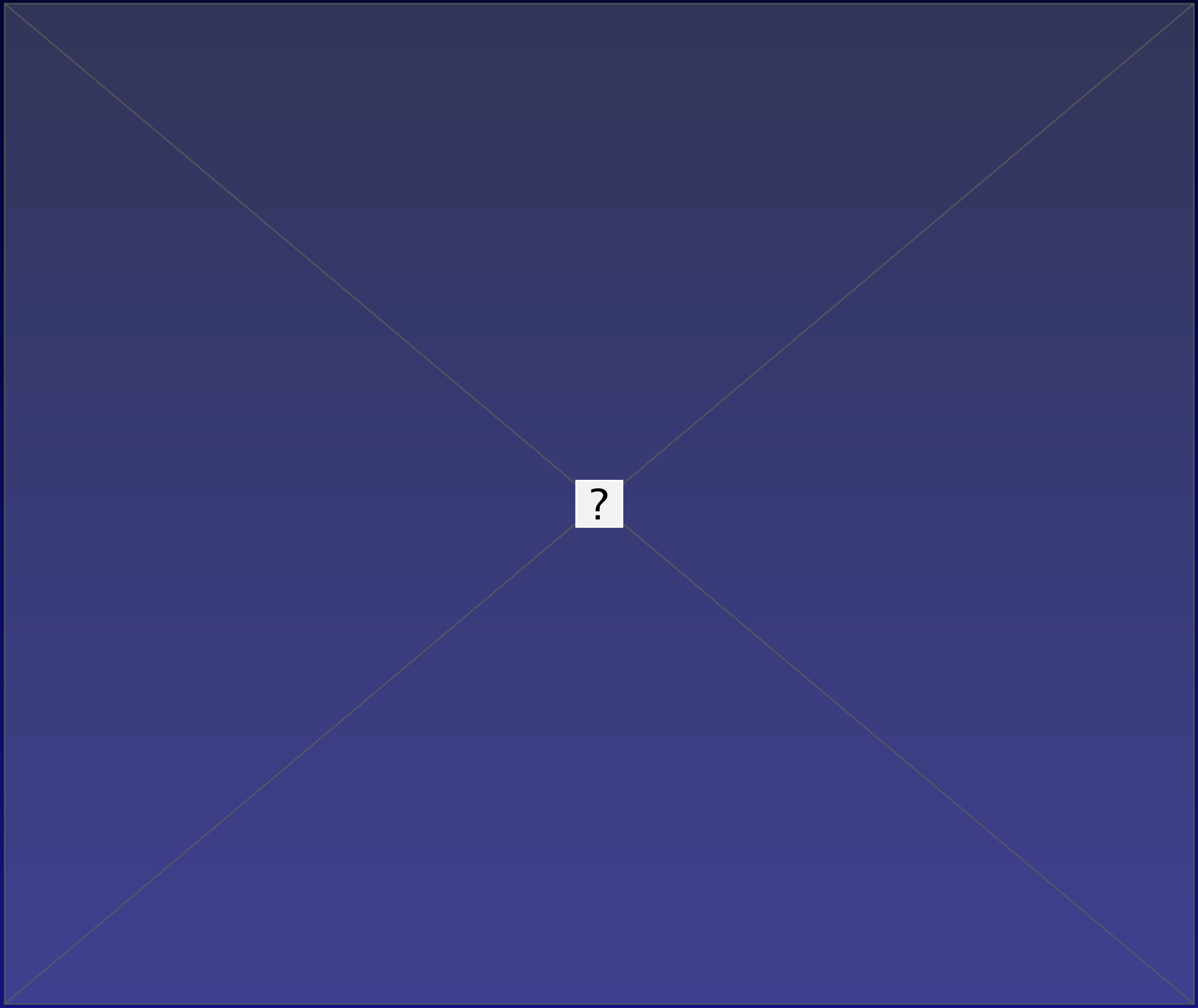




The leaflet opposite the fused leaflets is larger than the corresponding unfused leaflet.

Left- Non Coronary Leaflet Fusion.





Study Population n=139

117 Bicuspid Ao Valve

62 Aortic Coarctation

Bicuspid Valve in CoA= 62.5%

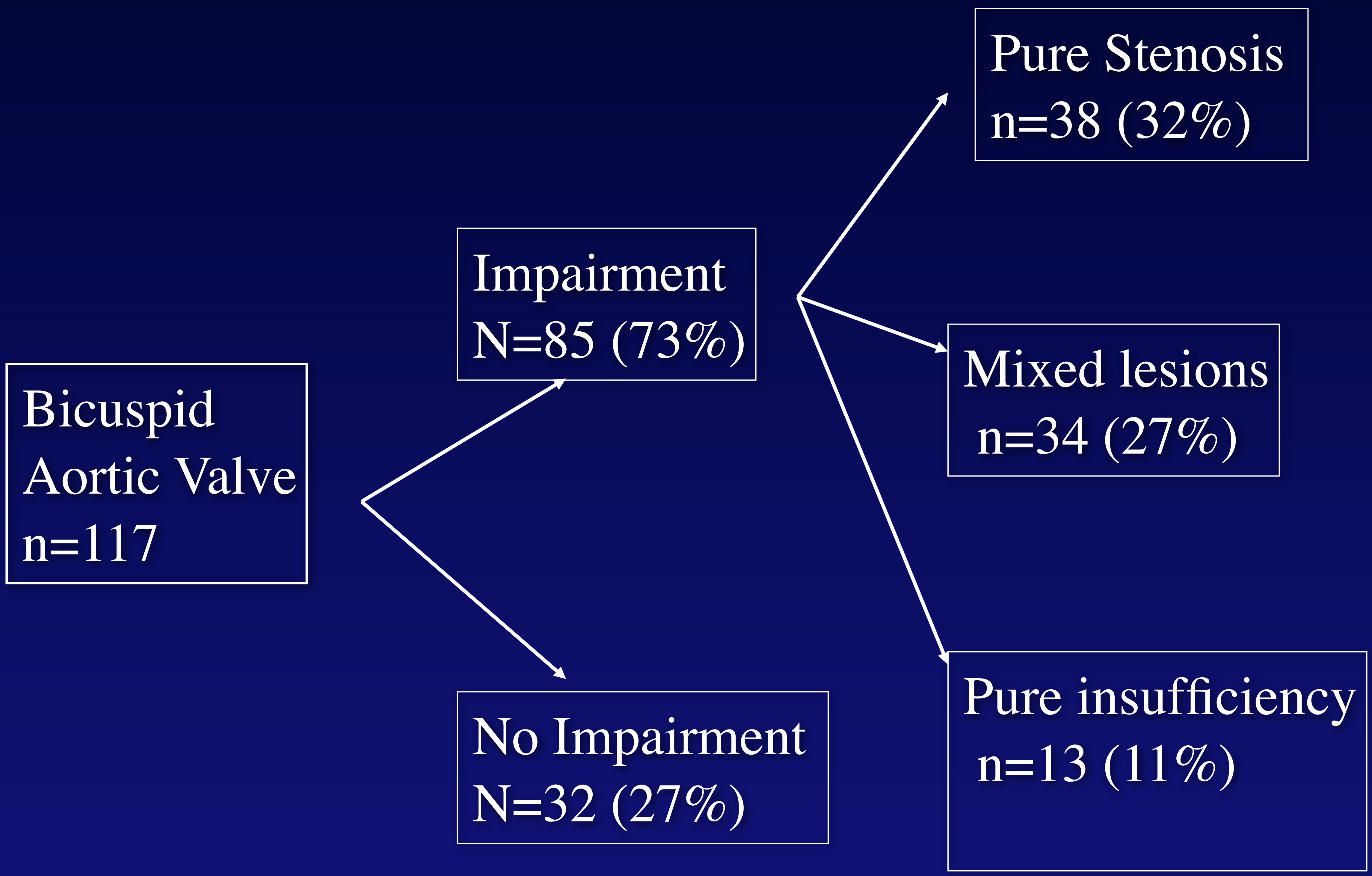
77 ø Coarctation

40 with Bicuspid aortic valve

22 with tricuspid aortic valve

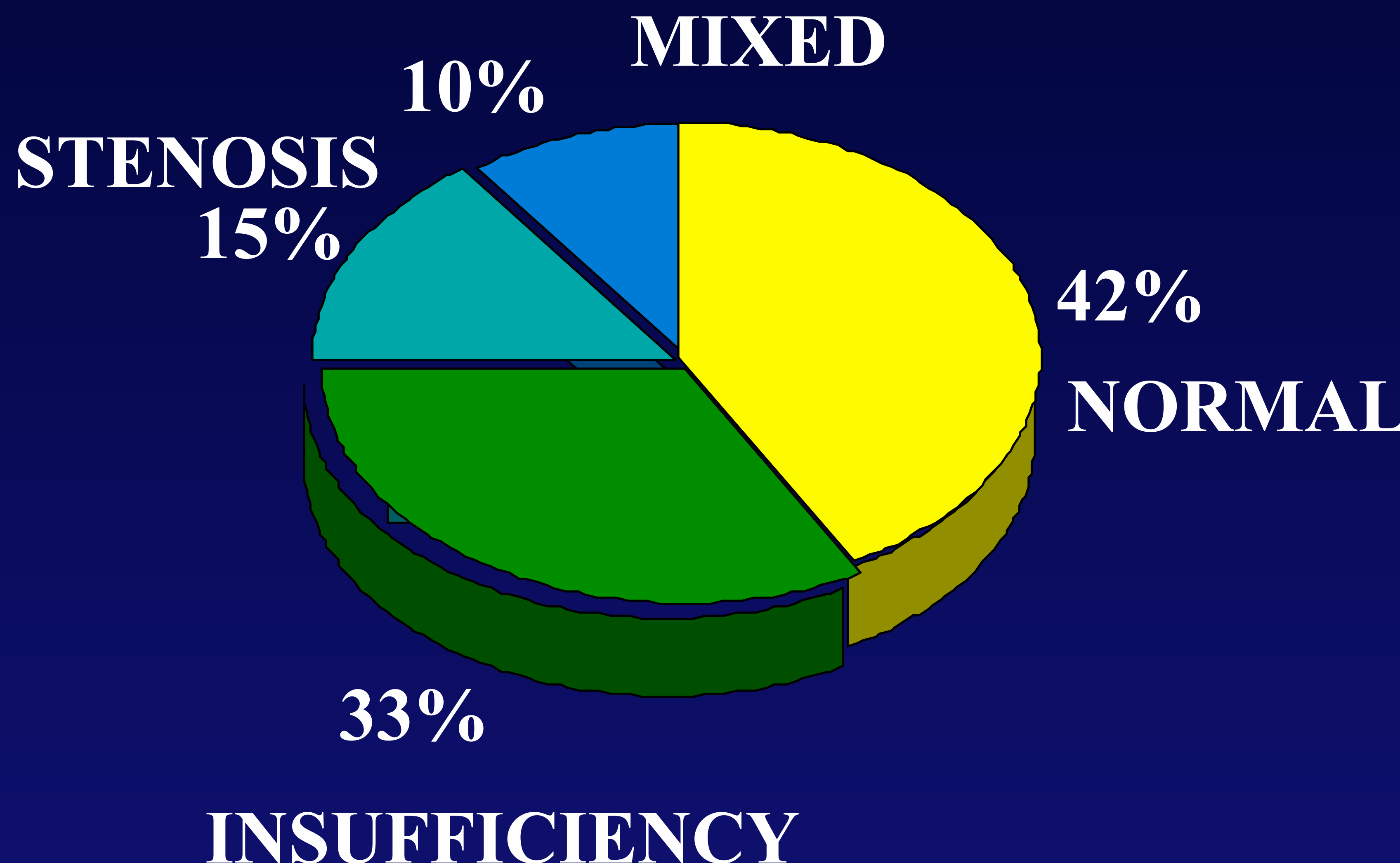
NHS.

Function of the Bicuspid Aortic Valve



NHS.

Valve Function : Adults <50 Yrs Of Age



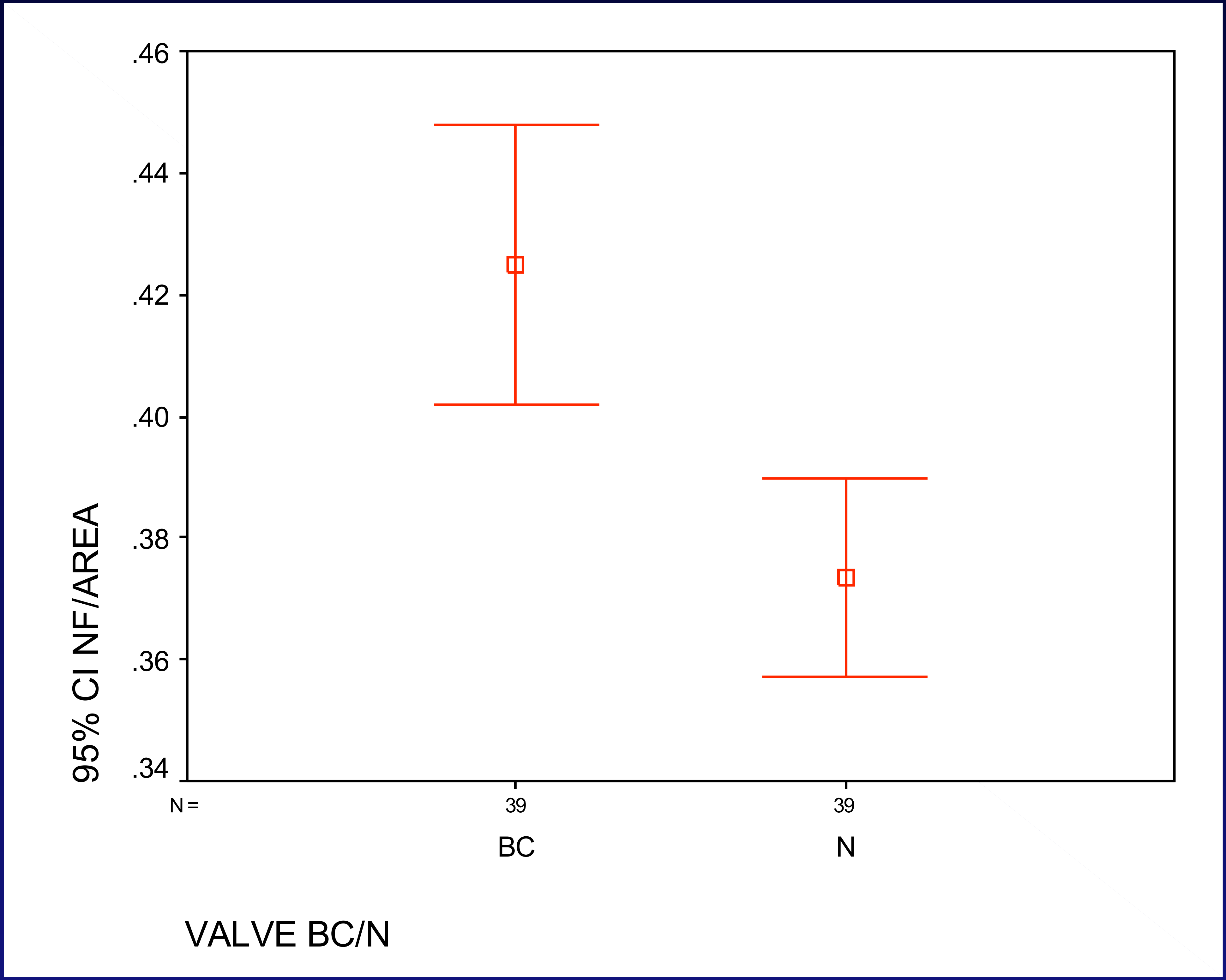
Hahn et al, JACC, 1992

Pachulski et al, Br Heart J, 1993

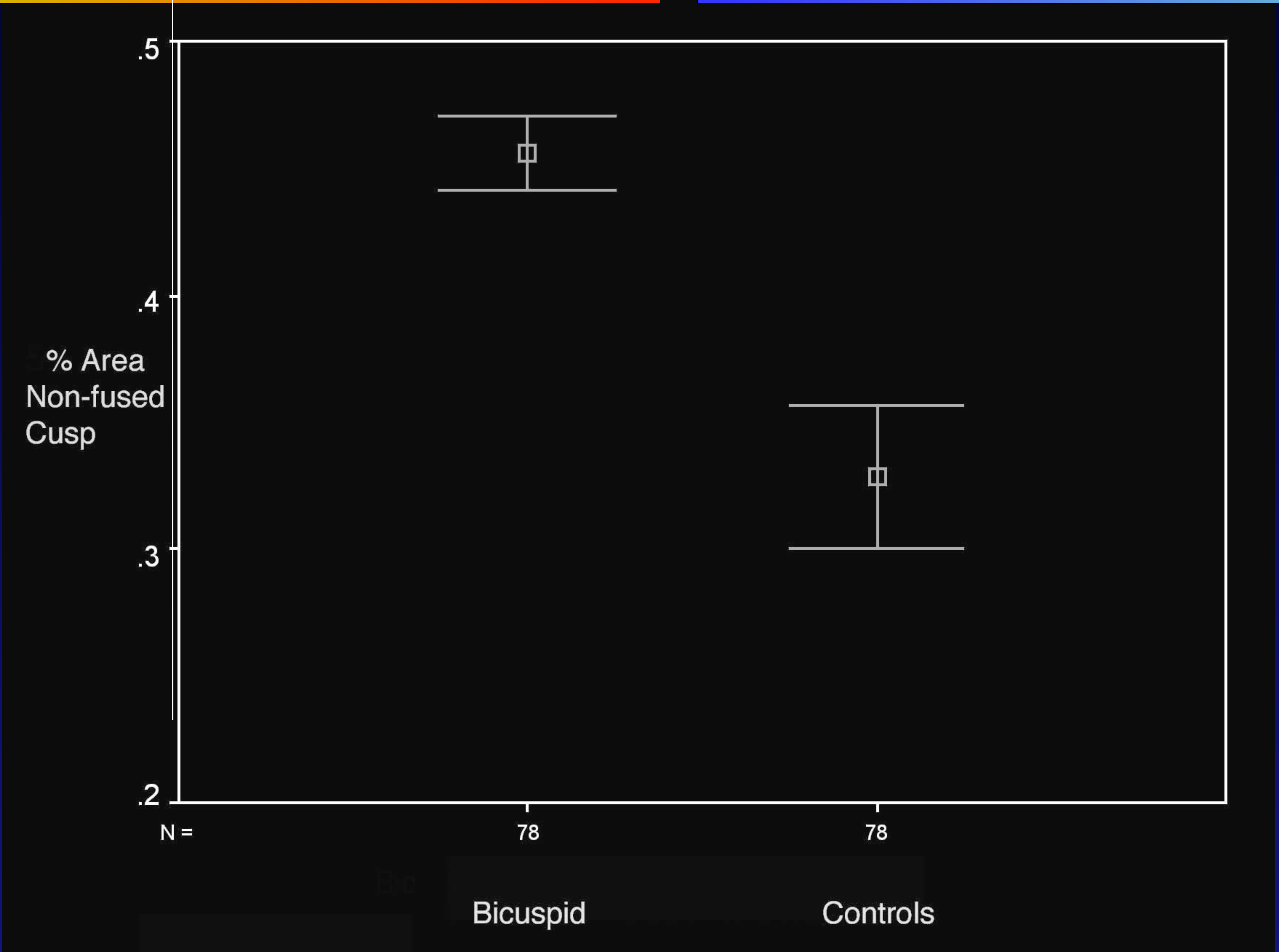
Results

- We found that there was a difference between the type of aortic valve fusion with or without coarctation.
- The patients with aortic coarctation had a higher frequency of fusion between the right and left coronary cusps - 87% versus 44% when no coarctation was found ($p < 0.001$).
- In contrast, patients with bicuspid valves but without coarctation had a higher incidence of fusion between the right and non-coronary cusps (39/78=50% versus 4/39 =10%) ($p < 0.001$).

Non-fused Cusp with CoA vs. Corresponding Cusp in Normals.

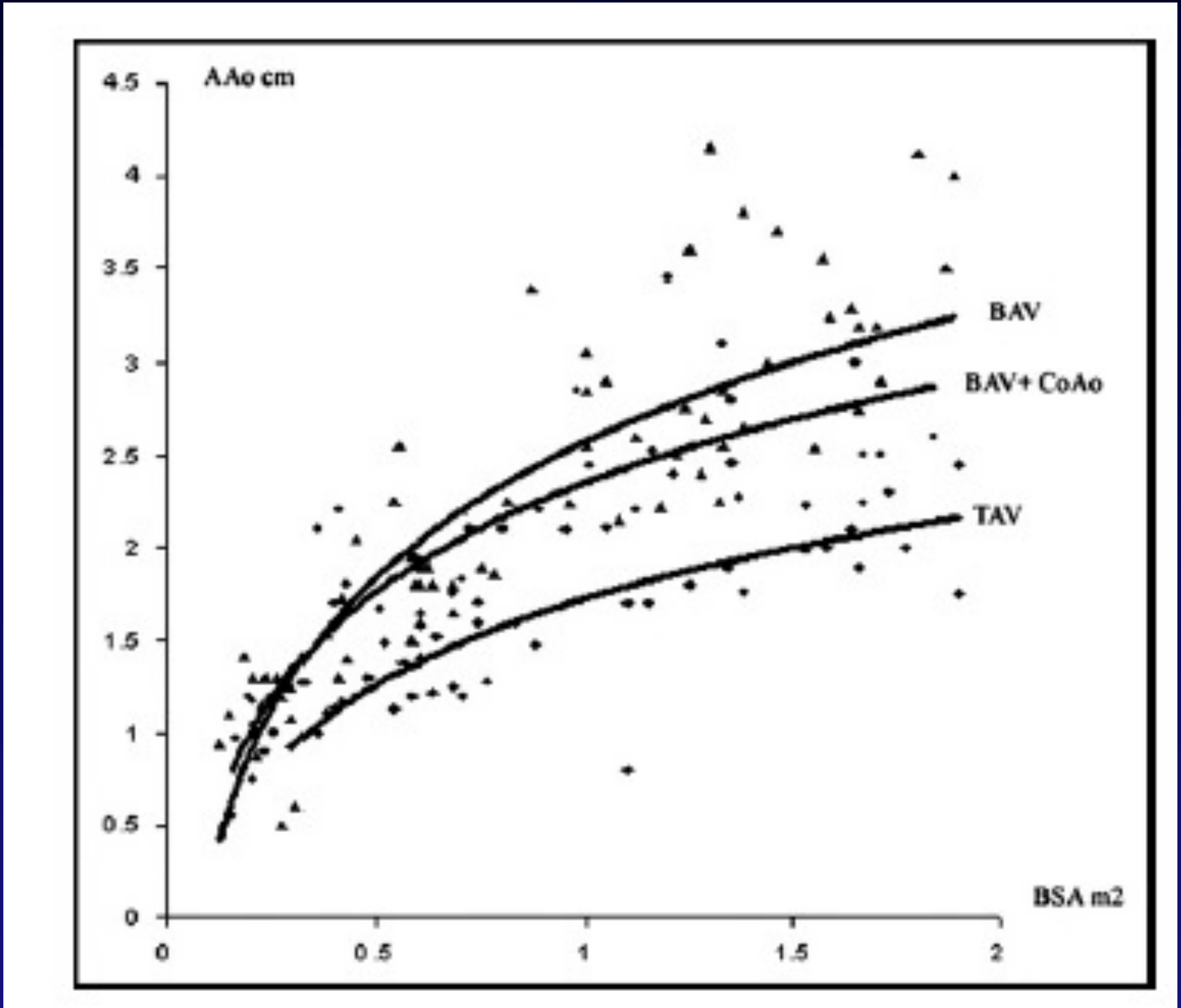


Non-fused Cusp In Bicuspid Valves vs. Corresponding Cusp in Normals.

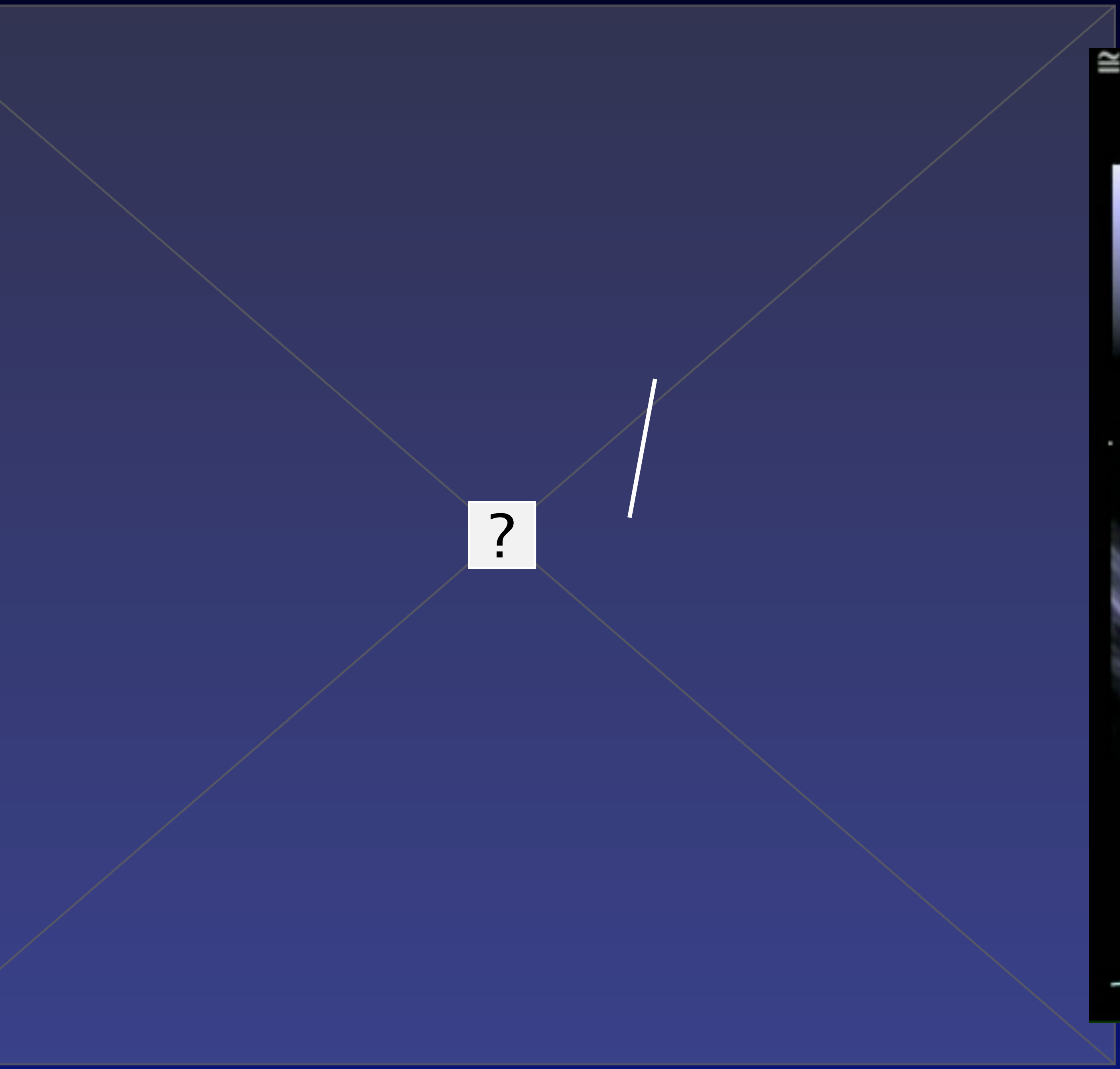


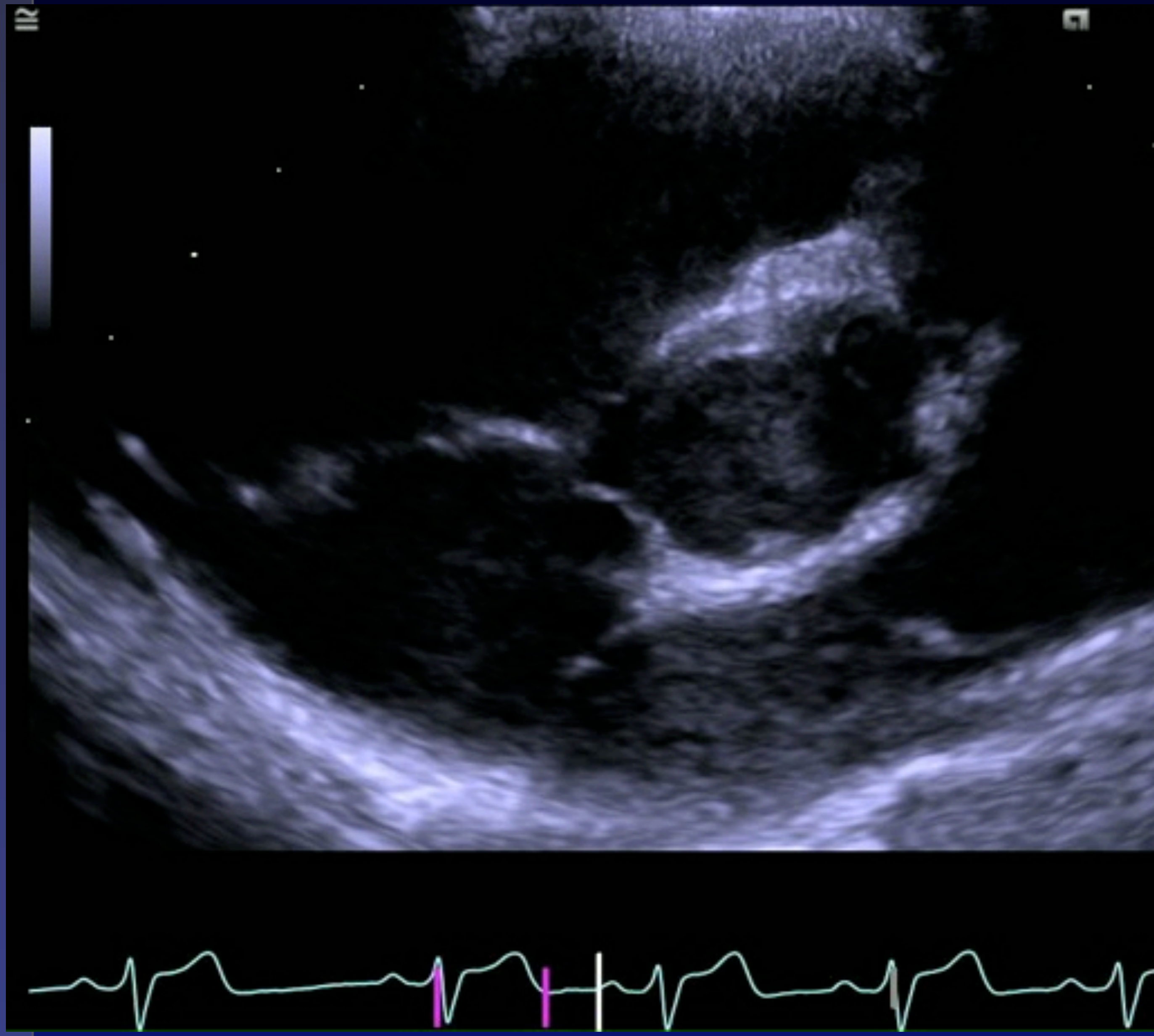
NHS.

Ascending Aortic Diameter Nl, BAV/ COA , BAV

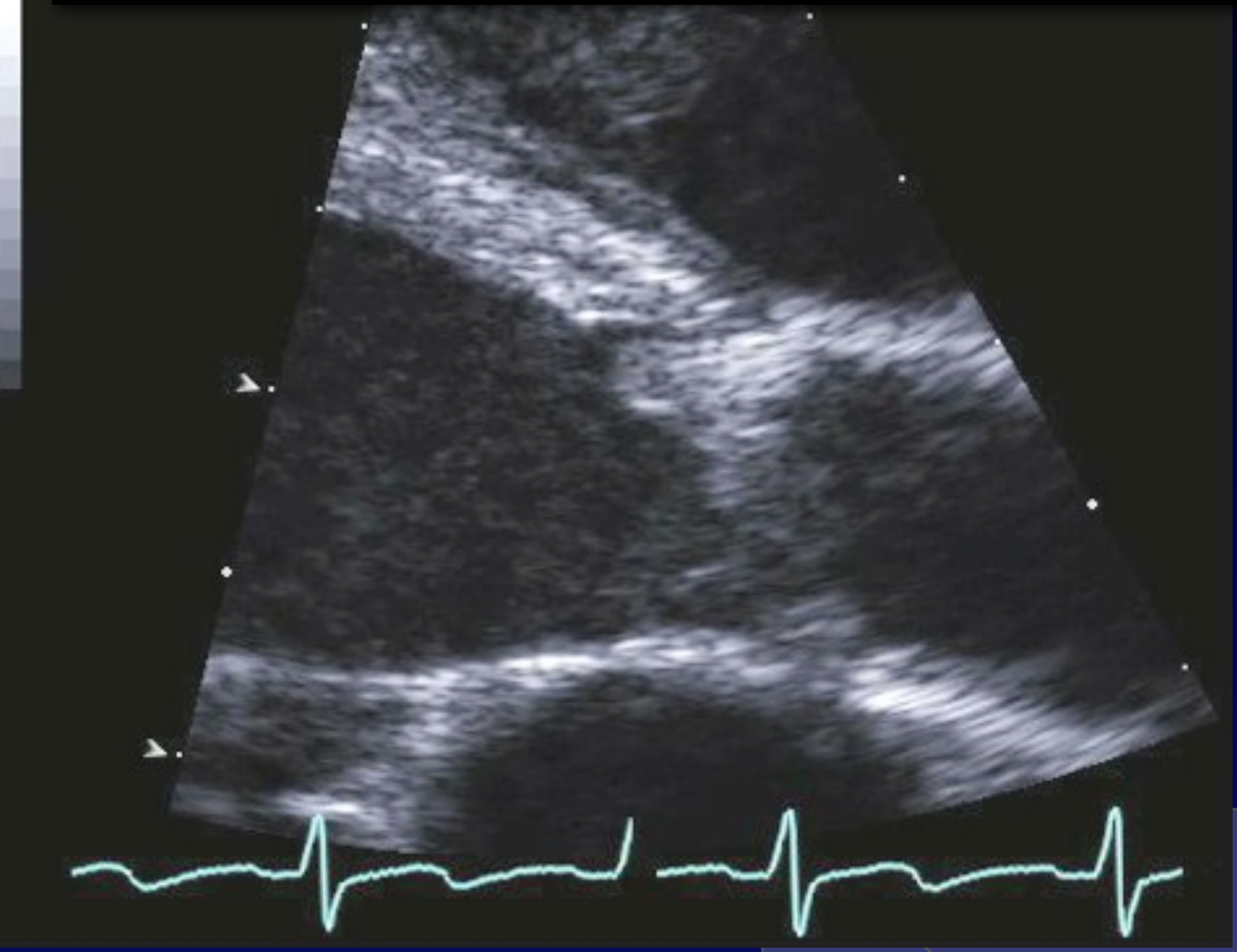


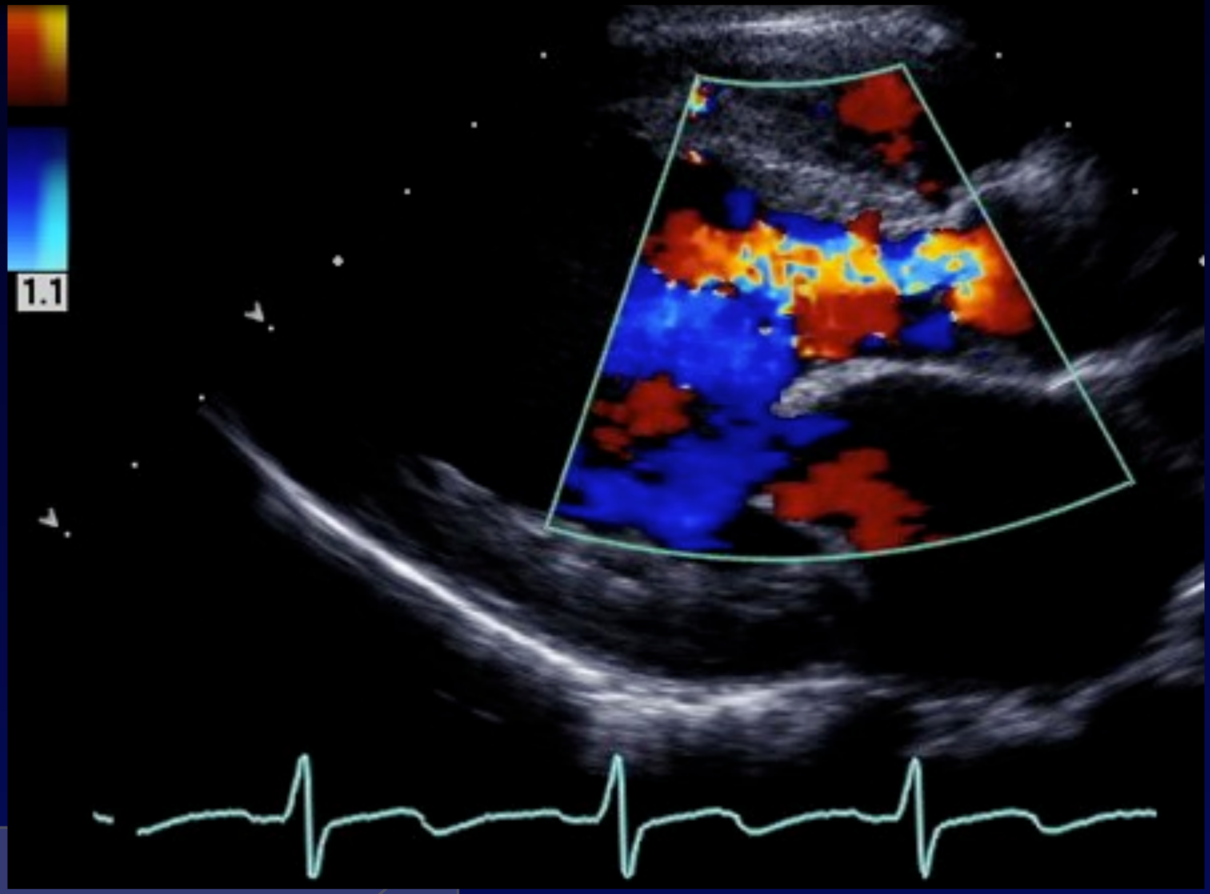
Cusp herniation with bicuspid Aortic valve





Bicuspid aortic valve with leaflet herniation + AI





Frequency of specific congenital cardiac lesions in patients with bicuspid aortic valves

Congenital Heart Defect Associated With BAVs	Cases
CoA	40 (34.2%)
Ventricular septal defect	16 (13.7%)
Subaortic stenosis	18 (15.4%)
Mitral abnormalities (Shone's included)	13 (11%)
Patent ductus arteriosus/interatrial septal defect	10 (8.5%)
Pulmonary atresia	3 (2.6%)
Hypoplastic left-sided cardiac syndrome	3 (2.6%)
Ebstein's anomaly	3 (2.6%)
Endocardial fibroelastosis	2 (1.7%)
Pulmonary stenosis	2 (1.7%)
Atrioventricular septal defect	2 (1.7%)
Partial anomalous pulmonary venous return	1 (0.8%)
Interrupted aortic arch	1 (0.8%)

Syndromes 4.2%: 3 Turner's, 1 Williams', 1 Down's Syndrome

Bicuspid Valve & Aortic Dissection

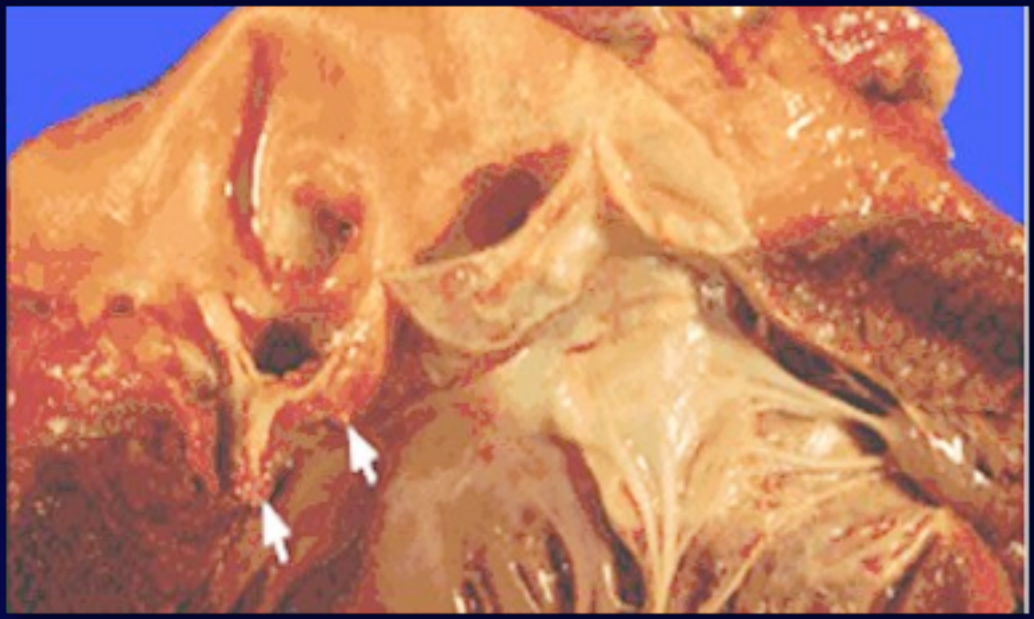
- The incidence of aortic dissection in patients with a bicuspid valve is 6%
- The incidence of aortic dissection in patients with a tricuspid valve is 0.7% Larson & Edwards, Am J Cardiol, 1984
- Aortic dissection is nearly 10 times more frequent in patients with a bicuspid valve

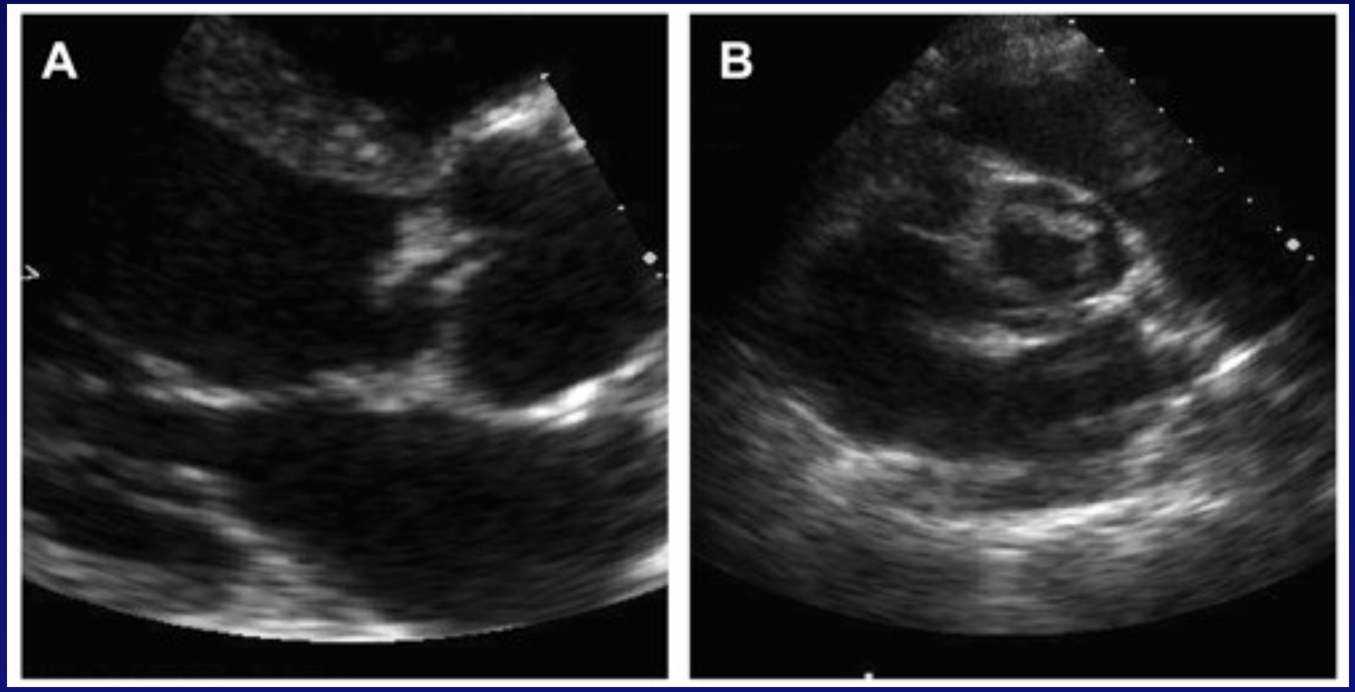
Aortic Dissection

- The incidence of a bicuspid valve is 28% in patients with aortic dissection younger than 40 years of age!

Gore, Arch Pathol, 1953

Bicuspid Aortic Valve: Infective Endocarditis





Infective endocarditis is responsible for ~50% of severe AR in BAV

1886: Osler: 1st associated BAV with infective endocarditis

1923: Lewis and Grant: >1/3 of pathologic specimens of aortic valve endocarditis were with BAV

Recent surgical series: 25-54% of infected aortic valves requiring AVR were BAV

Bicuspid Valve & Aortic Root Abscess

Pressure Recovery in Pediatric Aortic Valve Stenosis

R.E. Villavicencio,[1] T.J. Forbes,[1] R.L. Thomas,[2] R.A. Humes[1]

[1]Department of Pediatrics, Division of Cardiology, Wayne State University School of Medicine/Detroit Medical Center and Children's Hospital of Michigan, 3901 Beaubien Avenue, Detroit, MI 48201, USA

[2]Department of Pediatrics, Children's Research Center of Michigan, Detroit, MI 48201, USA

Prediction of Pressure Recovery

The degree of pressure recovery can be estimated from Doppler echocardiography using a previously validated equation based on principles of fluid dynamics [6, 7]. The measured peak continuous-wave Doppler gradients were corrected for pressure recovery with the following equation:

$$p_3 - p_2 = 4V_{cw}^2 \times (2\mathrm{AVA_c}/\mathrm{AOA}) \times [1 - \mathrm{AVA_c}/\mathrm{AoA}]$$

Vcw is the peak Doppler velocity

AoA is the
cross-sectional area of the ascending aorta

AVAc is the estimated
aortic valve orifice by the continuity equation

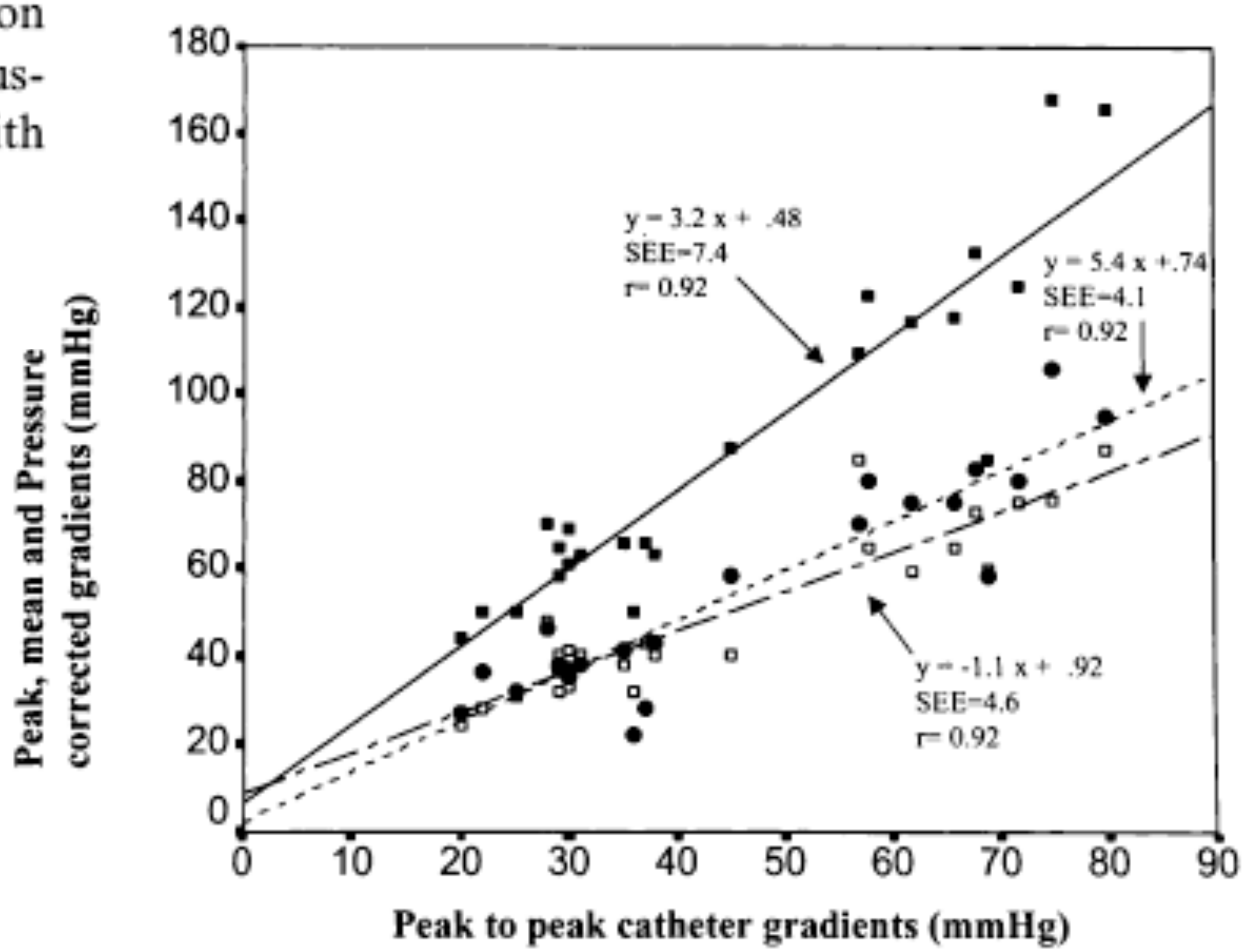
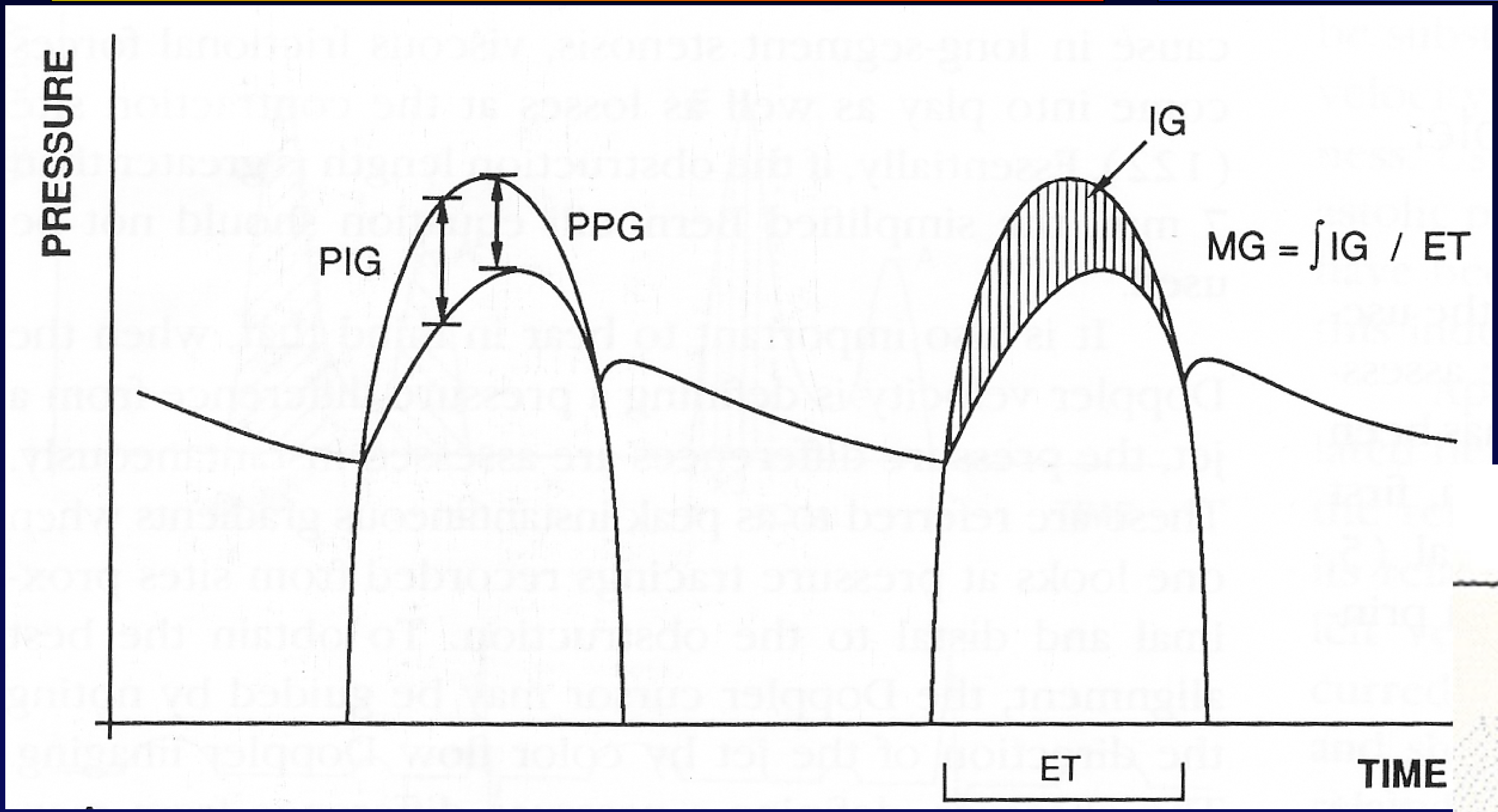
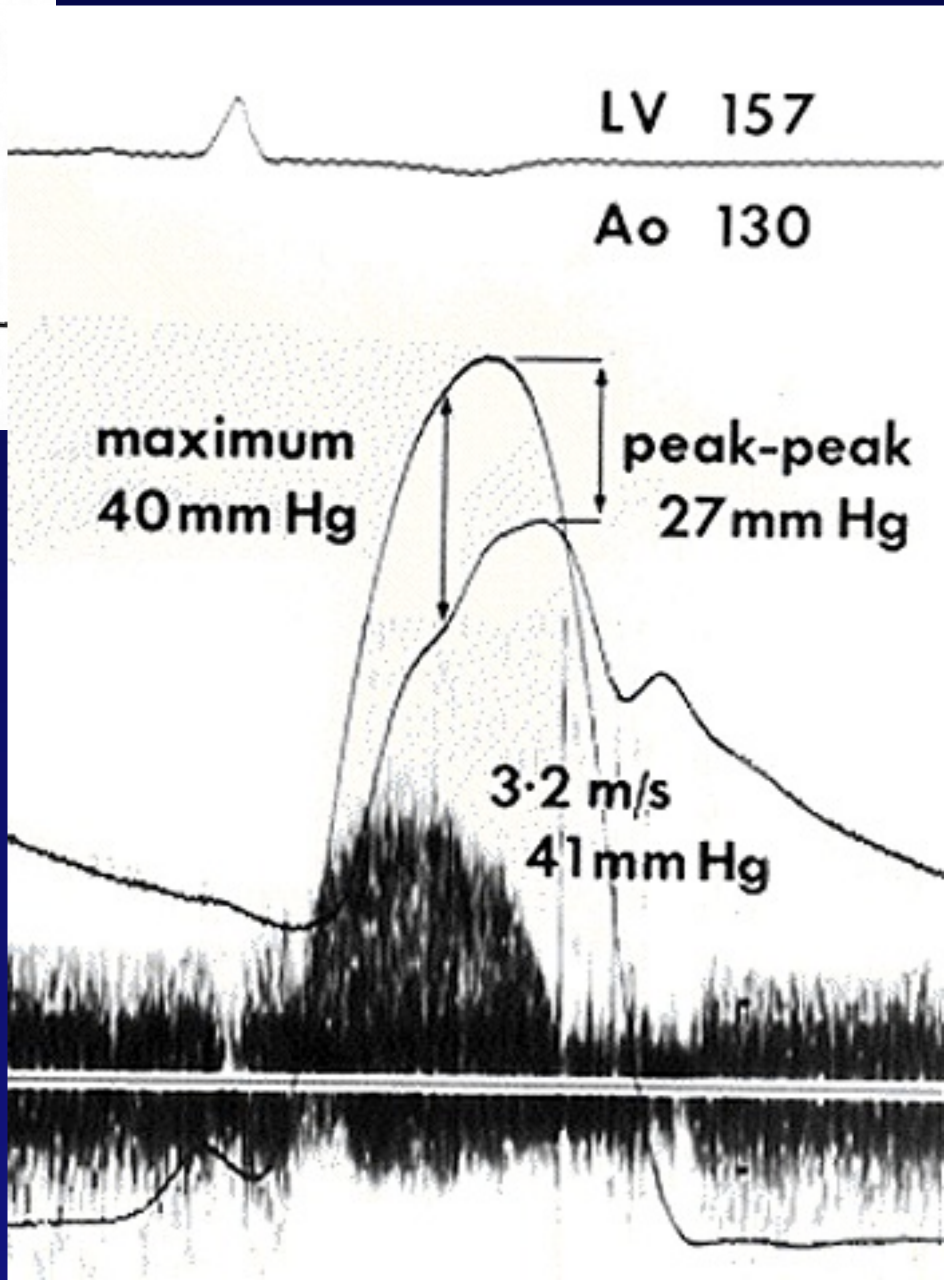


Fig. 2. Peak Doppler gradients (■), mean Doppler gradients (●), and pressure recovery corrected gradients (□) versus peak-to-peak catheter gradients. Similar linear relationship to the catheter gradients is appreciated for all three Doppler measurements.

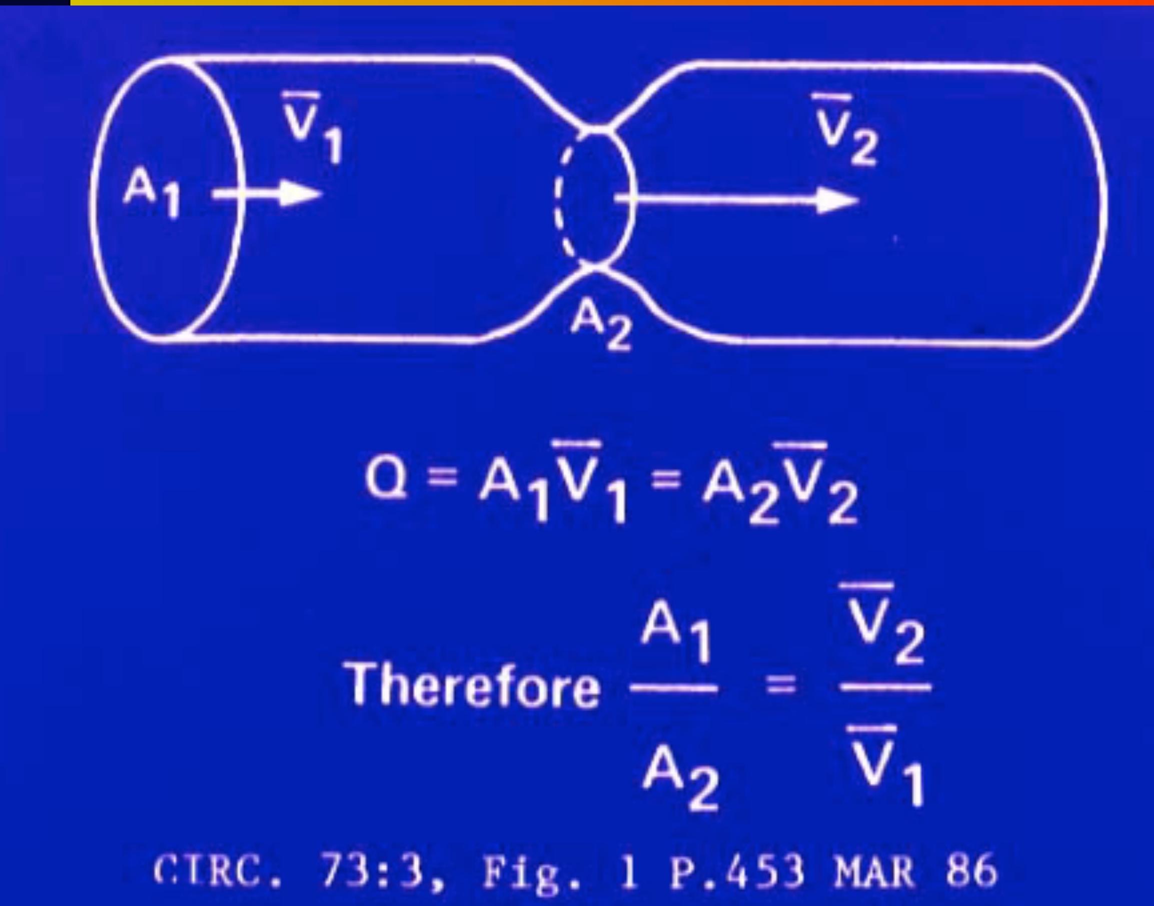
Pressure Drop Echo vs Catheter Estimates



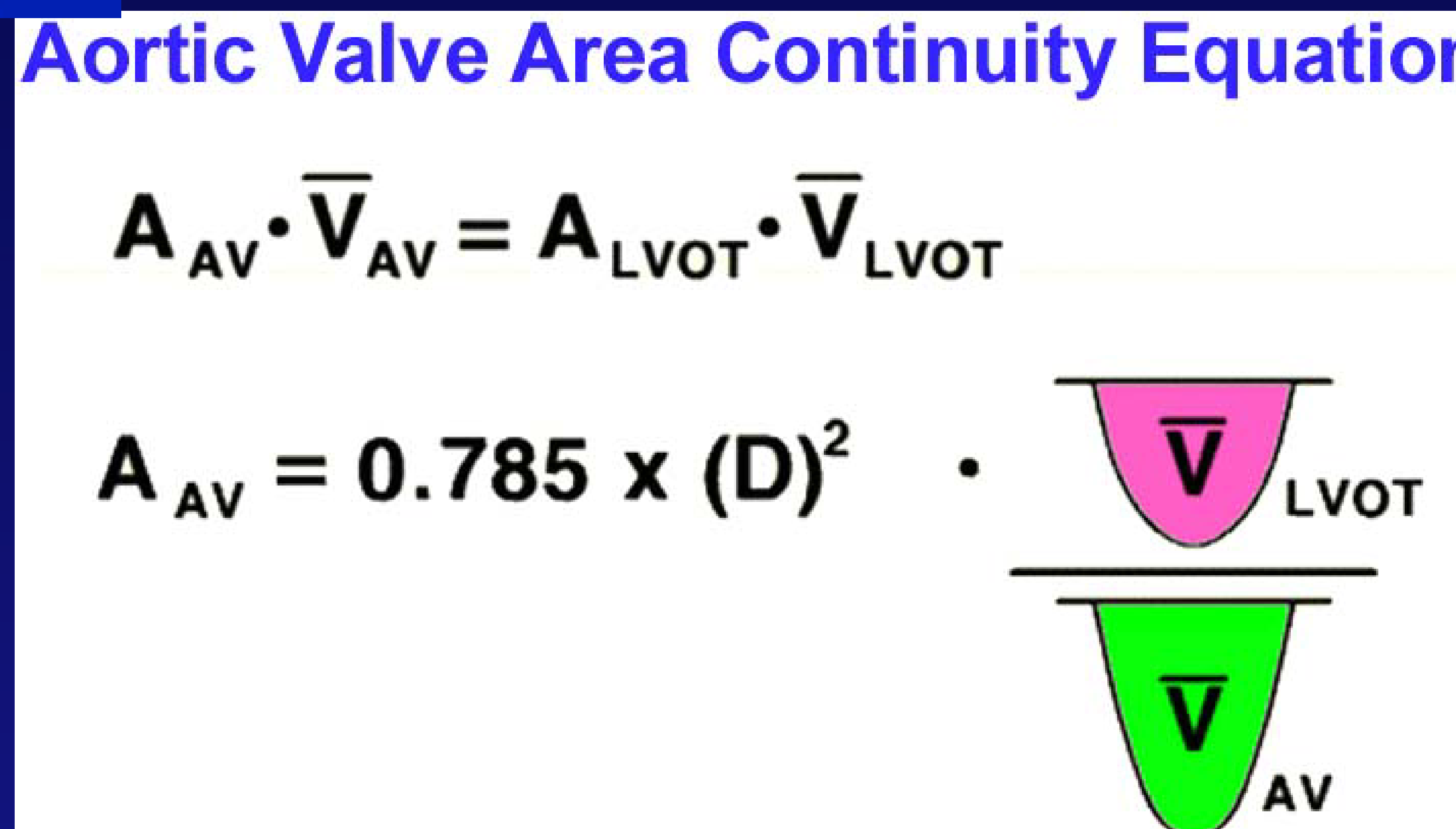


NHS.

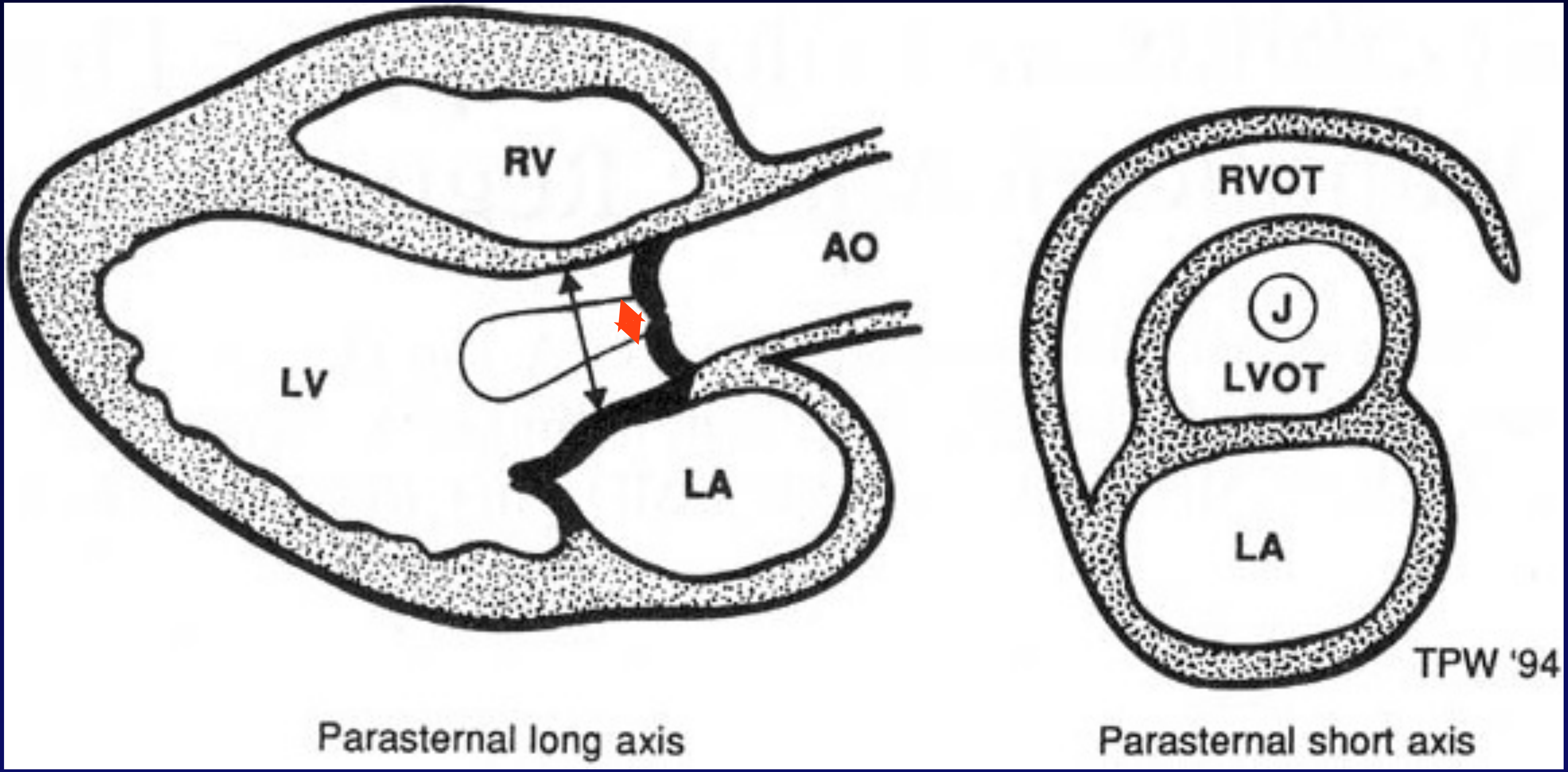
Continuity Equation



Critical for Volume Flow in
Flow Convergence Calculations



LVOT/jet Width Measurements

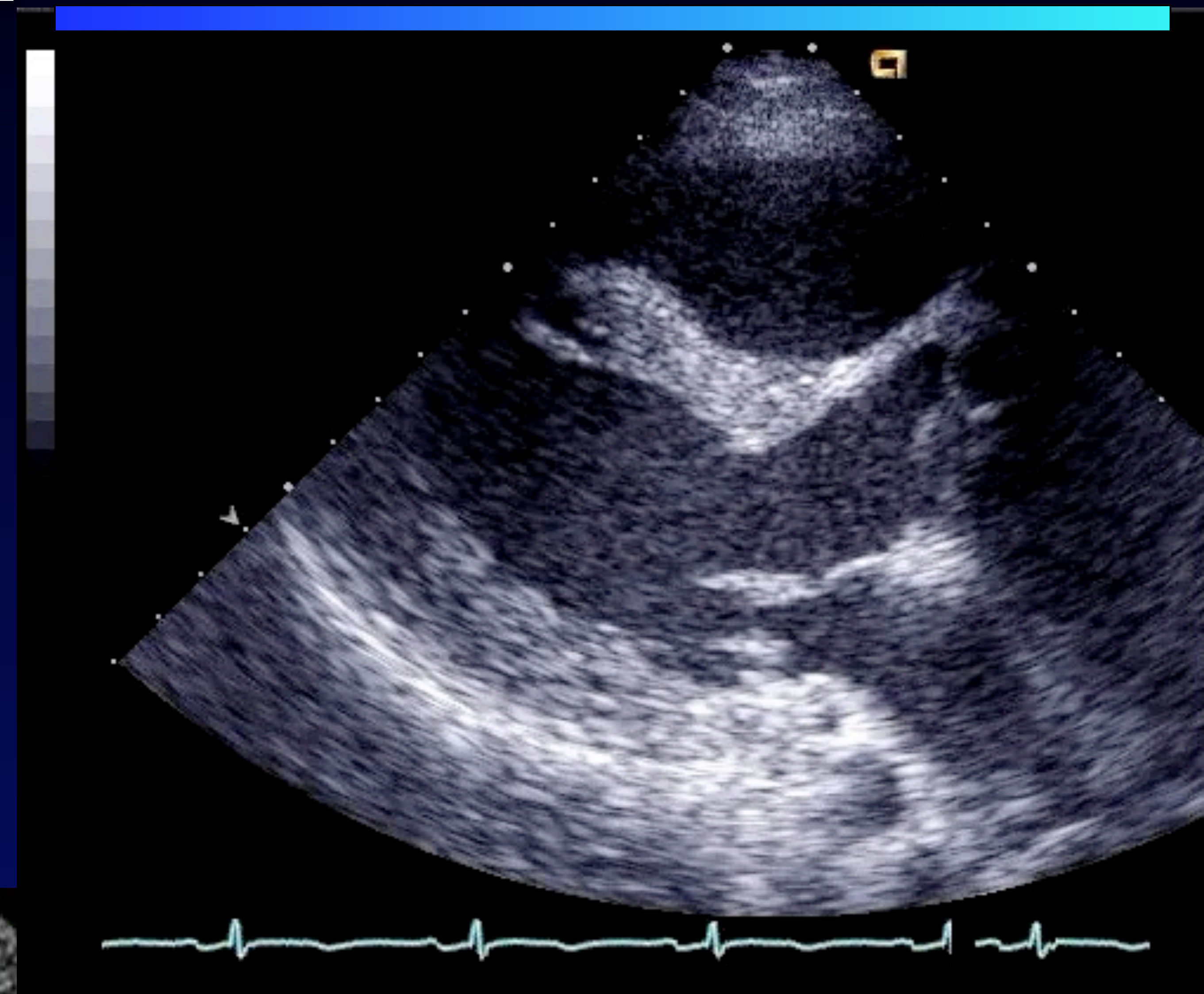


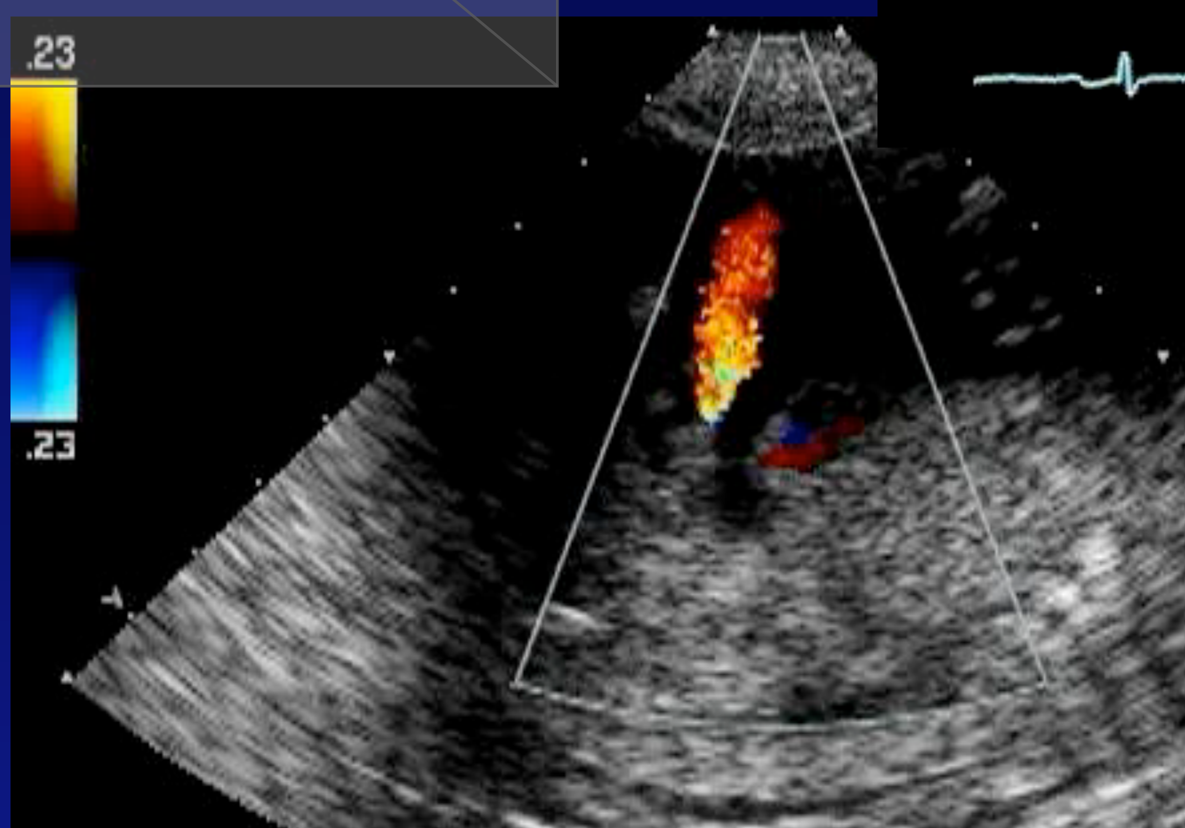
Doppler Evaluation of Aortic Regurgitation in Children

Tani LY, Minich LL, Day RW, Orsmond GS, Shaddy RE.
Am J Cardiol. 1997 Oct 1;80(7):927-31.

NHS.

Things aint always what they seem,
Skimmed milk masquerades as cream.





Echo Techniques for AI: Correlation with Angio

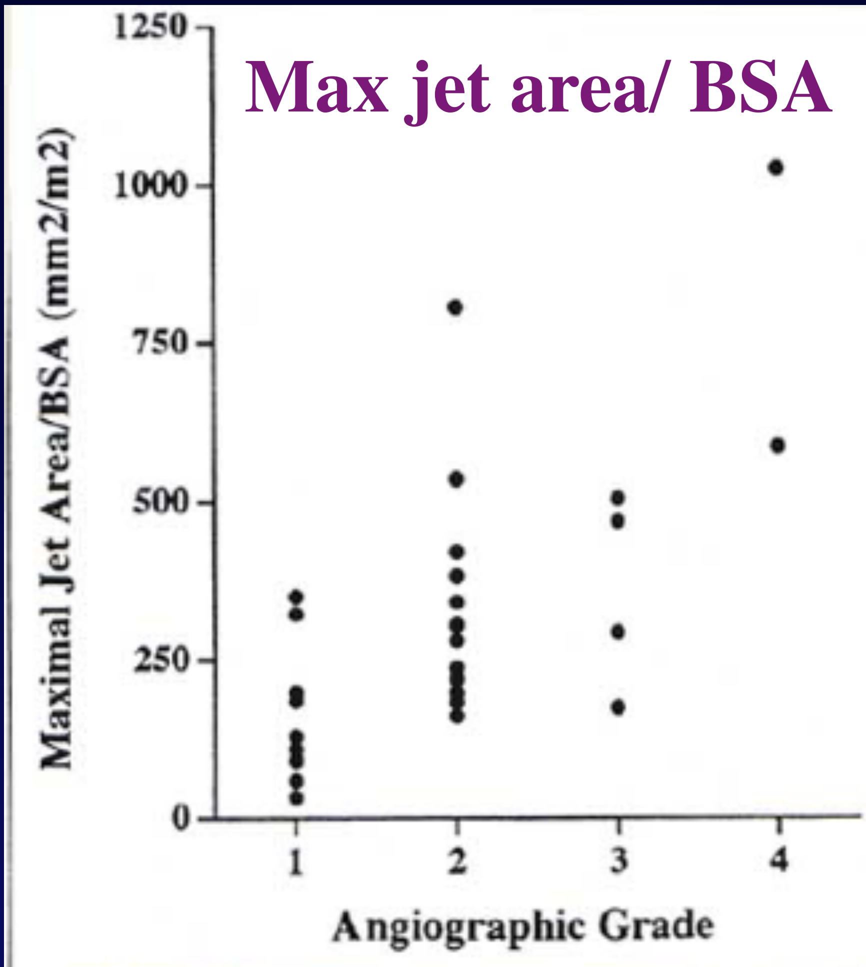


FIGURE 1. Maximum jet area/body surface area (BSA) compared with angiographic grade of aortic regurgitation. Means for 3+ and 4+ aortic regurgitation were significantly different, $p < 0.05$.

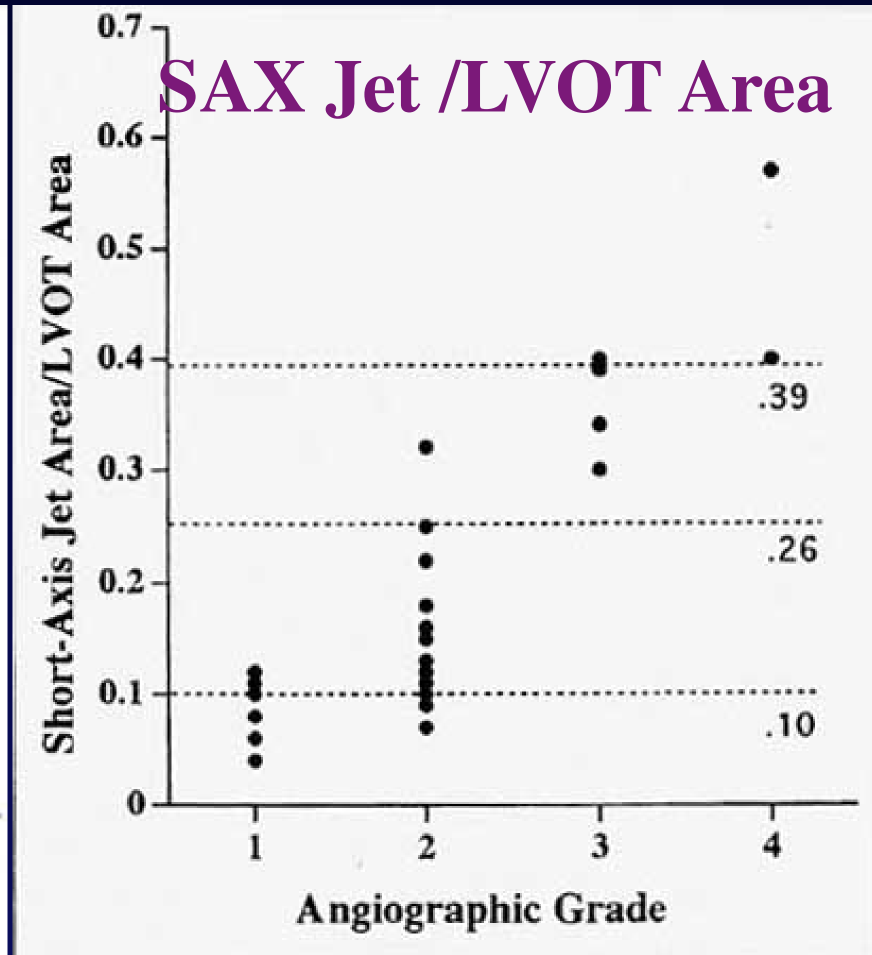


FIGURE 2. Short-axis jet area/left ventricular outflow tract (LVOT) area compared with angiographic grade of aortic regurgitation. Means were different between 2+ and 3+ and between 1+ and 2+ aortic regurgitation, $p < 0.05$.

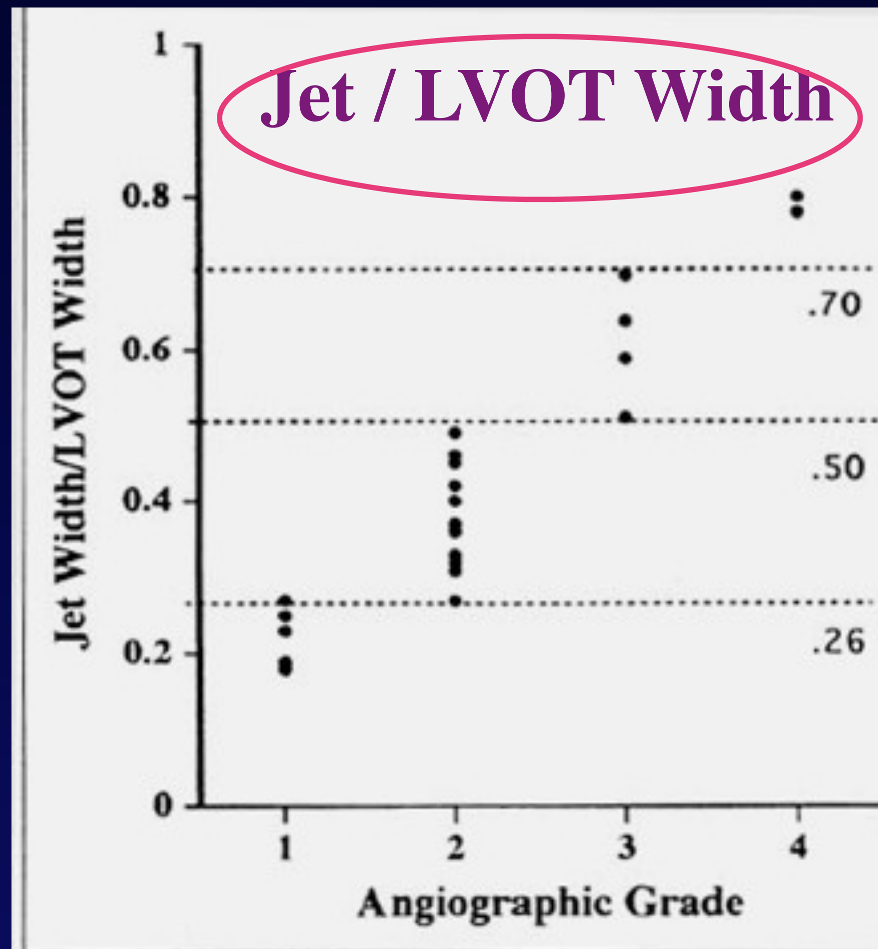


FIGURE 3. Jet width/left ventricular outflow tract (LVOT) width compared with angiographic grade of aortic regurgitation. Means were different between 3+ and 4+, between 2+ and 3+, and between 1+ and 2+ aortic regurgitation, $p < 0.05$.

TABLE III Doppler Indexes Versus Angiographic Grade of Aortic Regurgitation

Angio	No.	Jw/LVOTw	Jw/BSA$^{1/2}$	SaxJA/LVOT-A	SaxJA/BSA	JL/BSA$^{1/2}$	MaxJA/BSA	Rev/fwd VTI	Slope
1+	10	0.22 ± 0.03	4.0 ± 0.6	0.08 ± 0.03	38.7 ± 18.0	30.1 ± 15.6	156 ± 108	0.12 ± 0.08	3.14 ± 0.87
2+	14	0.37 ± 0.07†	6.5 ± 1.7†	0.15 ± 0.07*	73.1 ± 36.1	40.4 ± 8.7	328 ± 182	0.23 ± 0.29	4.31 ± 1.5
3+	4	0.61 ± 0.08†	10.8 ± 1.3†	0.36 ± 0.05†	142 ± 31.0†	48.1 ± 7.5	360 ± 154	0.69 ± 0.20*	4.31 ± 1.5
4+	2	0.79 ± 0.01*	17.5 ± 3.8†	0.49 ± 0.12	317 ± 4.2†	49.8 ± 3.5	804 ± 312*	0.73 ± 0.16	5.16 ± 2.2

*$p < 0.05$; †$p < 0.005$ compared with next lower grade.

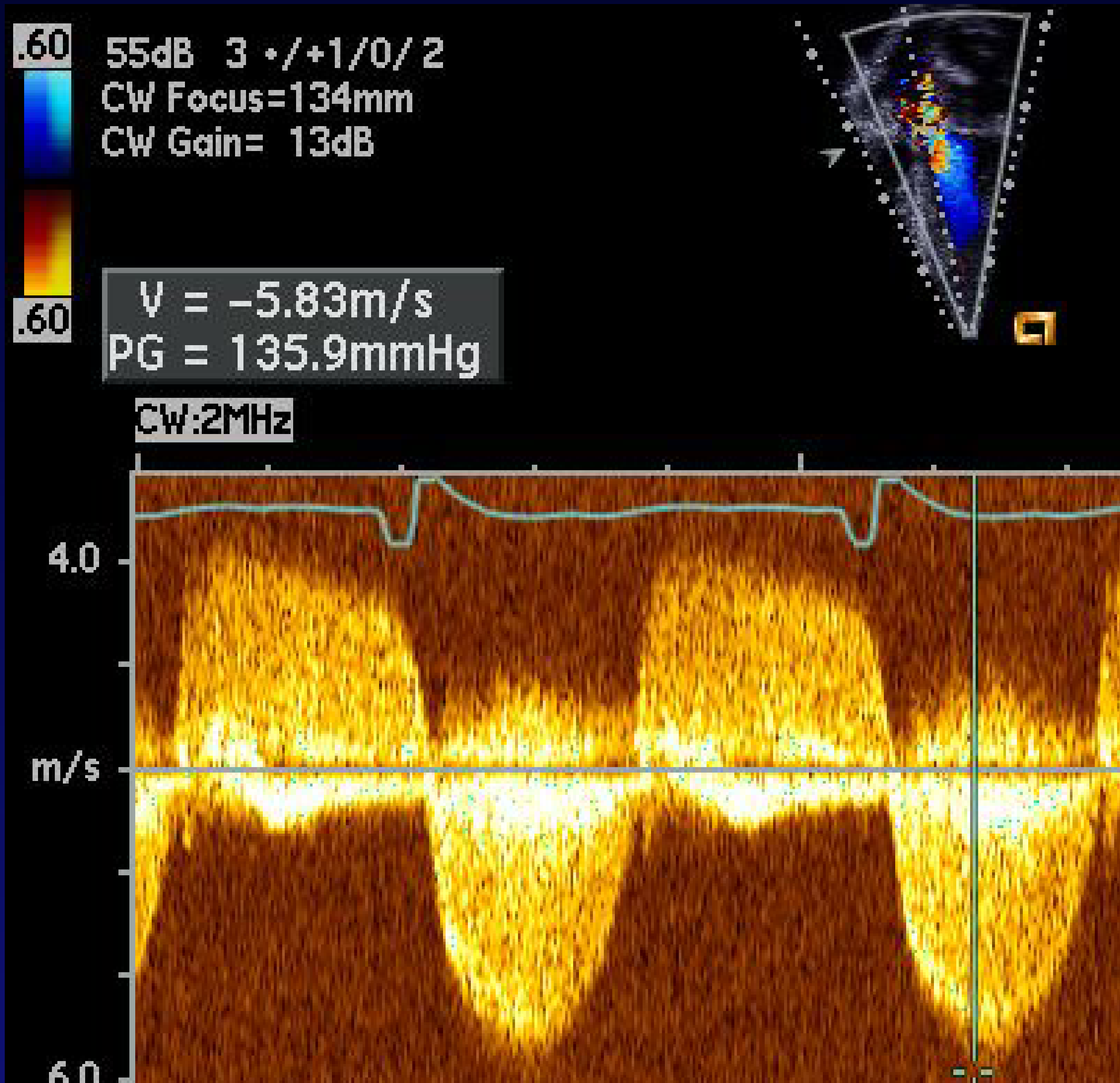
Data are expressed as mean ± SD.

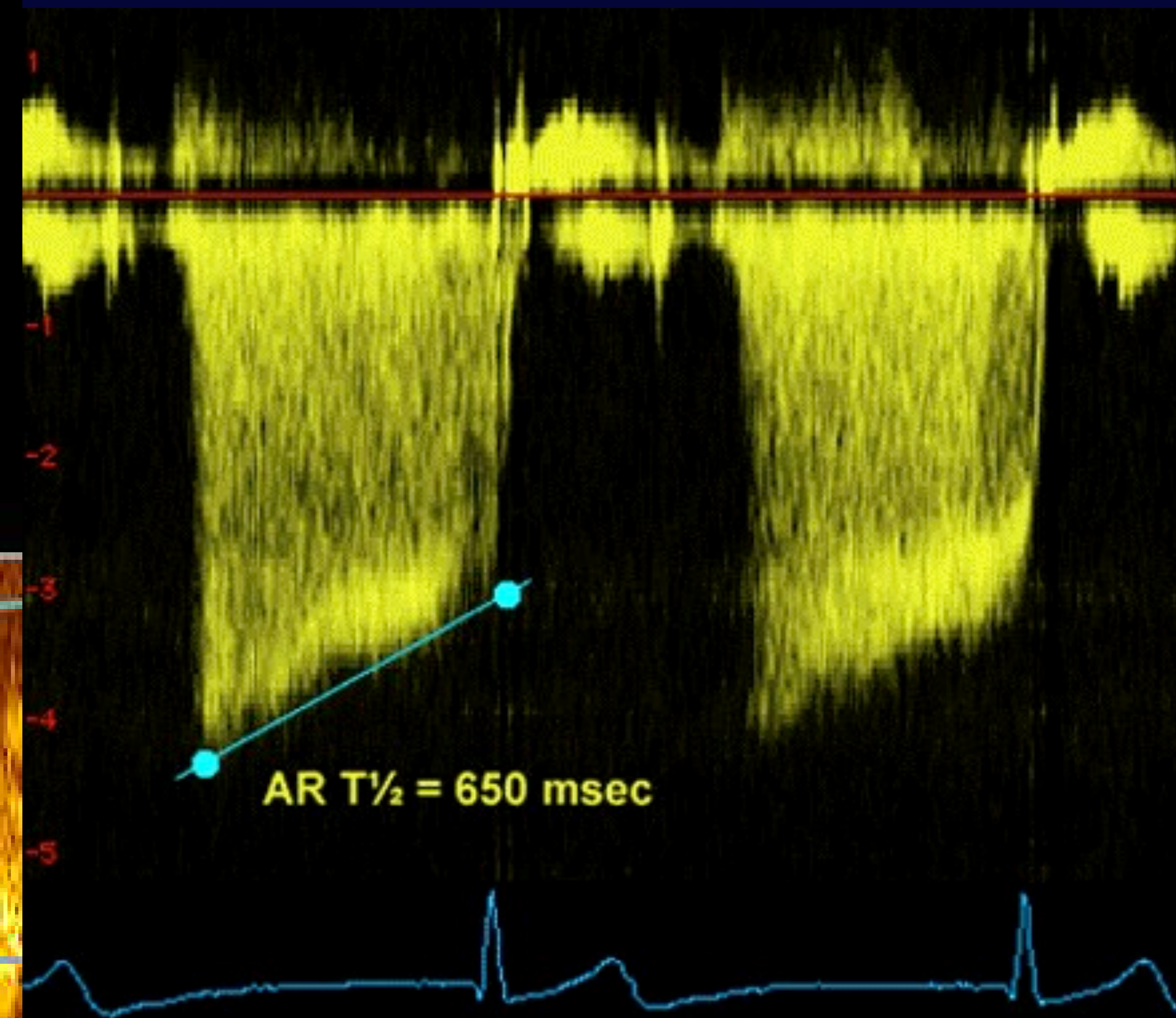
Angio = angiographic grade of aortic regurgitation; BSA = body surface area; JL = jet length; Jw = jet width; LVOTw = left ventricular outflow tract width; LVOT-A = left ventricular outflow tract area; MaxJA = maximal jet area; Rev/fwd VTI = ratio of reverse and forward velocity time integrals from the abdominal aorta; SaxJA = short-axis jet area.

Jet measurements = millimeters or square millimeters; body surface area = square meters; slope = meters per square seconds.

Tani, Minnich et al

Aortic Regurgitation: Pressure 1/2 time

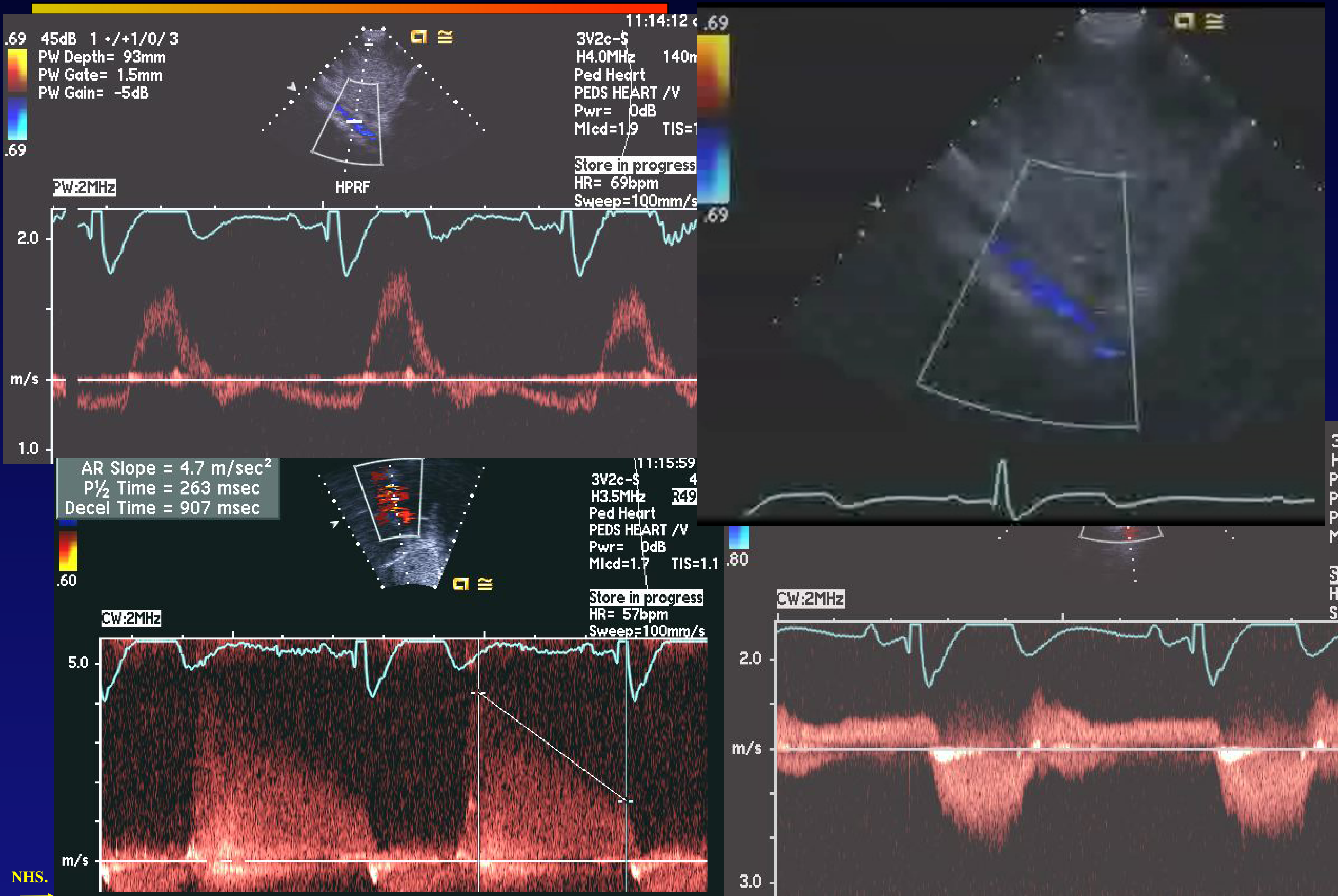




NHS.

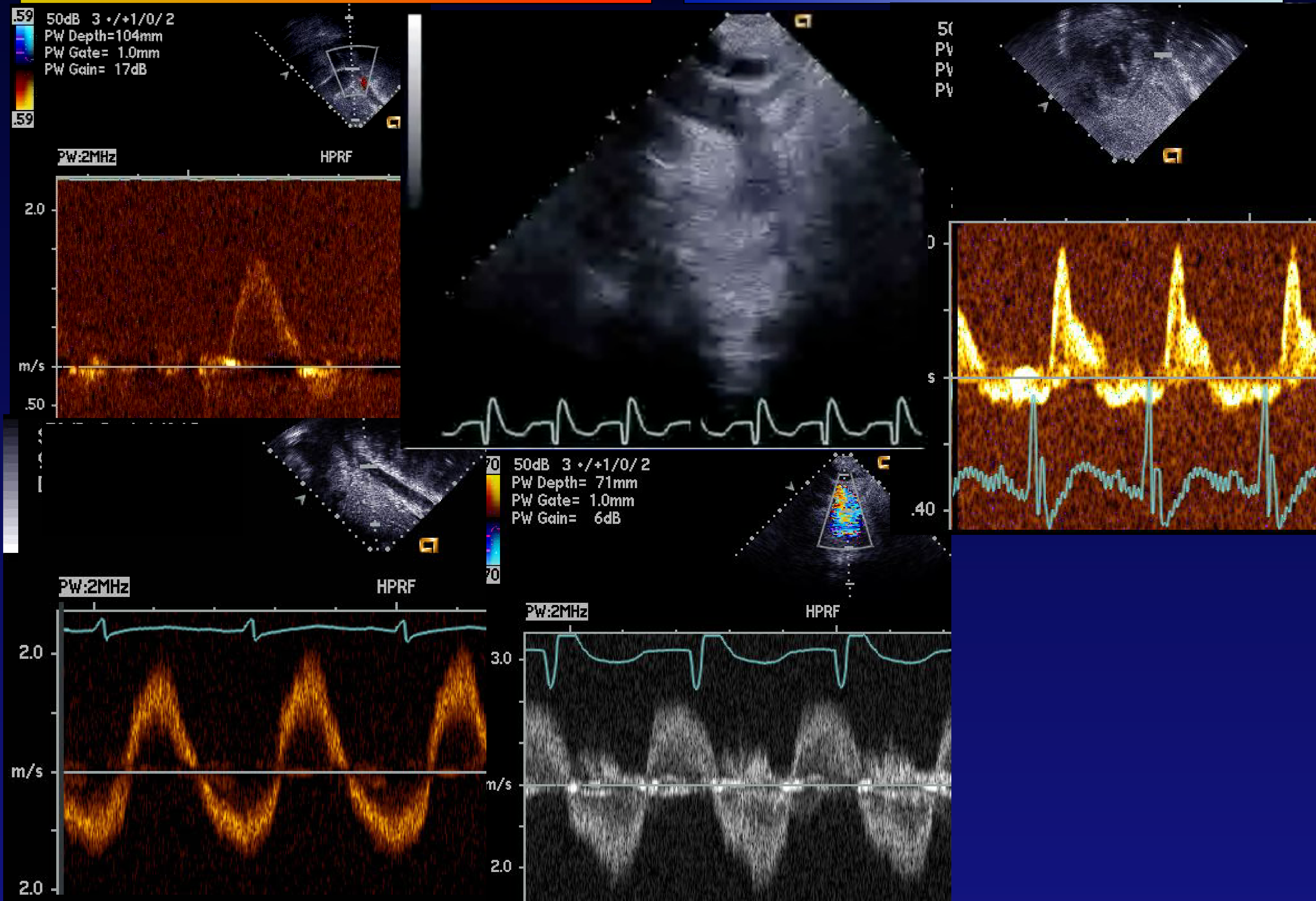
Aortic Regurgitation- How Severe?

Severe!

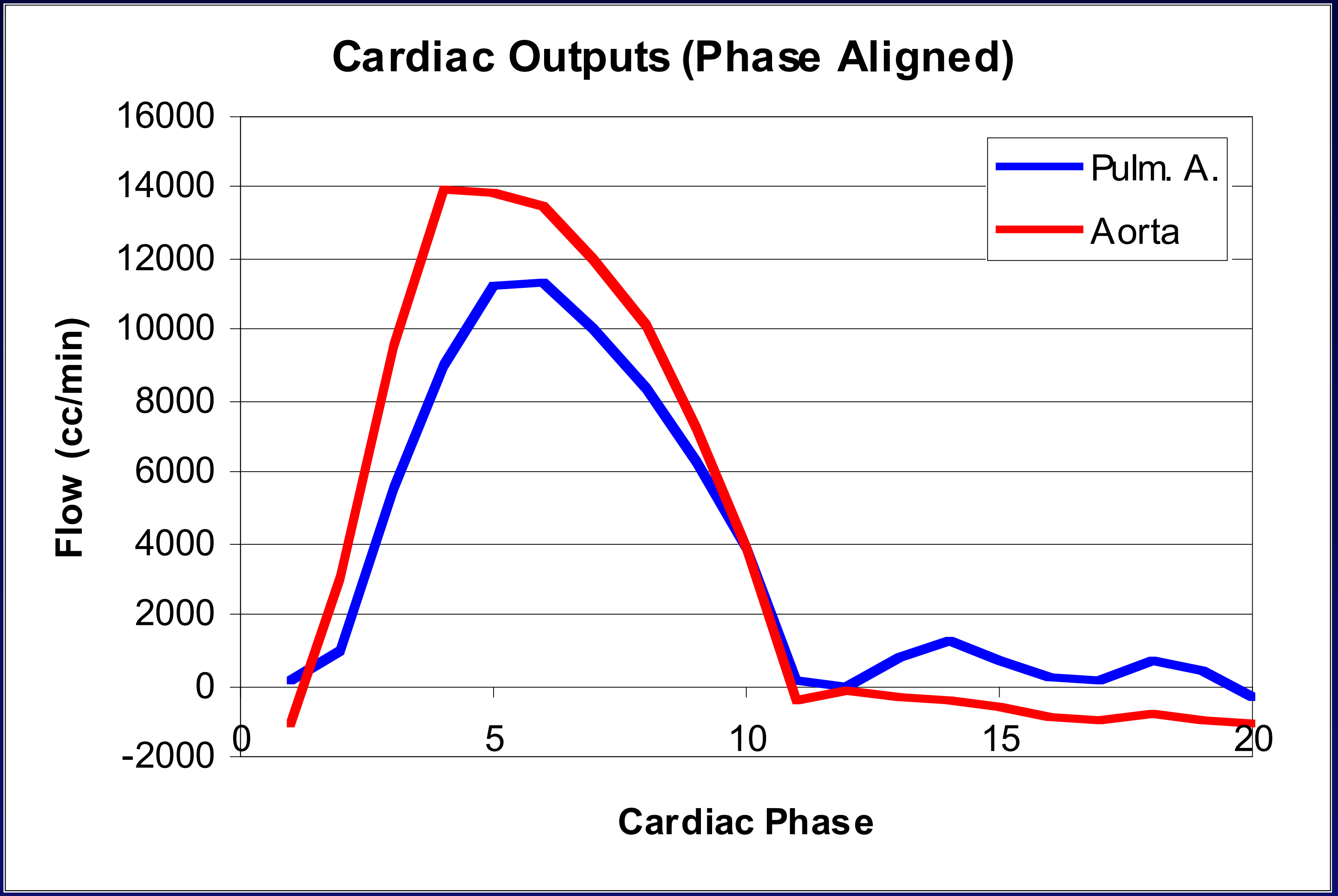


NHS.

AR: Retrograde Flow



MRI: Calculation of flows and outputs



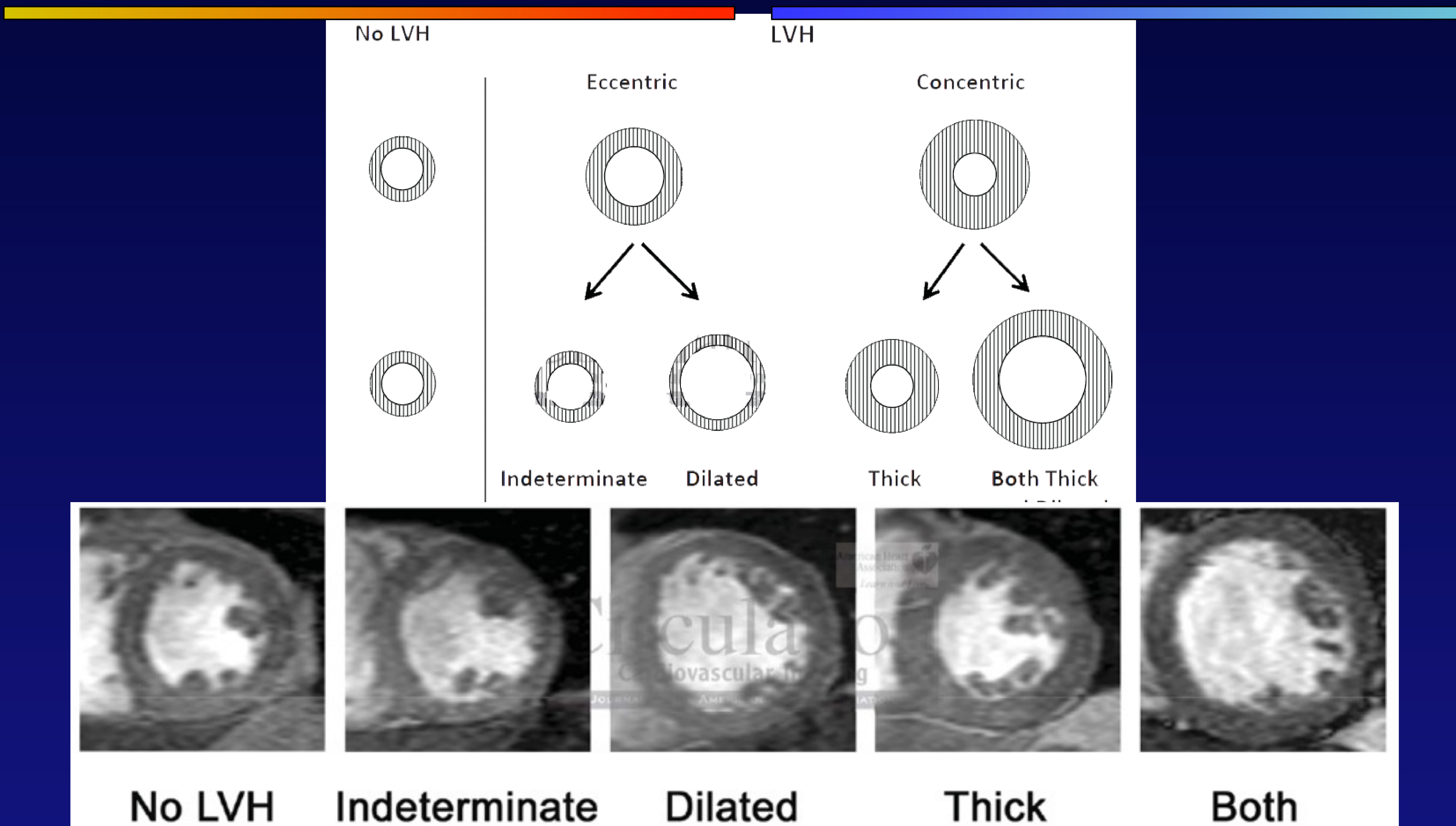
		Aortic	Pulmonary	Mitral	Tricuspid
Regurgitant Flow	L/min	0.39	0.02	-0.12	0.26
Regurgitant Volume	cc	4.26	0.22	-1.53	3.26
Regurgitant Fraction	%	8.90%	0.57%	-3.23%	6.93%

Ventricular Geometry: The Dallas Heart Study

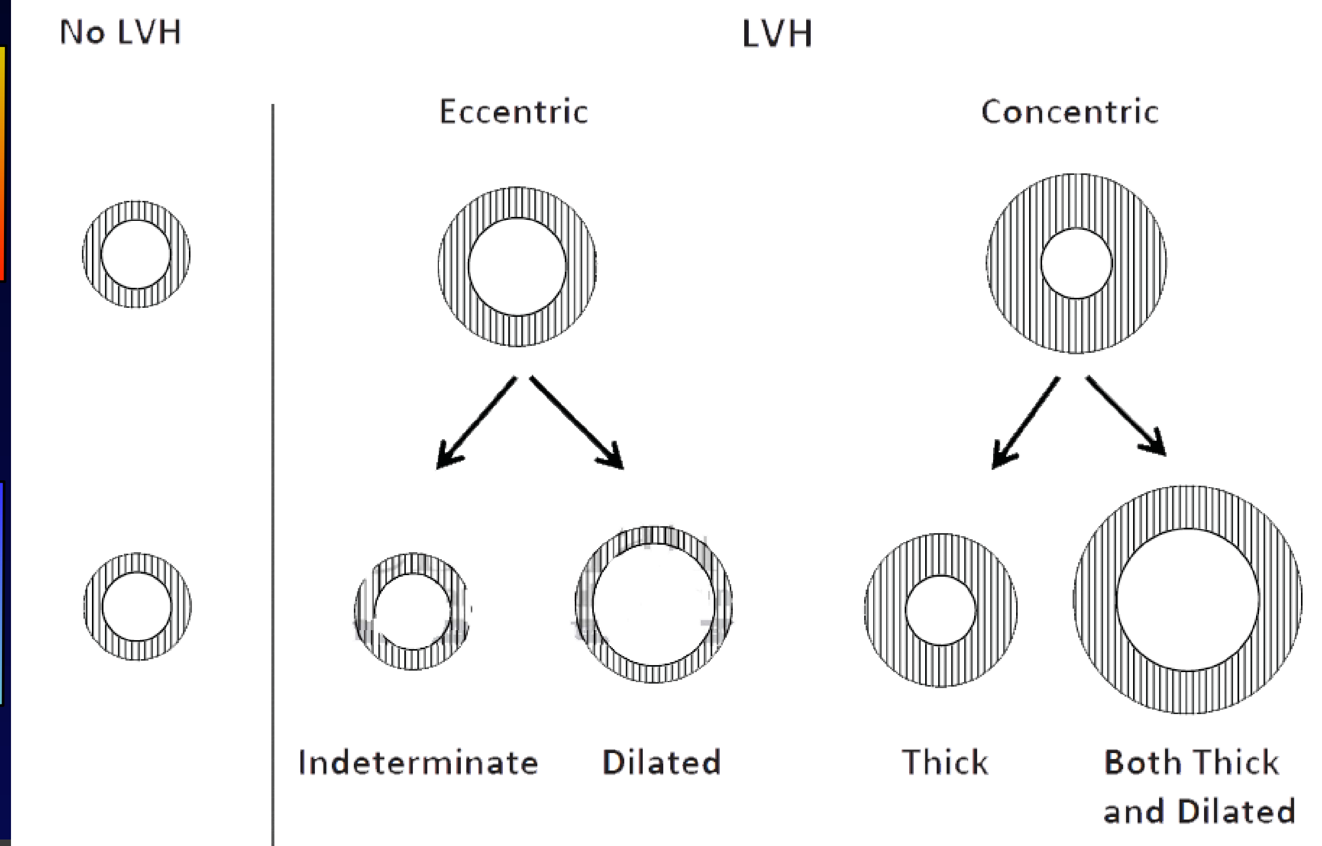
A Four-Tiered Classification of Left Ventricular Hypertrophy Based on Left Ventricular Geometry: The Dallas Heart Study

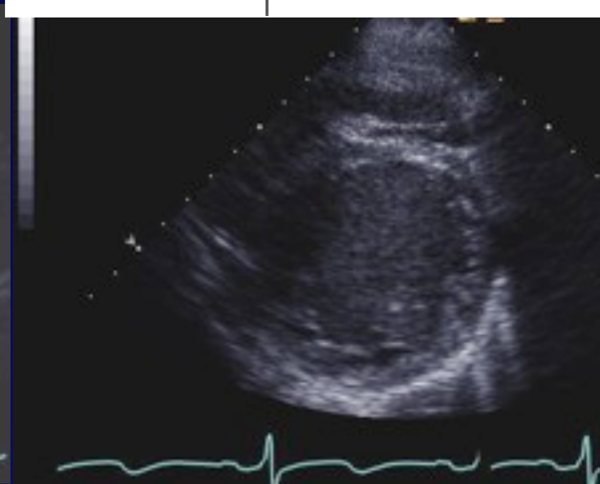
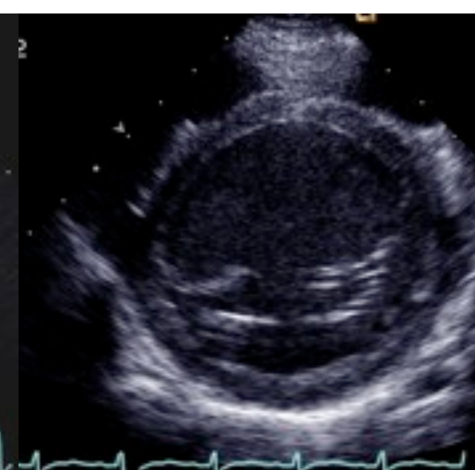
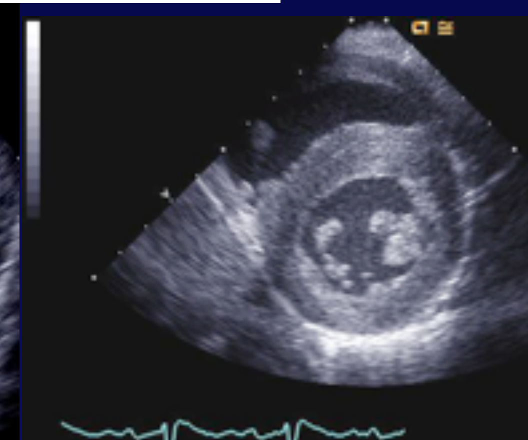
Michel G. Khouri, Ronald M. Peshock, Colby R. Ayers, James A. de Lemos and Mark H. Drazner

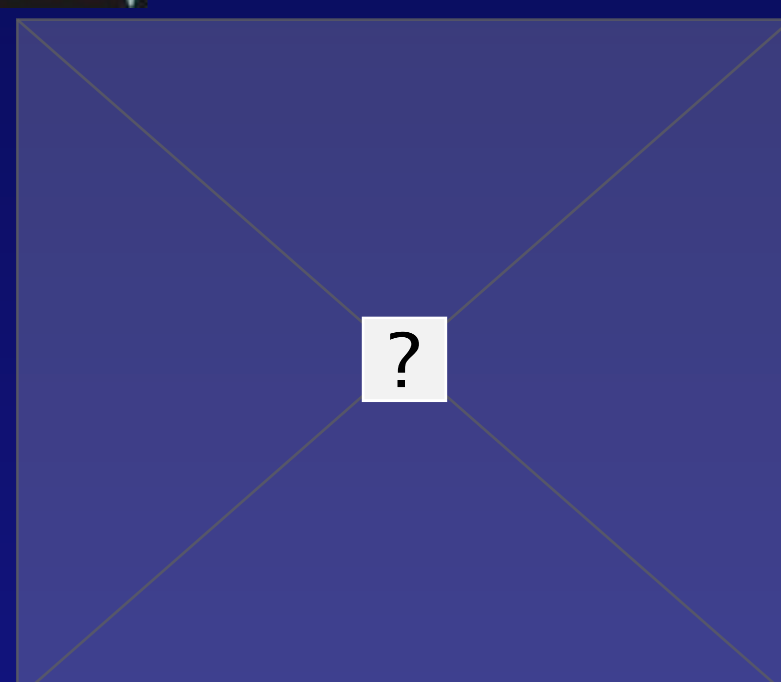
Circ Cardiovasc Imaging published online Jan 8, 2010;



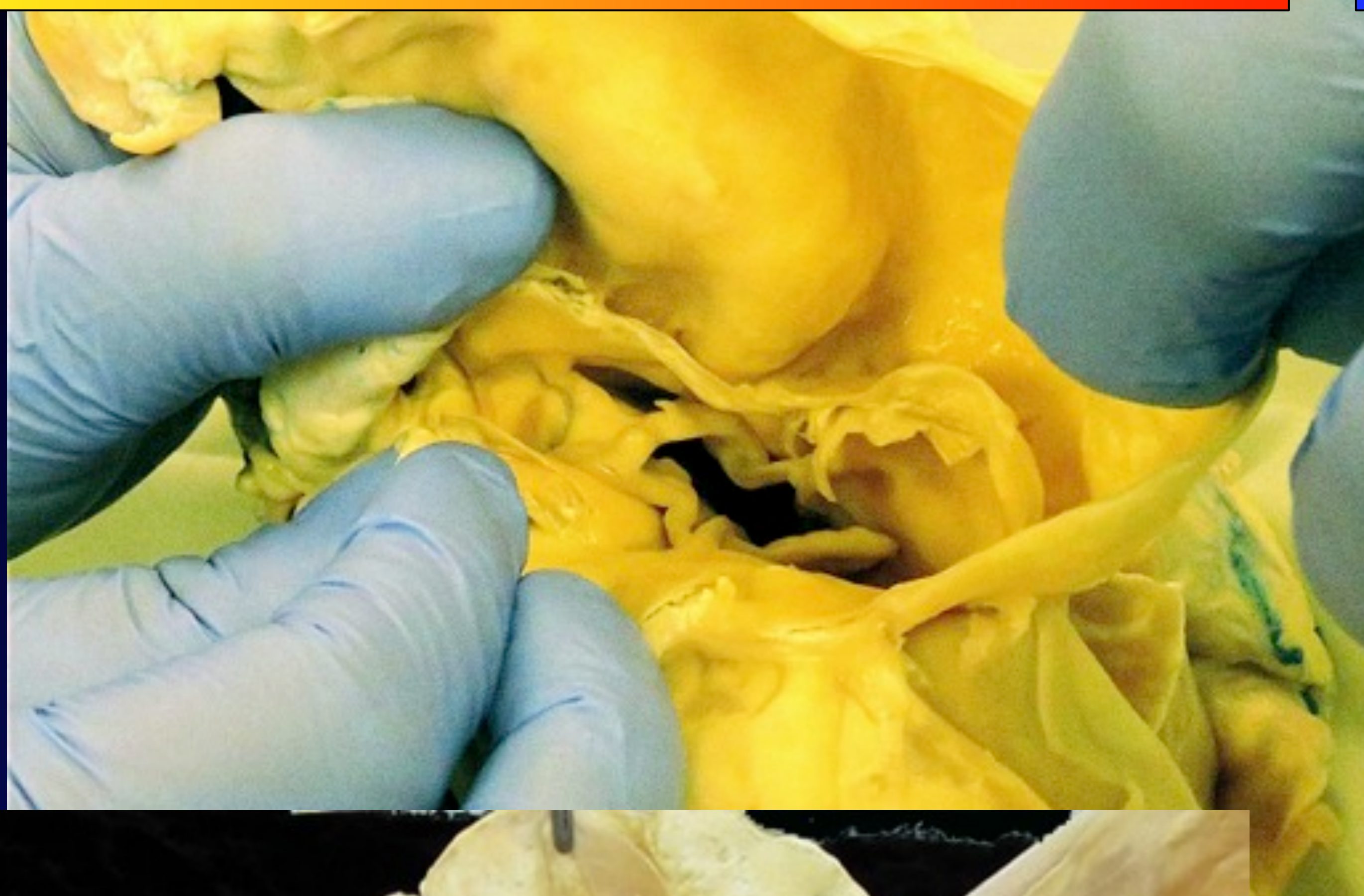
Can Echo Do This?

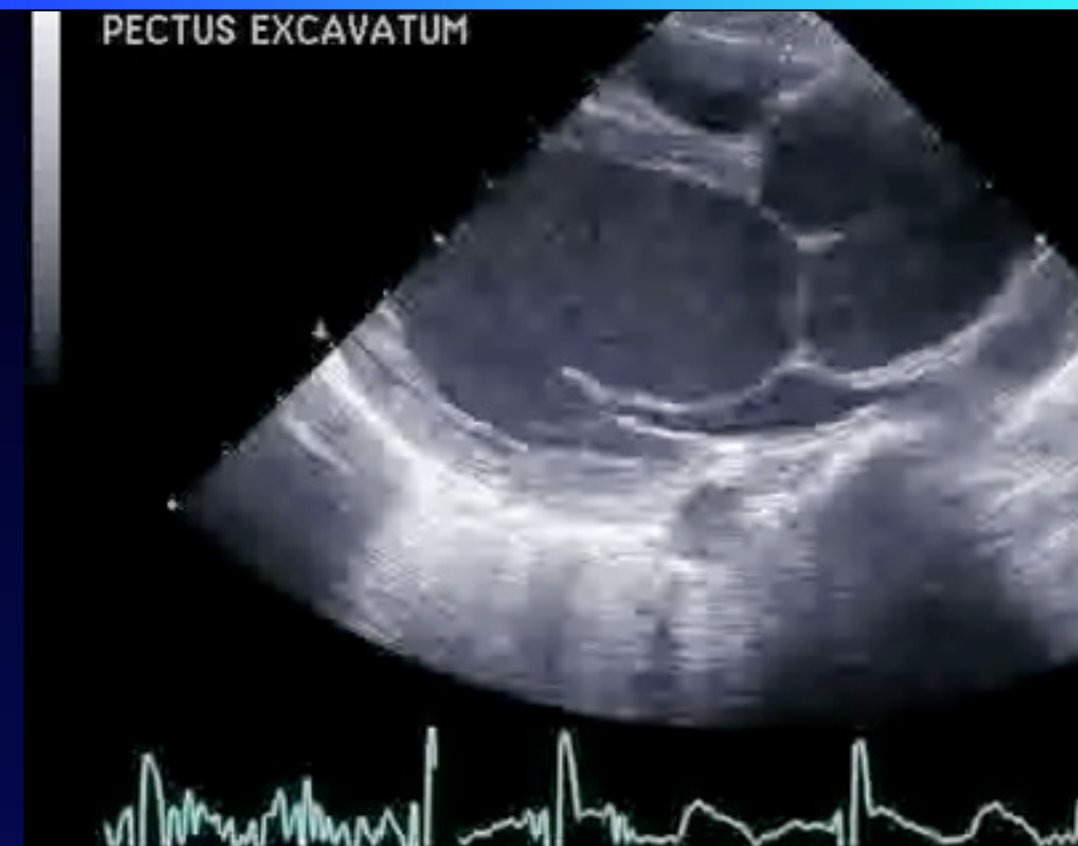


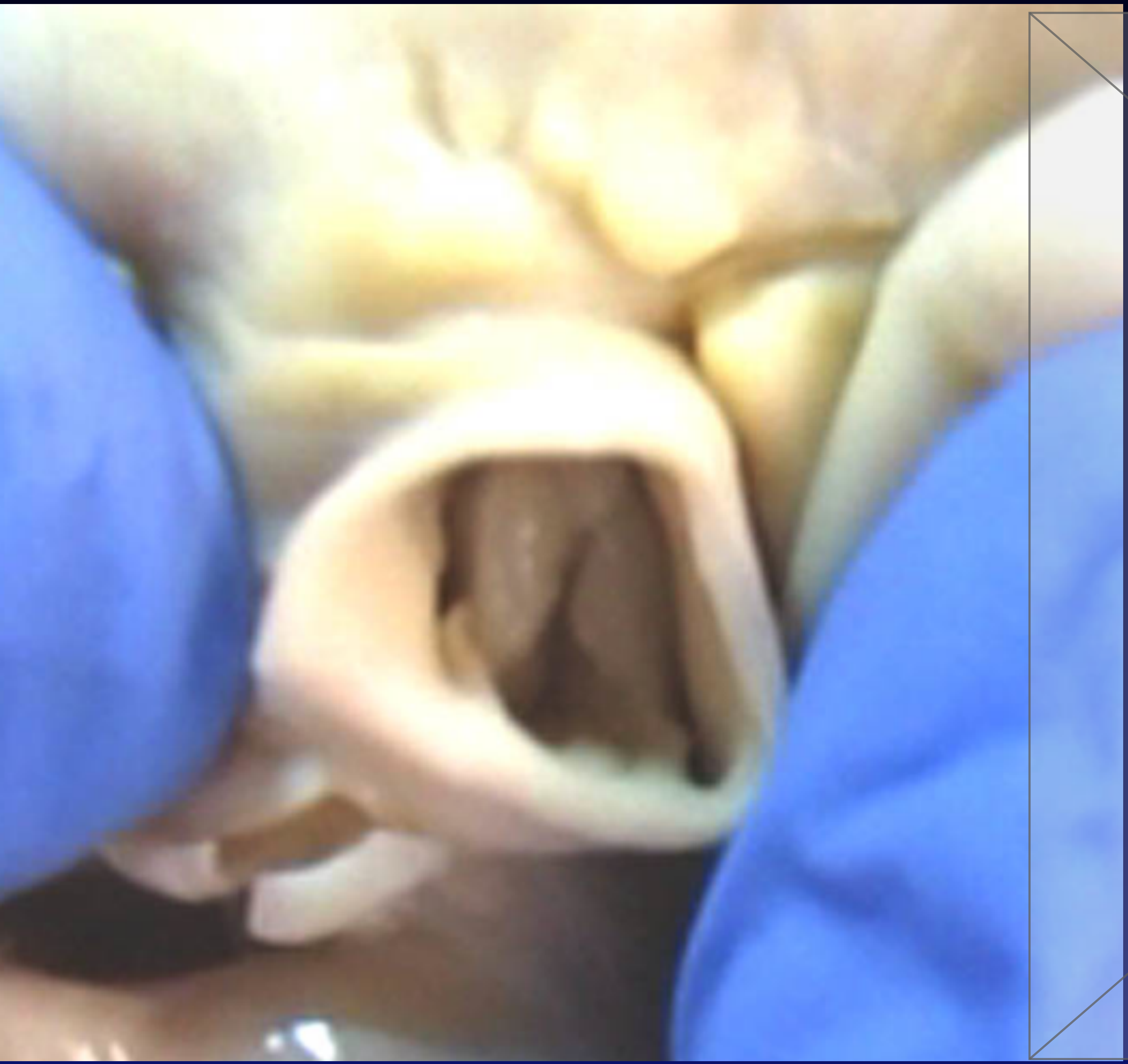



Dilated aortic root and dissection

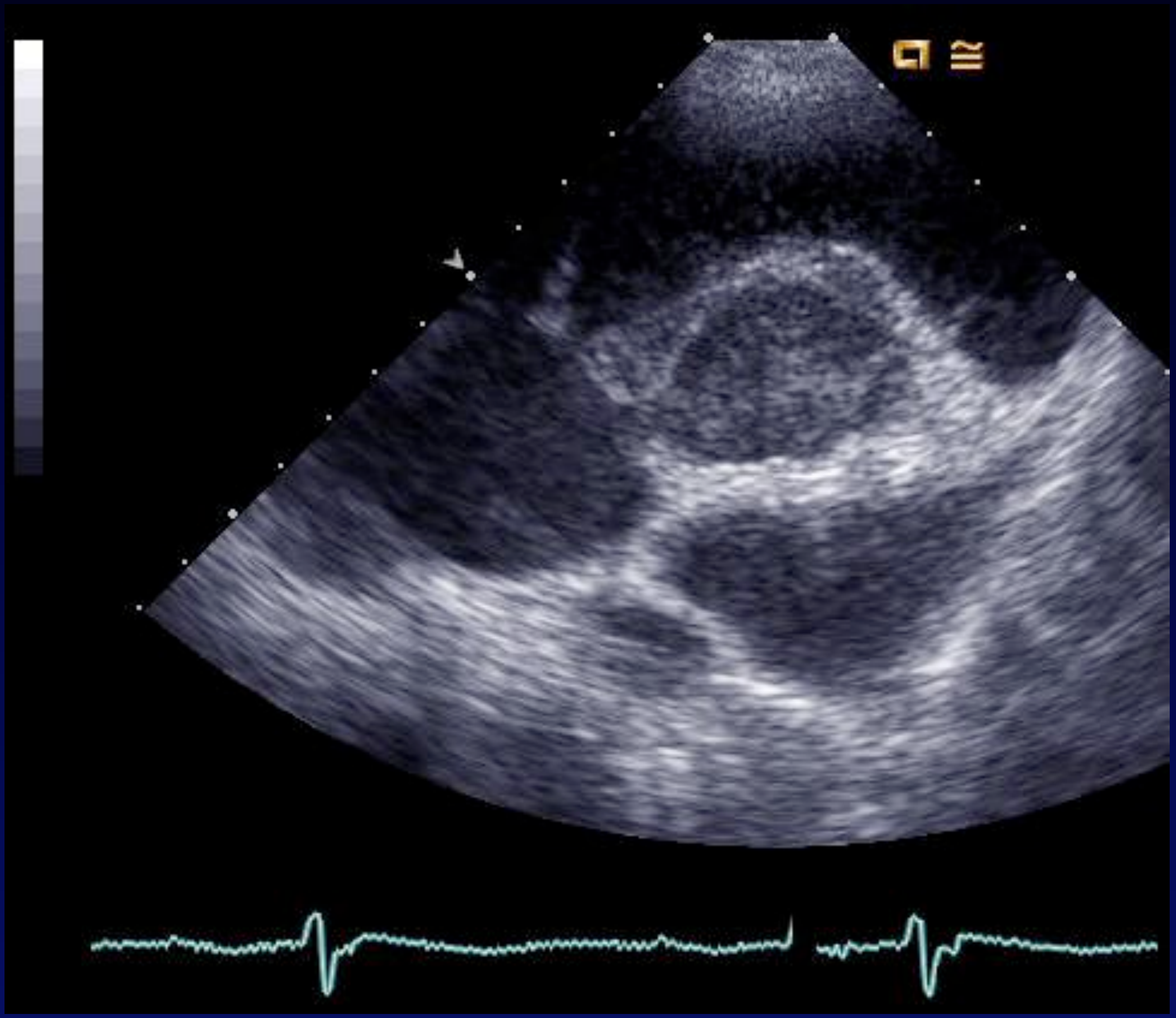




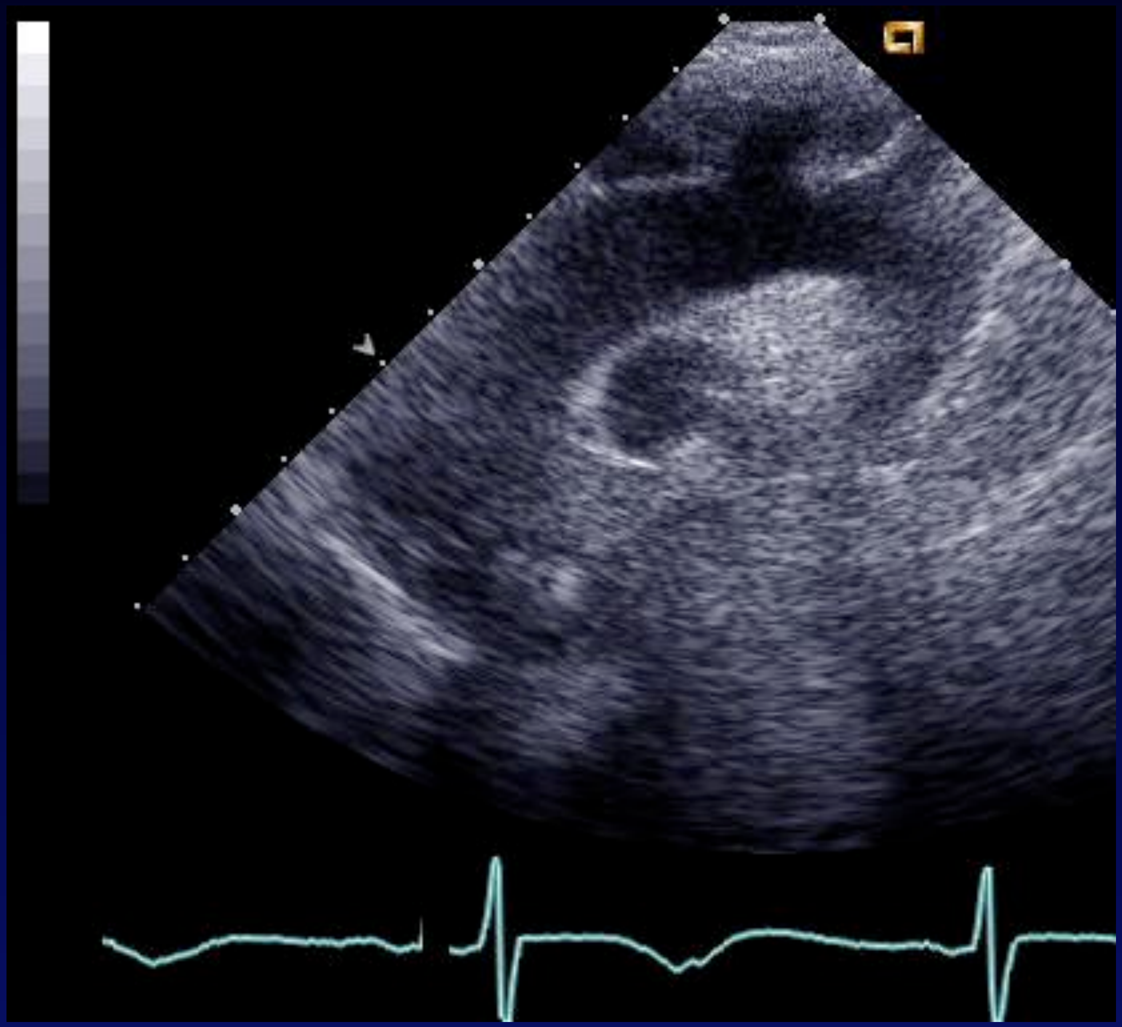
Pseudo Unicuspid Aortic Valve

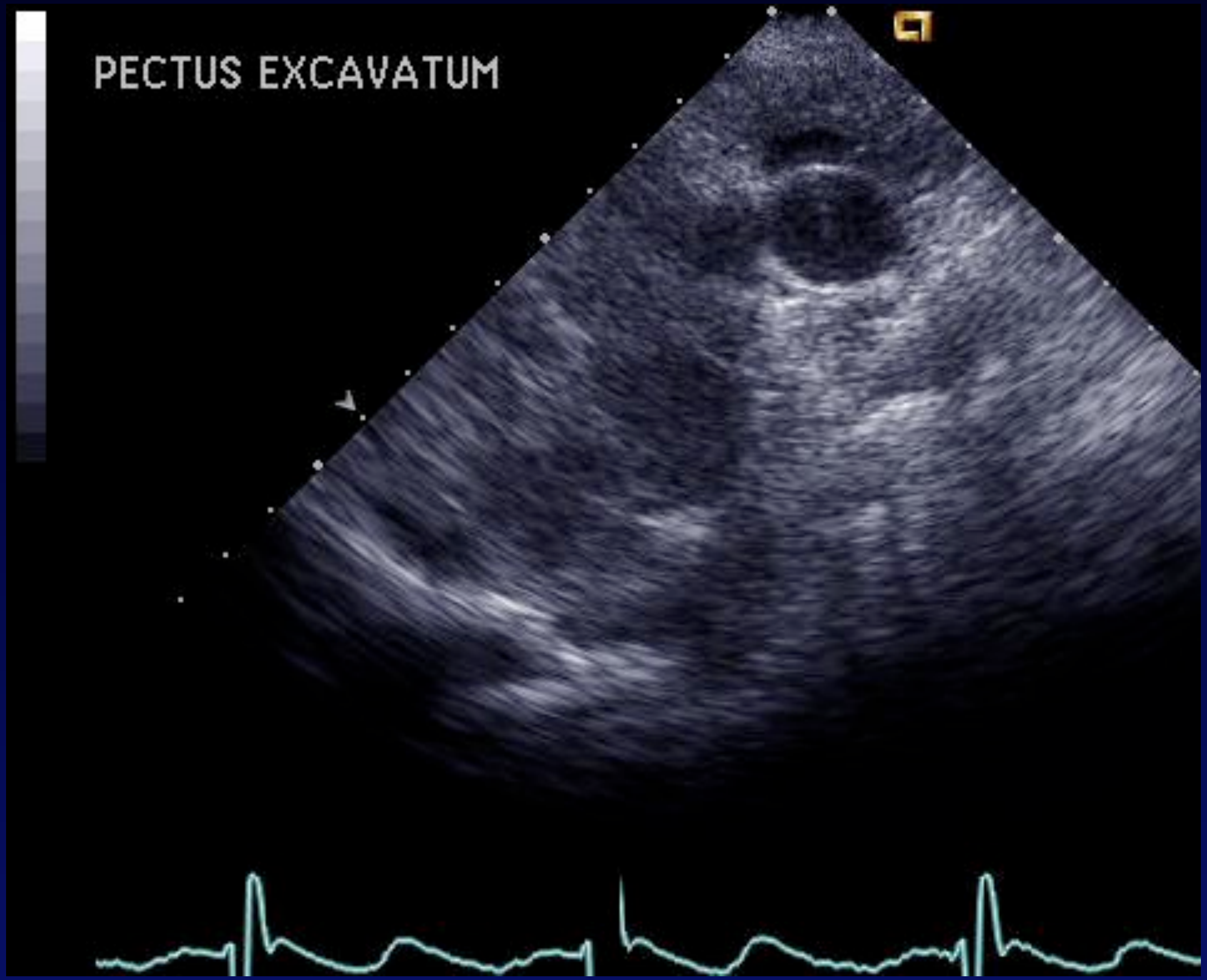


The dilated aortic root in cross section.

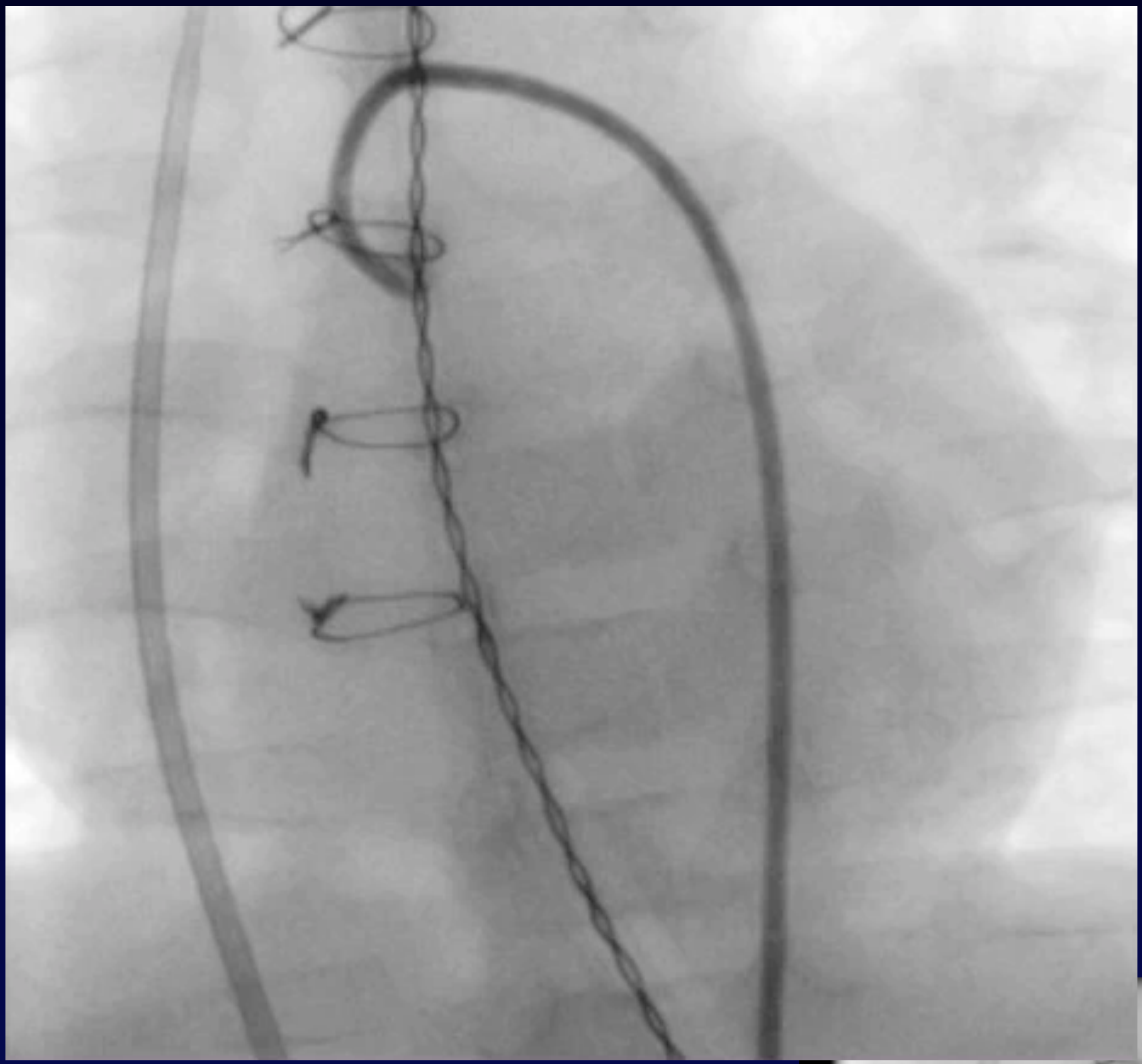


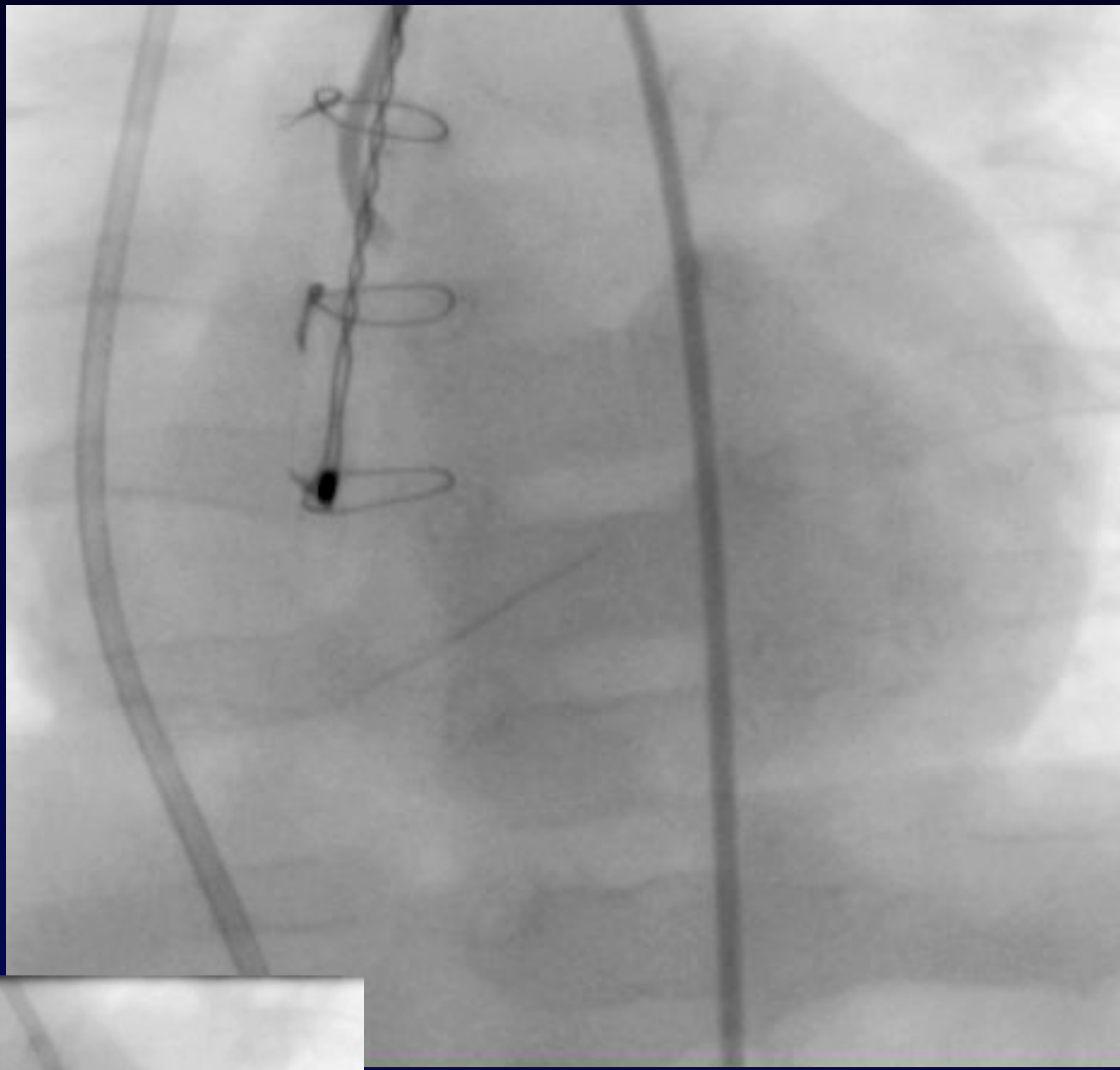
Suprasternal views of dilated aorta

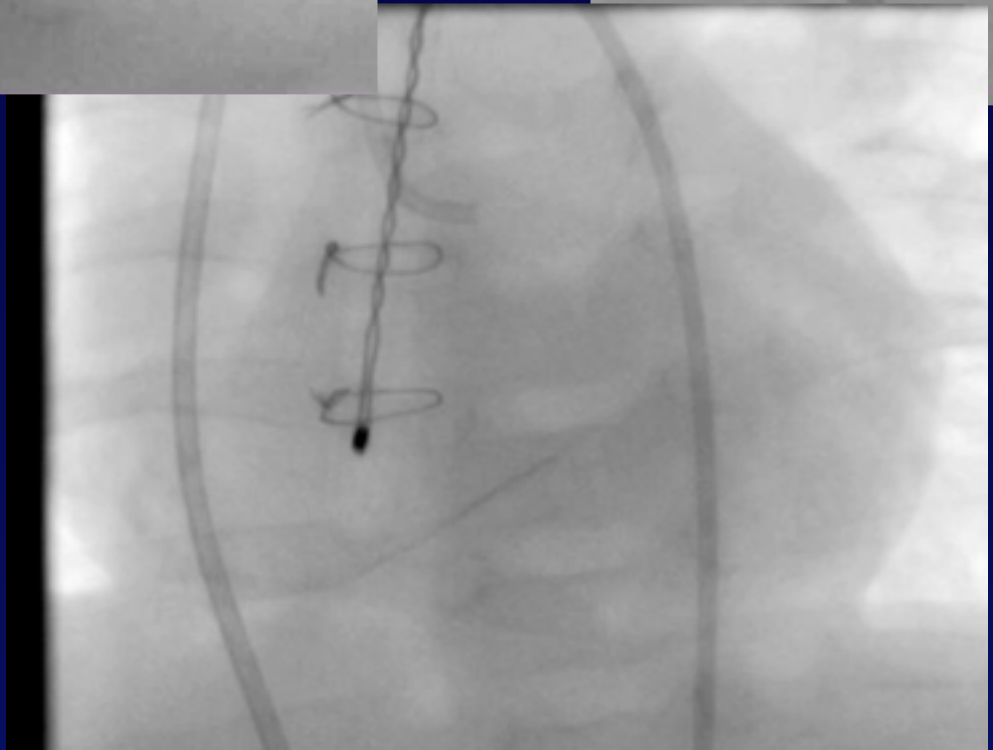




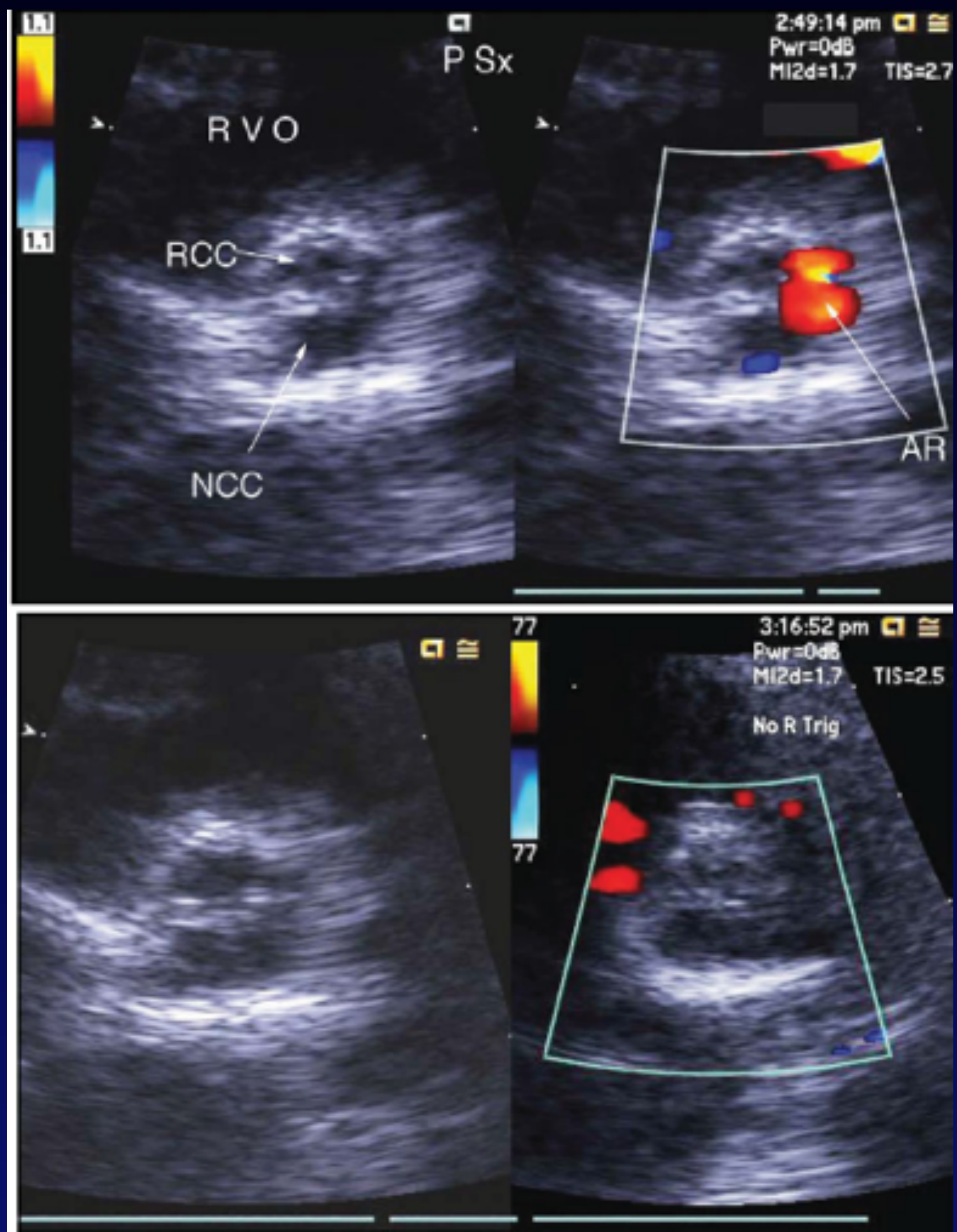
Right & Non- Coronary Cusp Injection

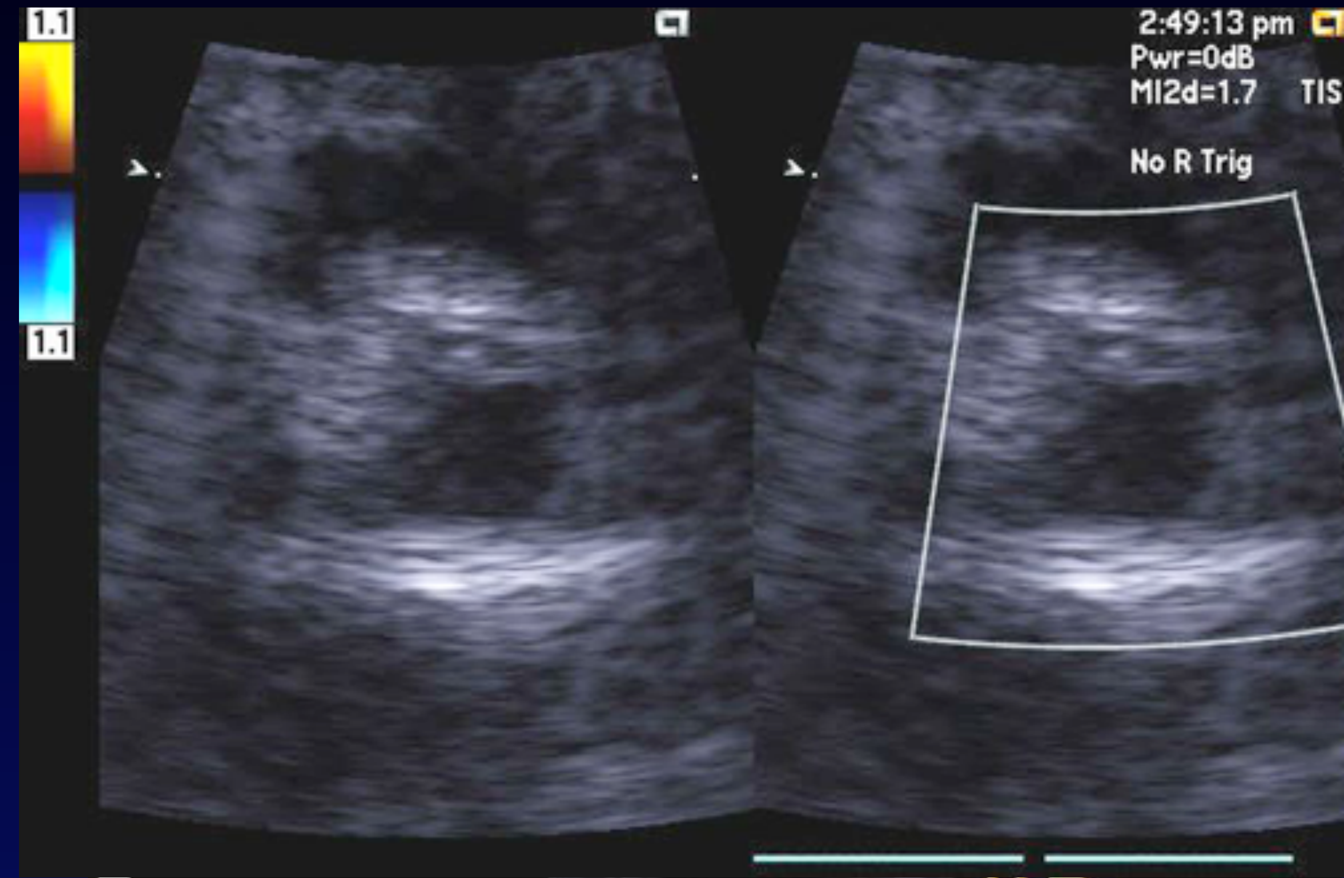


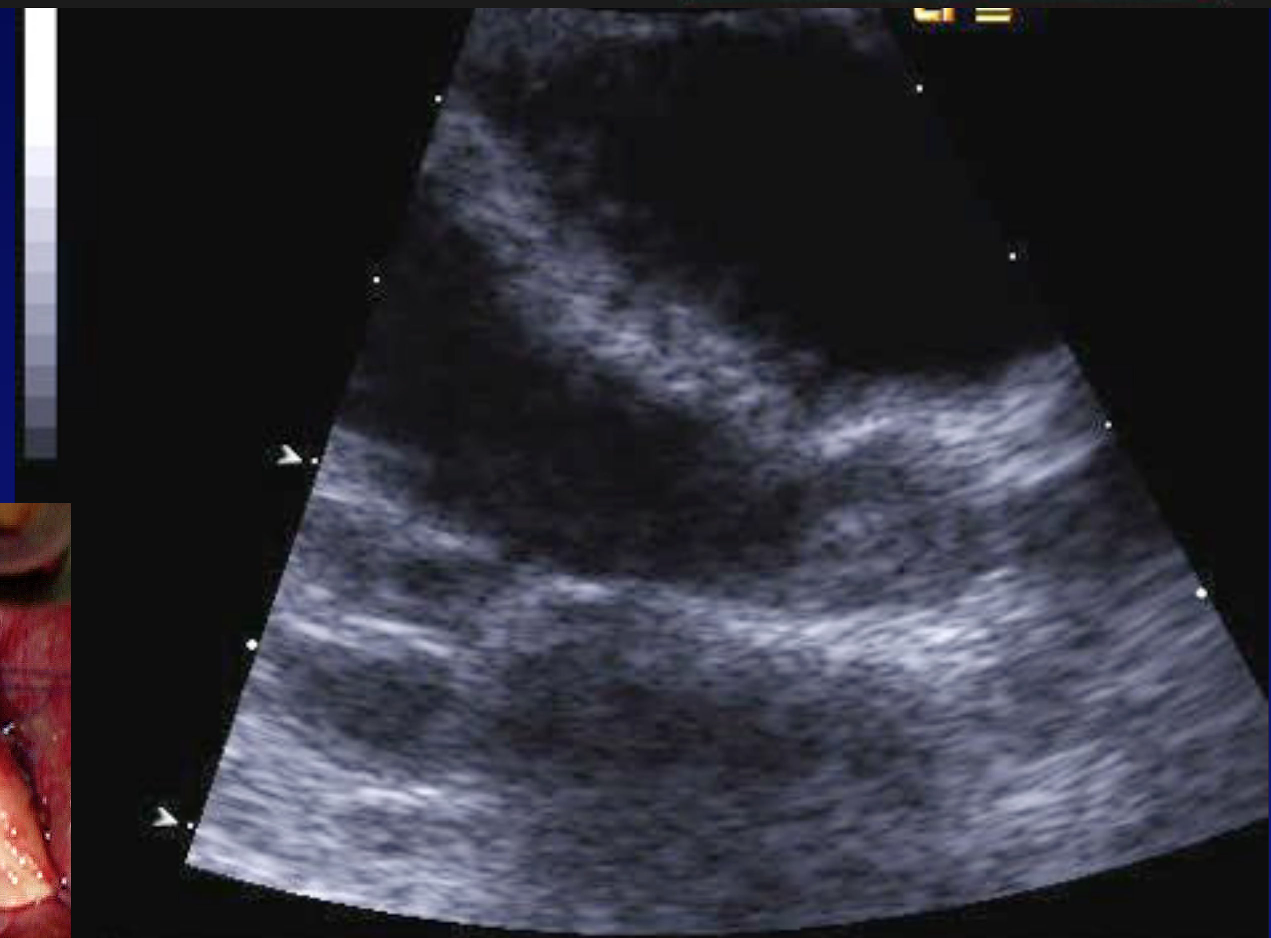


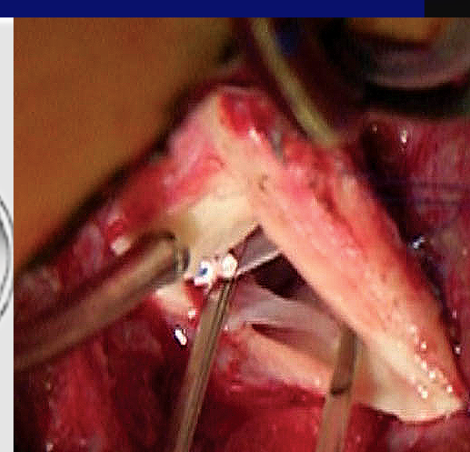


Aortic Regurgitation due to a Hypoplastic Left Sinus of Valsalva.

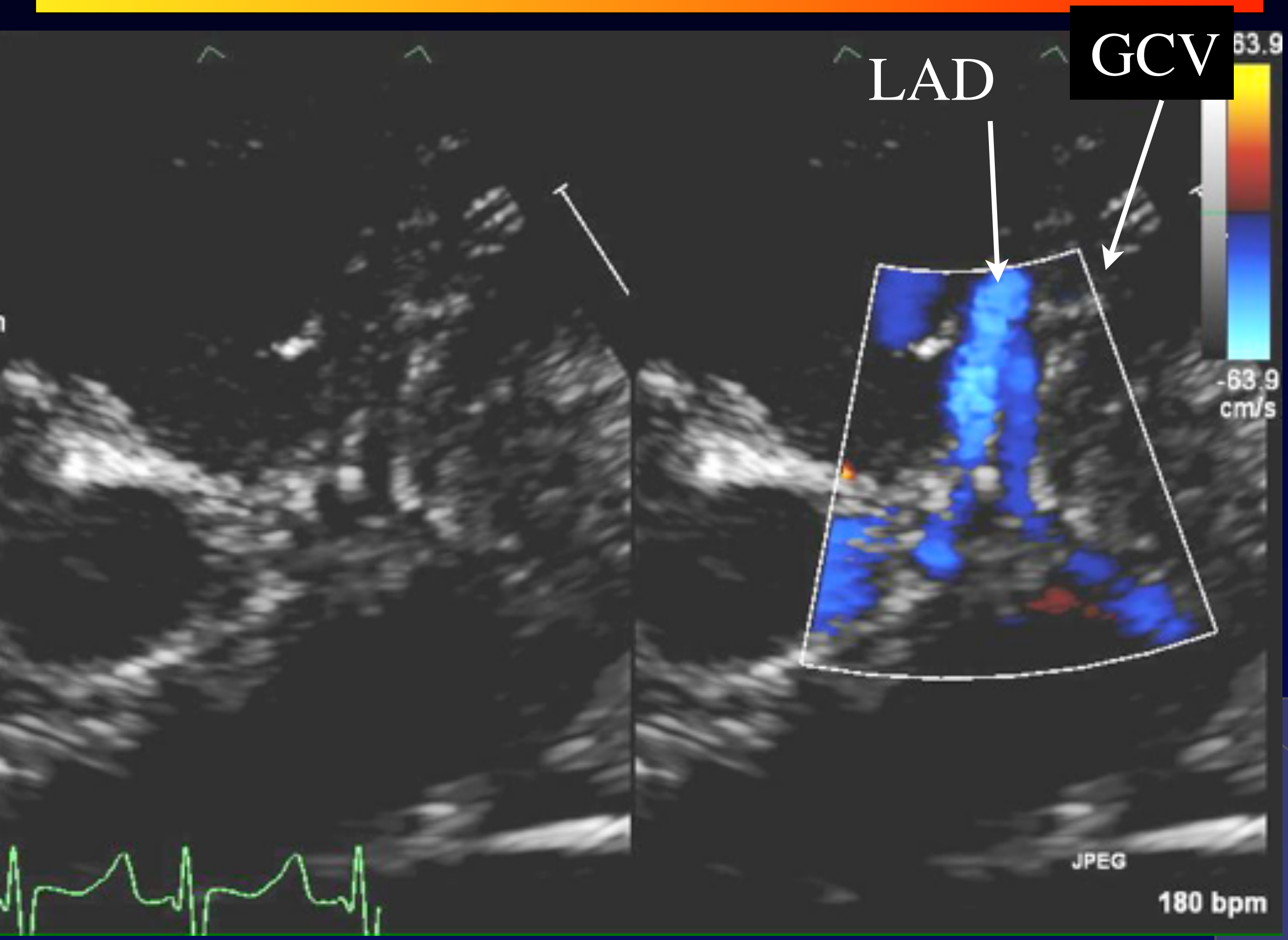








Case 2 Intermittent signs of Ischemia, like ALCAPA



Bellhouse and Bellhouse

Steady flow

With a steady velocity of 62·3 cm/sec in the aorta, the cusps were positioned as in figure 2 with a dividing streamline meeting the sinus ridge at *S*, opposite the centre of the cusp tip. With the use of dye, the vortex pattern, generated by a complex inflow–outflow system at the downstream end of the sinuses, could be observed. Each vortex occupied the entire sinus, but the core was located near

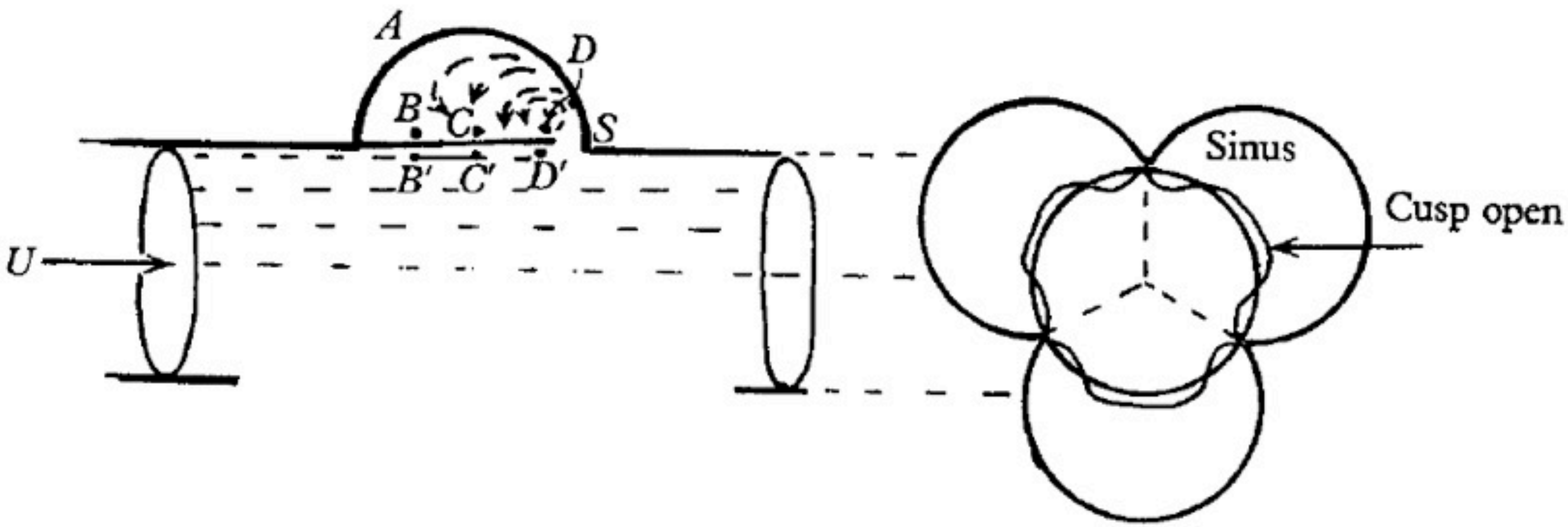
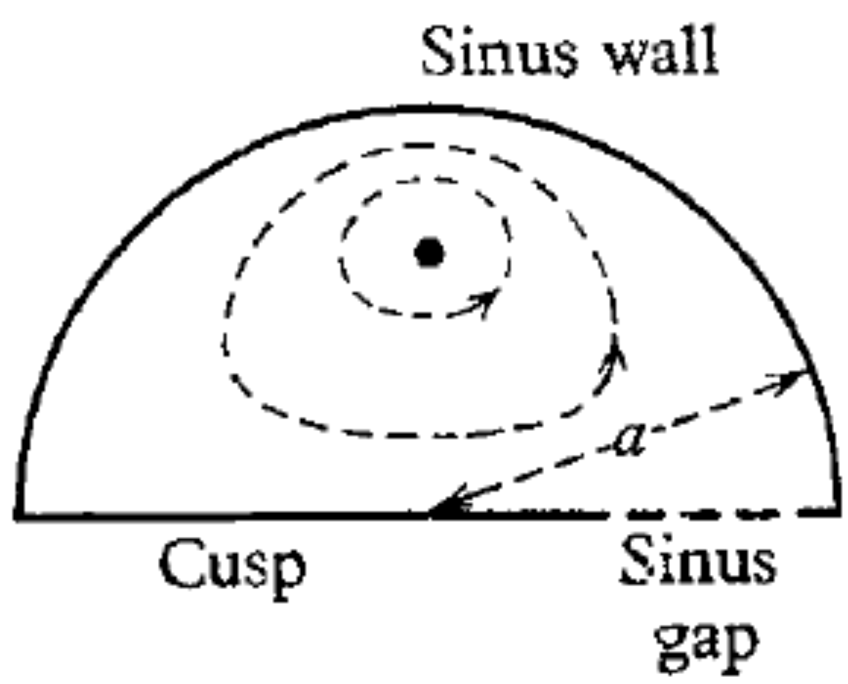


FIGURE 2. Streamlines for steady flow.

Bellhouse and Bellhouse



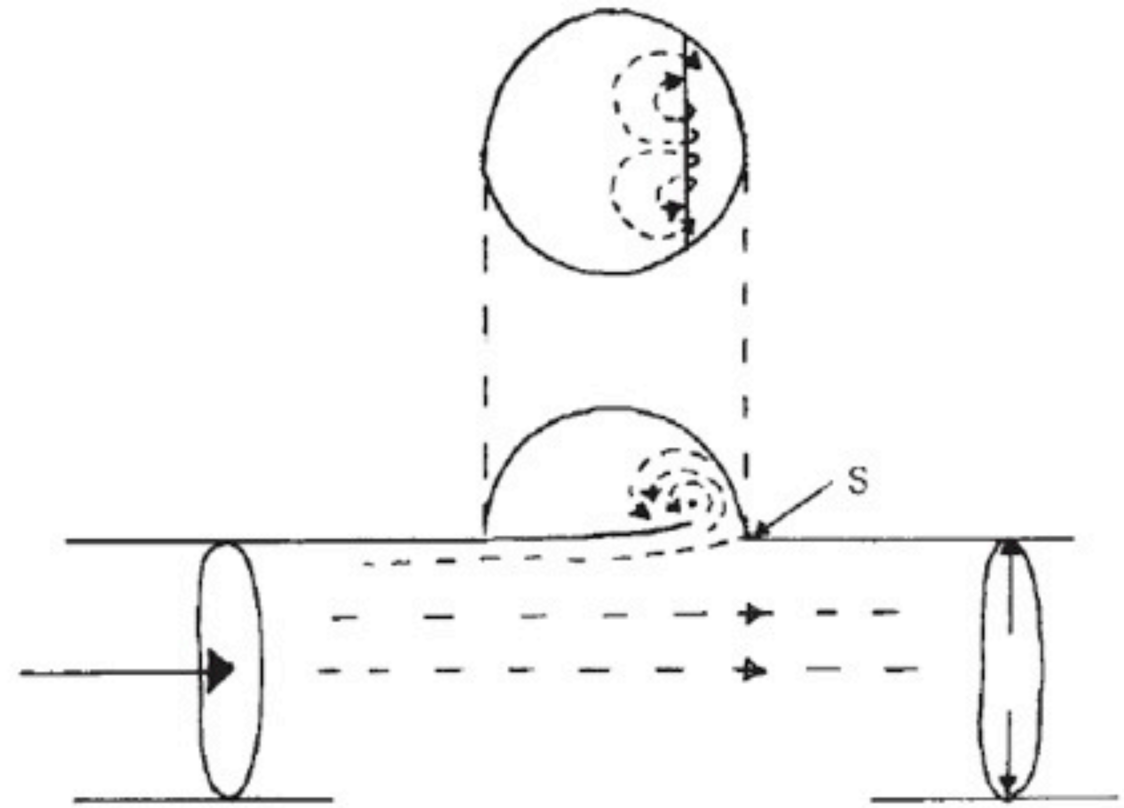


Fig. 1. Quasi-steady phase. Stagnation point at S.

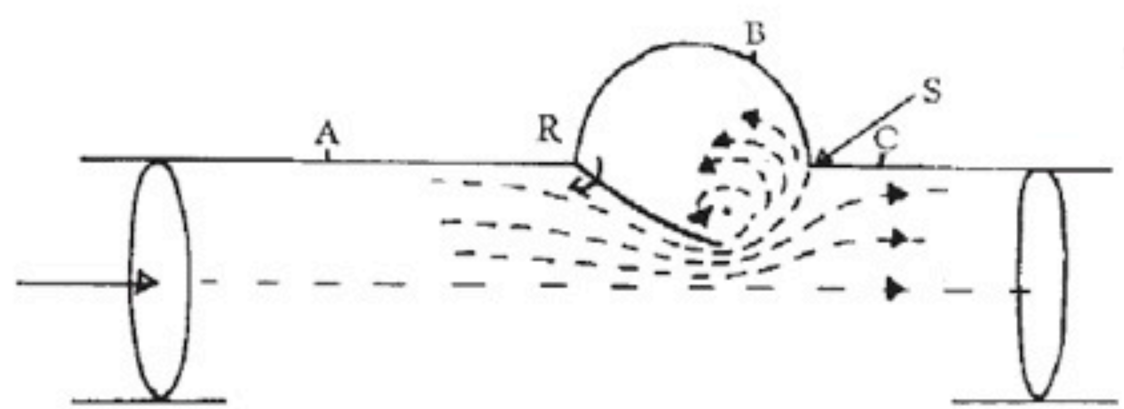
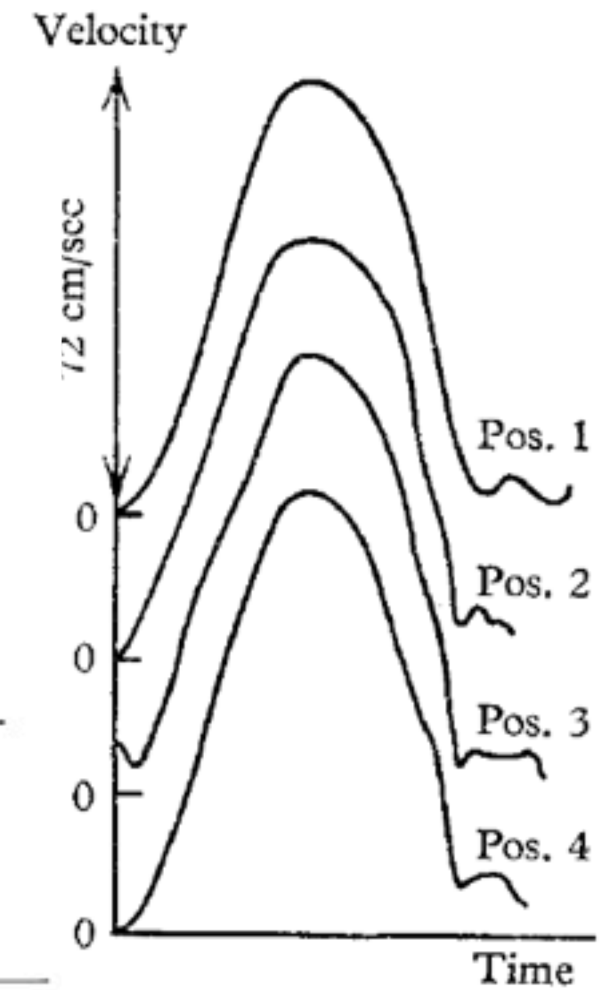


Fig. 2. Deceleration phase.



END