



# A Potpourri of Fascinating Cases to Learn from

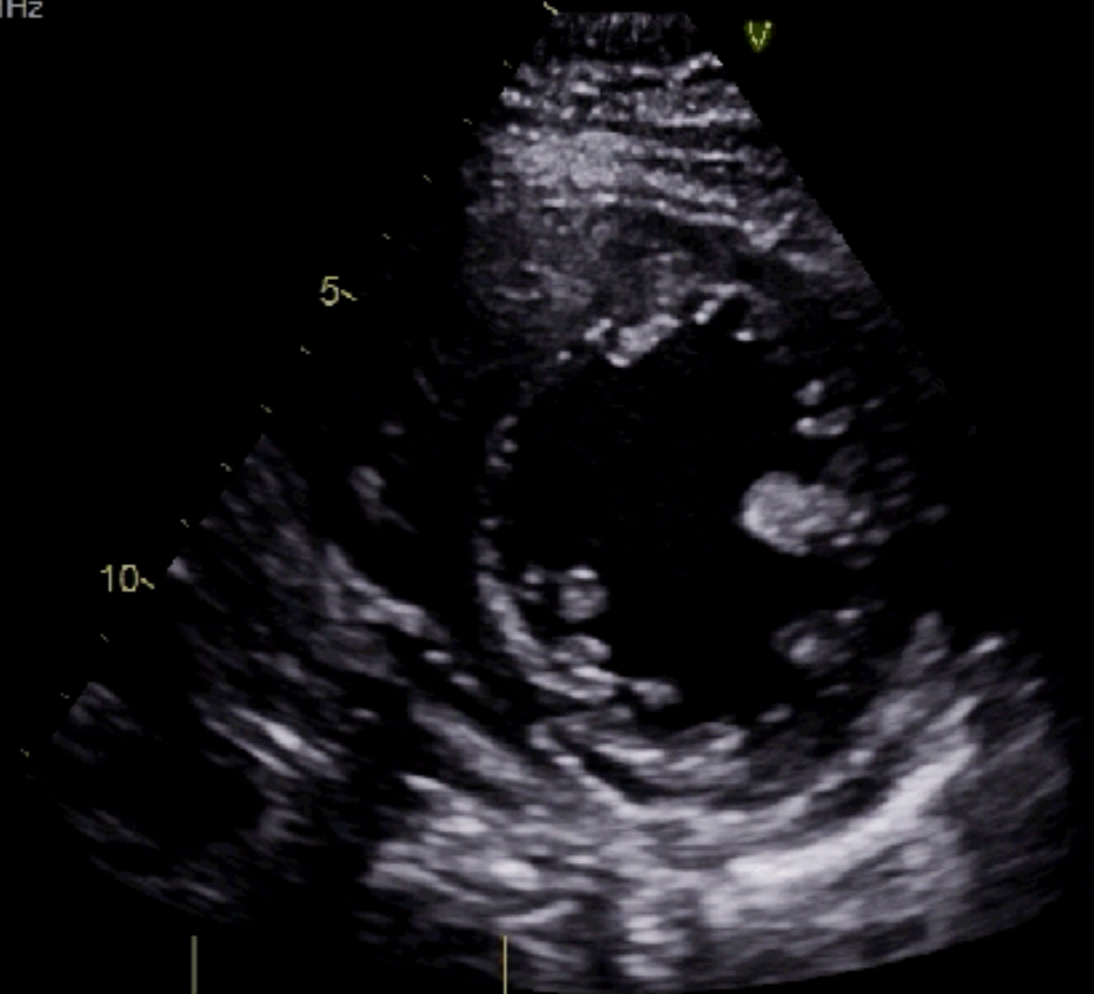
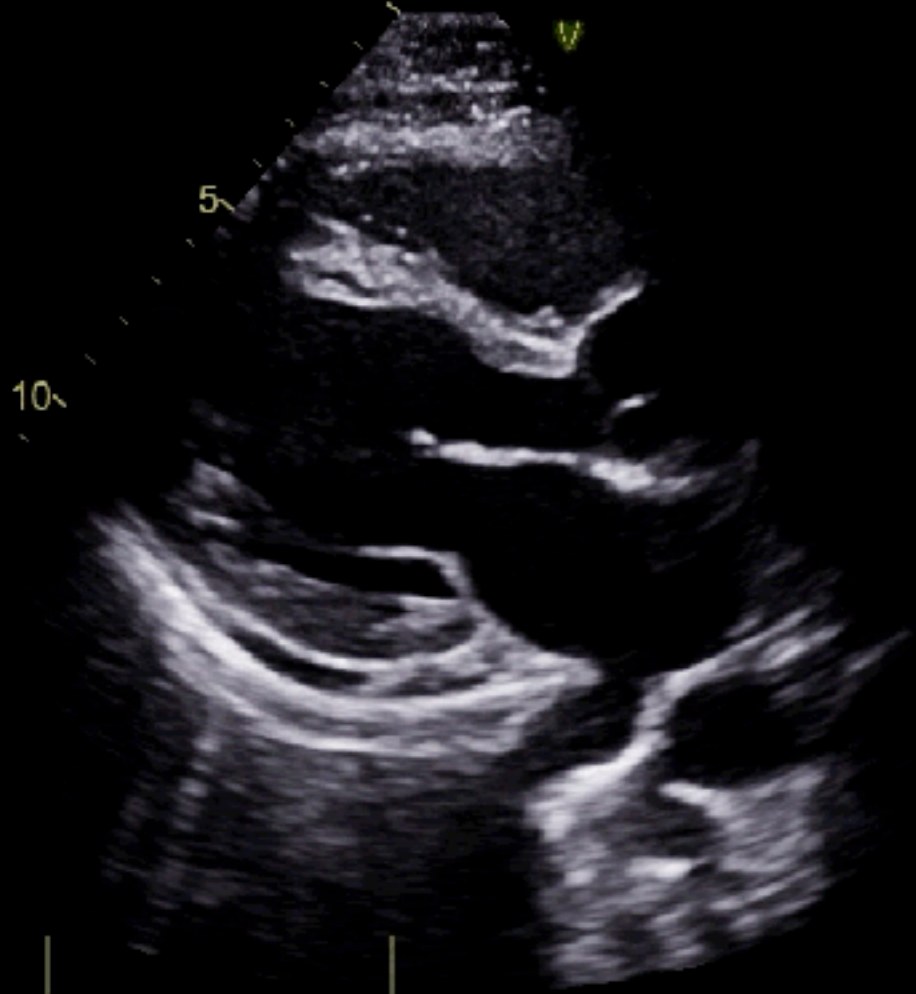
Sangeeta B. Shah MD  
Associate Professor Medicine  
VCU Health  
Richmond, Virginia

# Case 1

- 24 y.o. woman referred by cardiology for history of congenital heart disease and 'drop attacks'
- Age 5 had percutaneous closure of secundum ASD with 33mm CardioSeal
- Age 17 admission for hemoptysis
- Poor functional capacity.
- Vital BP 102/69 both arms; Pulse Ox 98%; HR 76 bpm; BMI of 37kg/m<sup>2</sup>
- Physical exam 2/6 holosystolic at apex and trace LE edema
- Meds: Spirinolactone and lactulose

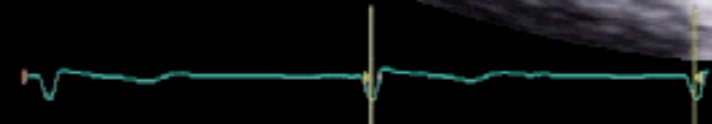
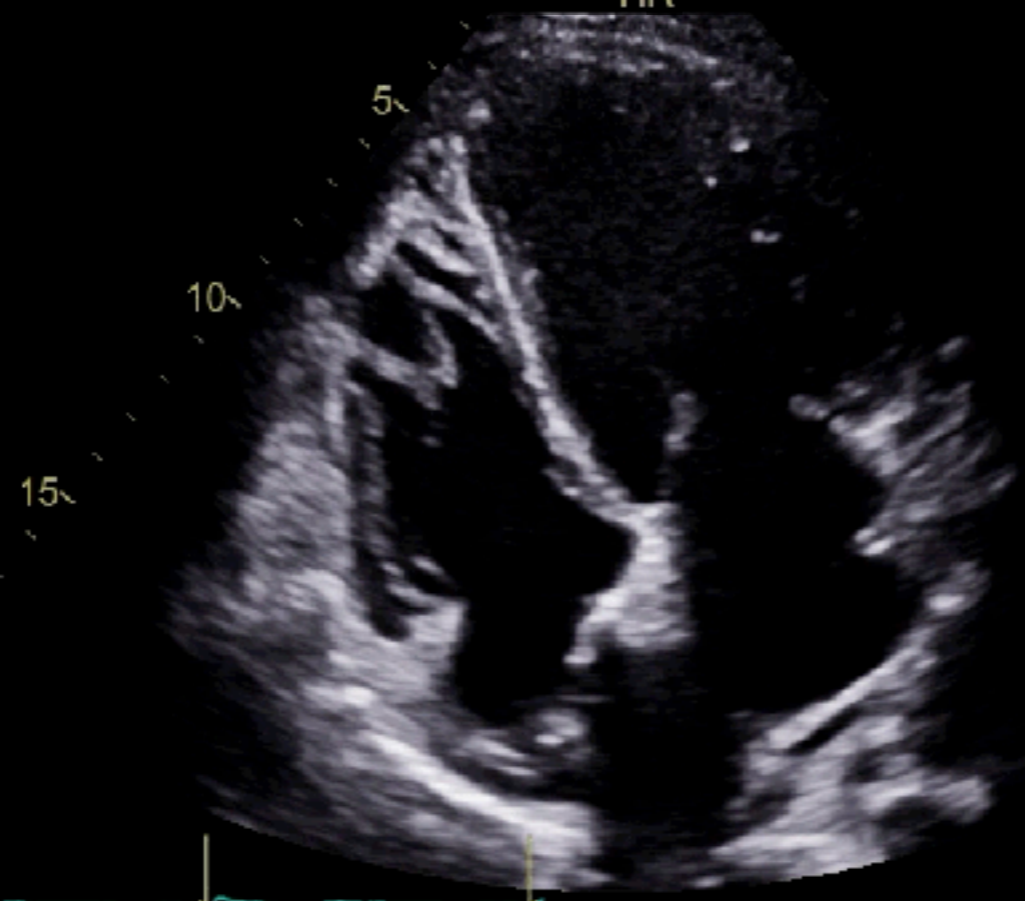


3.577  
1.7 MHz/3.3 MHz  
3(t): 3 dB  
impr: 60 dB  
14.0 cm



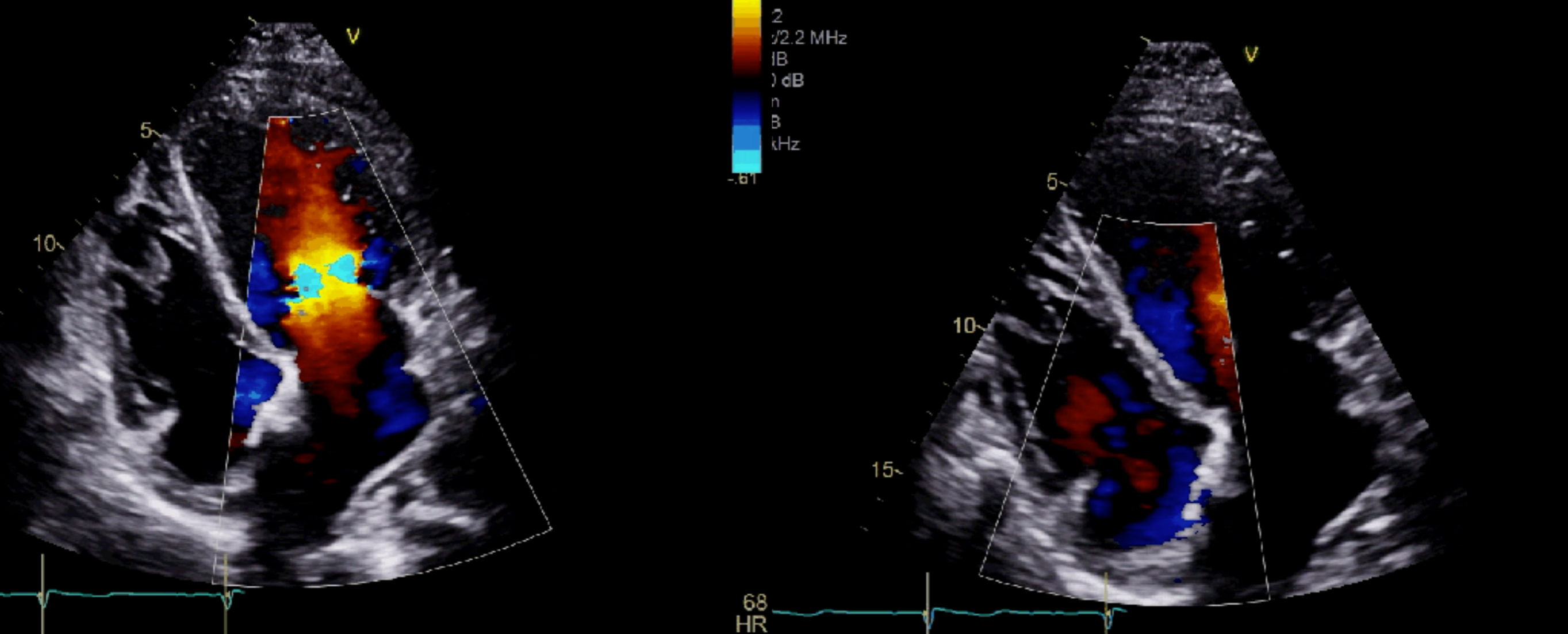
U: 19.0 cm

66  
HR

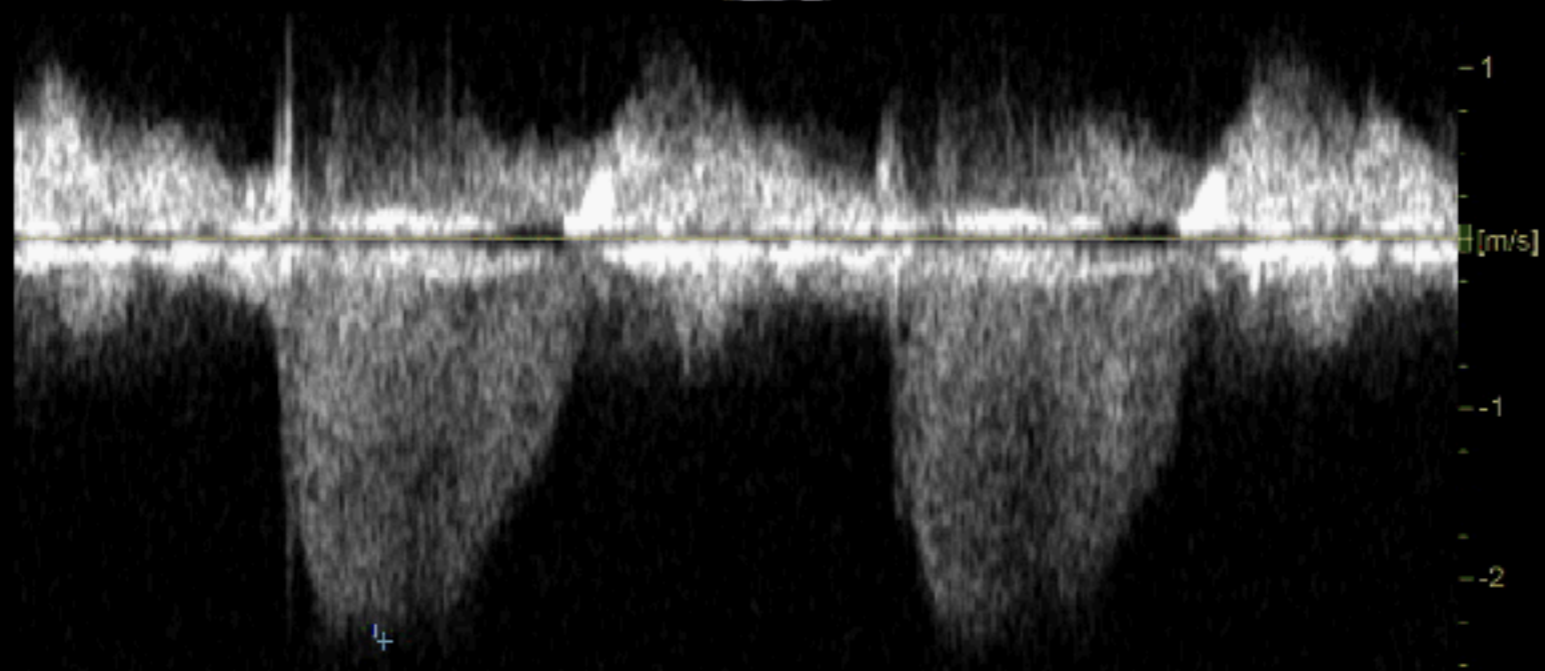
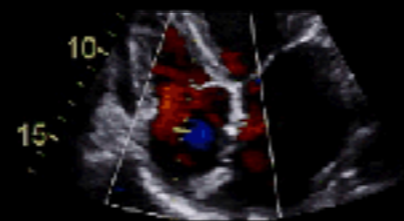


67  
HR





AG(t): 3 dB  
 Compr: 60 dB  
 D: 17.0 cm  
 G(c): -4 dB  
 PRF: 3.5 kHz  
 G(d): 0 dB



M5Sc  
YCX Echo

MI 1.4  
TIs 0.8

Soft

ACE  
FPS: 57  
f: 1.7 MHz/3.3 MHz  
AG(t): 0 dB  
Compr: 60 dB  
D: 18.0 cm

**Saline Contrast**

**REST**



71  
HR

# TTE

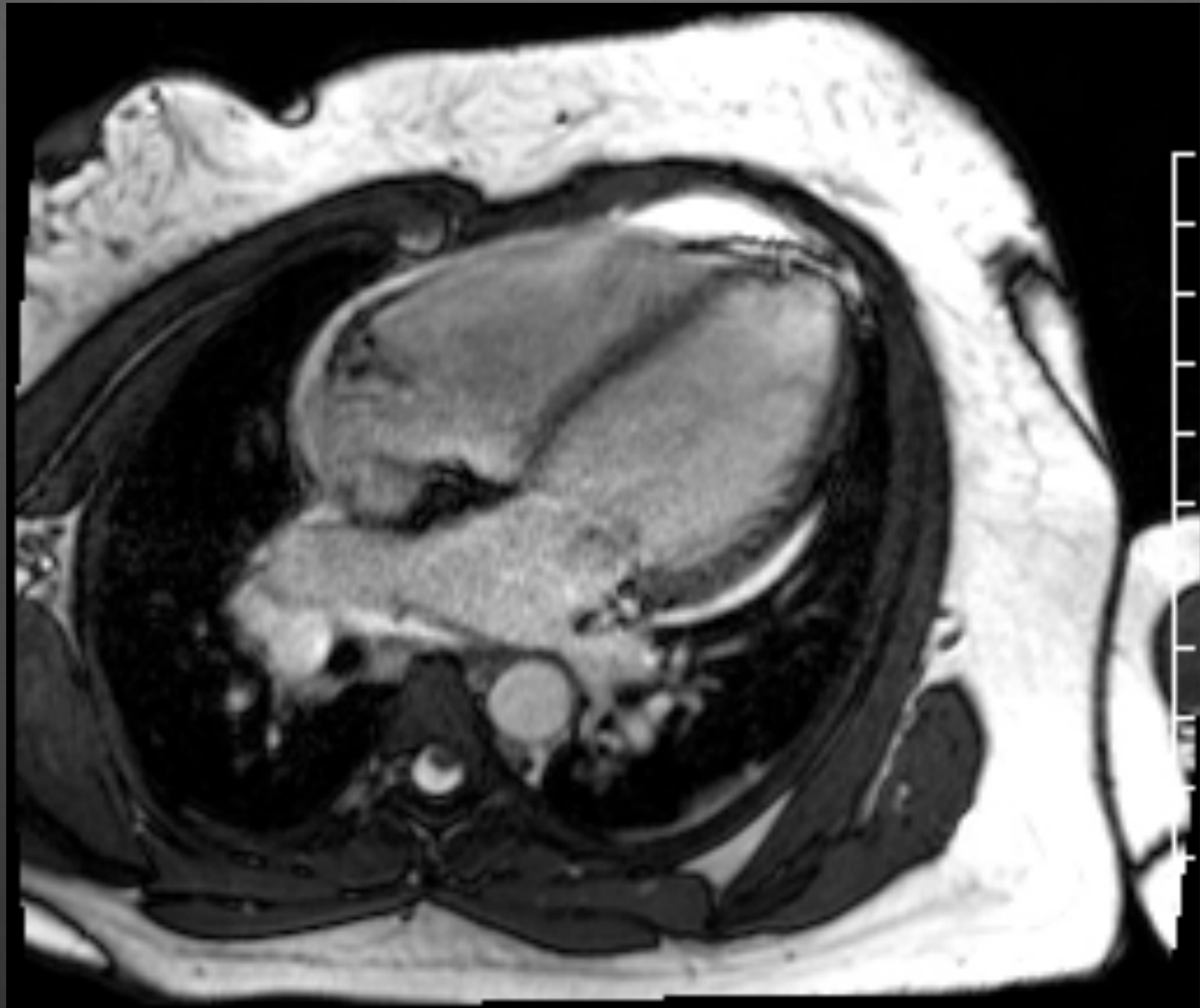
- Mildly dilated right heart with normal RV systolic function
- Mild TR with estimated RV systolic pressure of 27mmHg
- Severe LAE (66cc/m<sup>2</sup>)
- Mild to moderate MR
- LVEF of 55-60%
- ASD device noted. Late shunting suggestive of pulmonary AVM



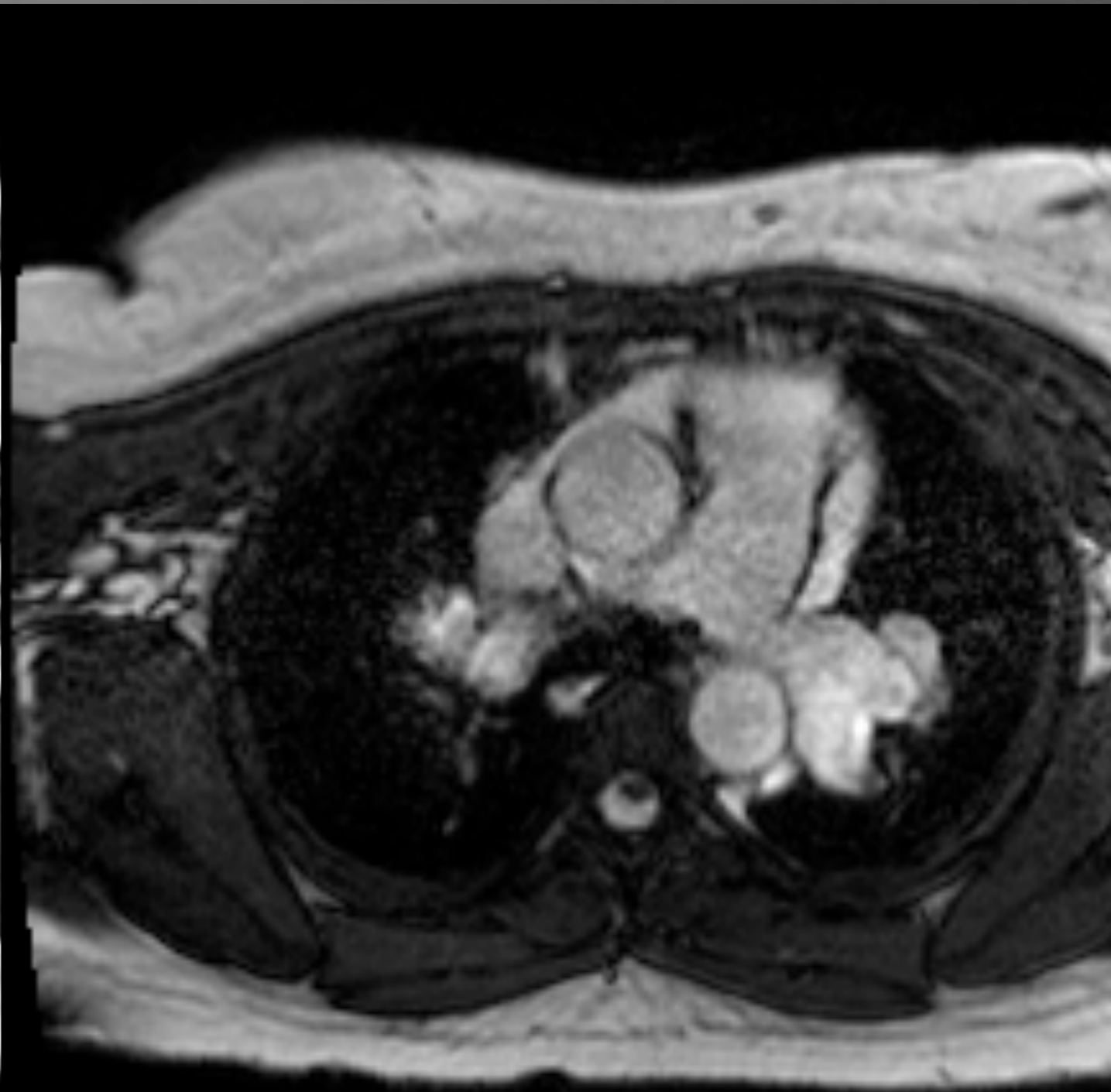
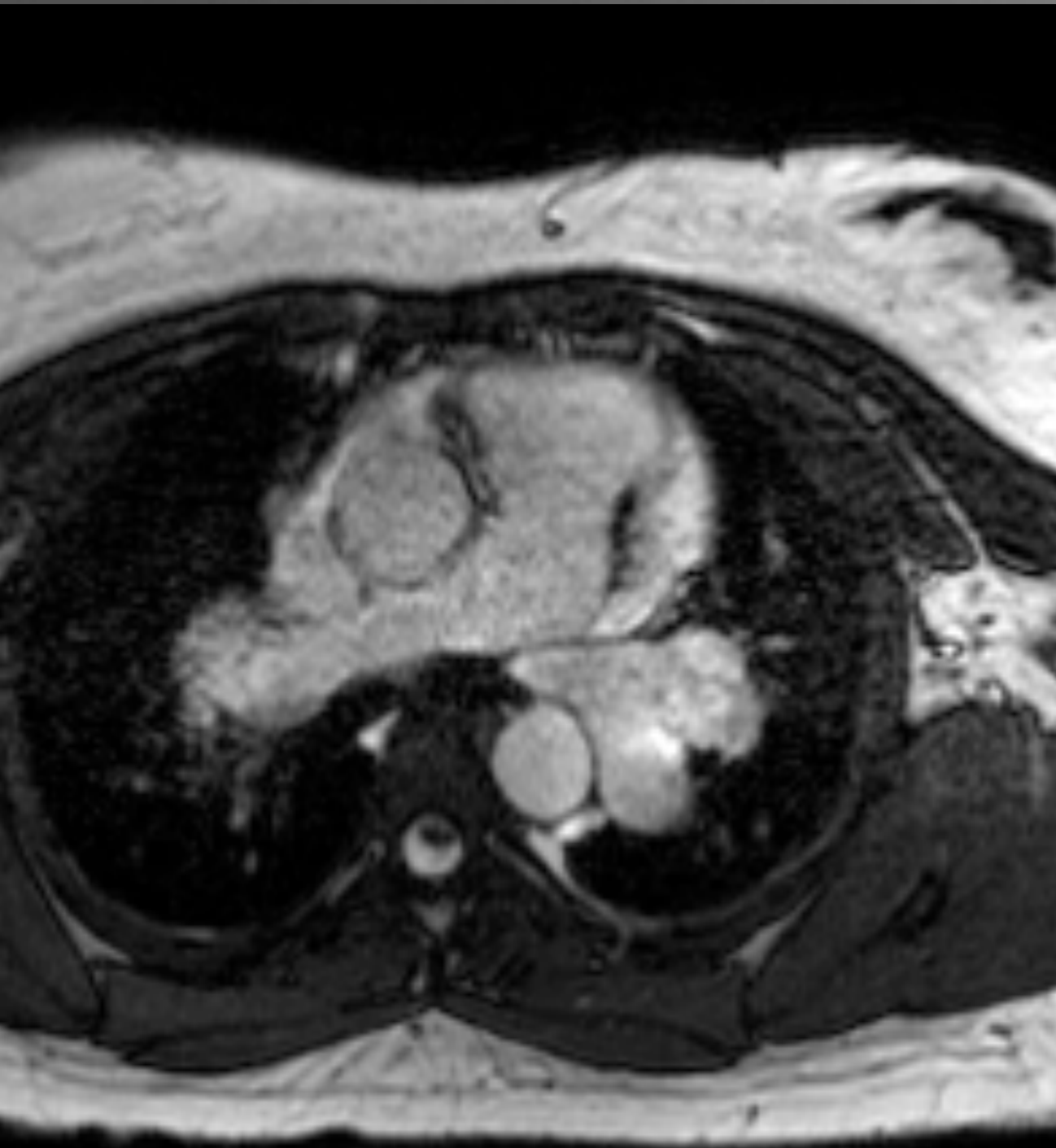
# CPET

- Peak  $\text{VO}_2$  14.5 mL/kg/min (RER 1.11)
- Hypotension with exercise 89/63mmHg (baseline 105/66)
- No hypoxemia











**Aorta**  
**111 ml/beat**



**PA**  
**130 ml/beat**



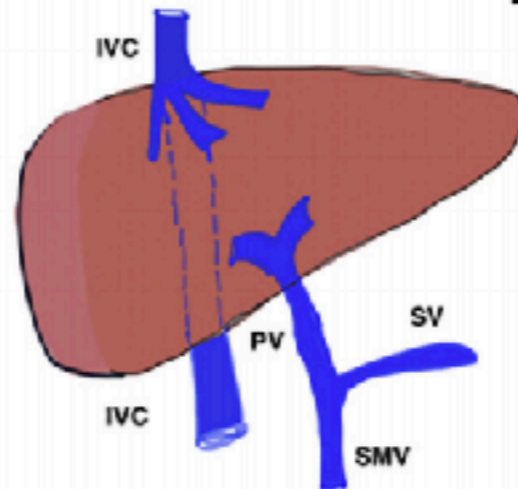
# Cardiac MRI

- RAE
- LAE
- Normal RV volumes with RVEF of 47%
- Increased LV volumes with LVEF of 55%
- Dilated PA with greater flow in the right PA >> Left PA with mild LPA stenosis
- Qp:Qs 1.2:1
- Nodularity of liver

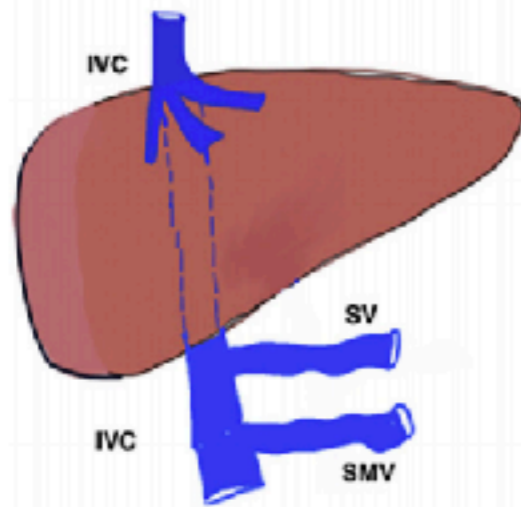


# Abernethy

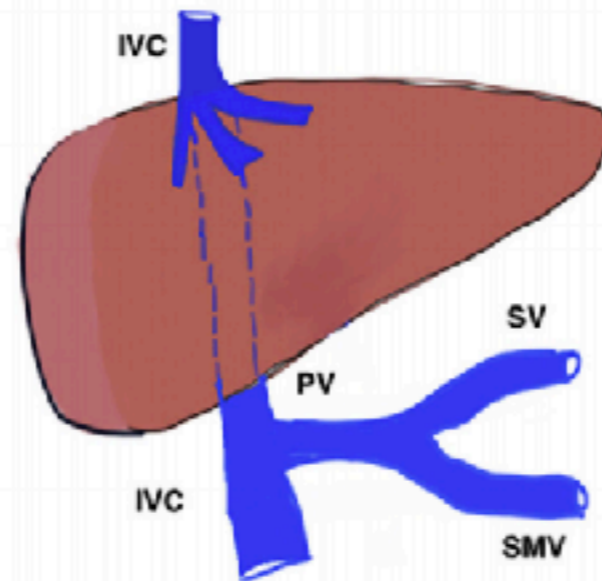
## Normal anatomy



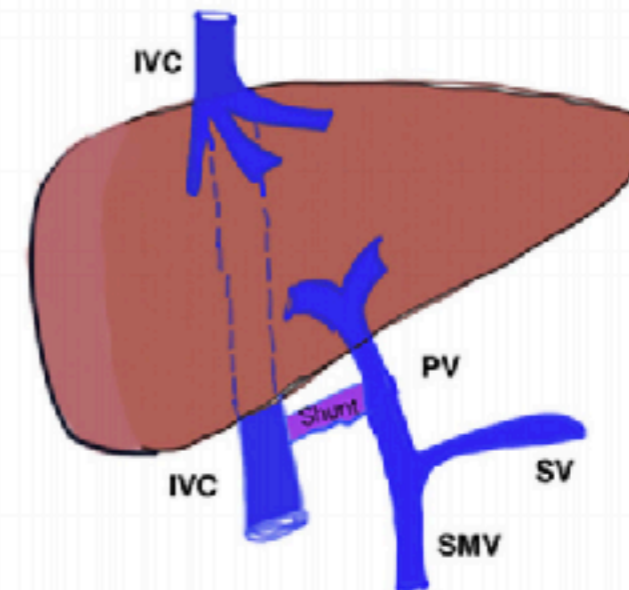
## *Abernethy malformation*



**Type 1a**



**Type 1b**



**Type 2**



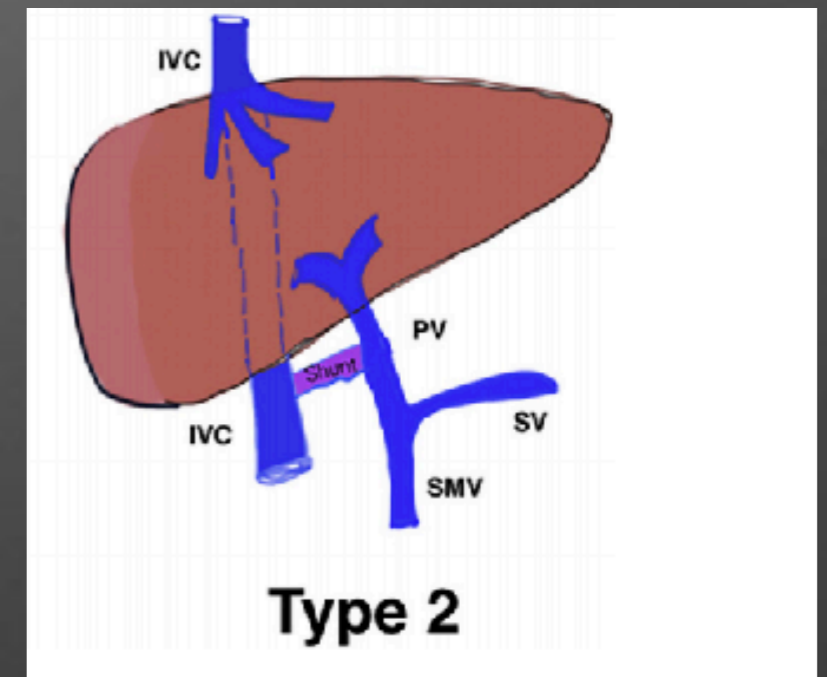
# Symptoms

- Hepatic encephalopathy
- Hepatopulmonary syndrome
- Pulmonary hypertension
- Nodular Liver Lesions- HCC and adenomas



# Plan

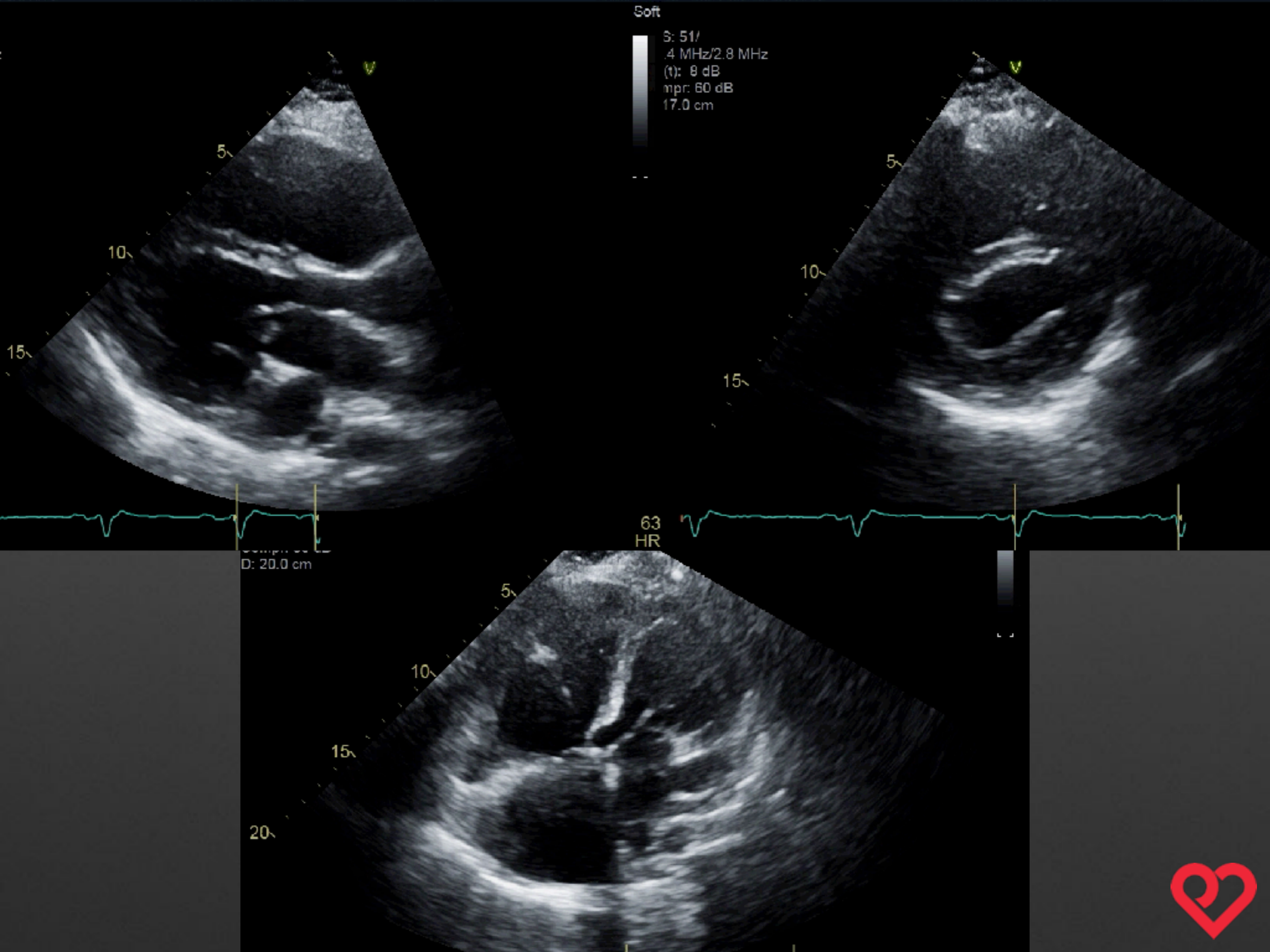
- Monitor for exercise hypoxemia
- AFP/Liver MRI
- Pulmonary hypertension
- Can consider closure of the shunt with IR



# Case 2

- 35 y.o. woman referred by CTS for history of congenital heart disease and severe tricuspid regurgitation
- Tetralogy of Fallot with AV Canal Defect
- Age 6 repair of AV canal defect and surgical pulmonary valvulotomy and one month later repair of TV with annuloplasty ring
- Poor functional capacity. TIA dx with atrial fibrillation with DCCV
- Vital BP 122/76 R and 104/74 L; Pulse Ox 98%; HR 77 bpm; BMI of 28kg/m<sup>2</sup>
- Physical exam 2/6 holosystolic at RLSB
- Meds: Eliquis





Soft

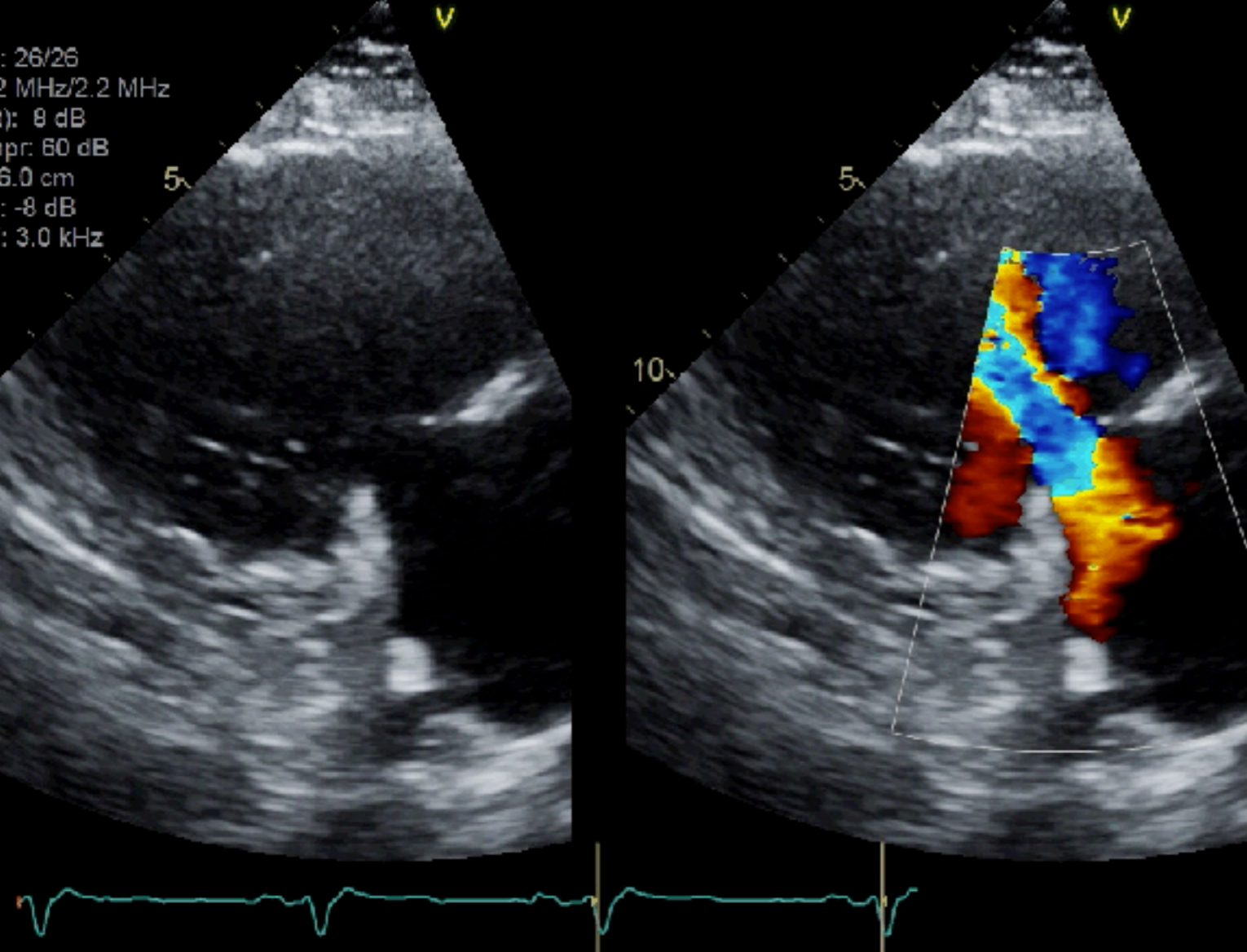
S: 51/  
.4 MHz/2.8 MHz  
(t): 8 dB  
mpr: 60 dB  
17.0 cm

63  
HR

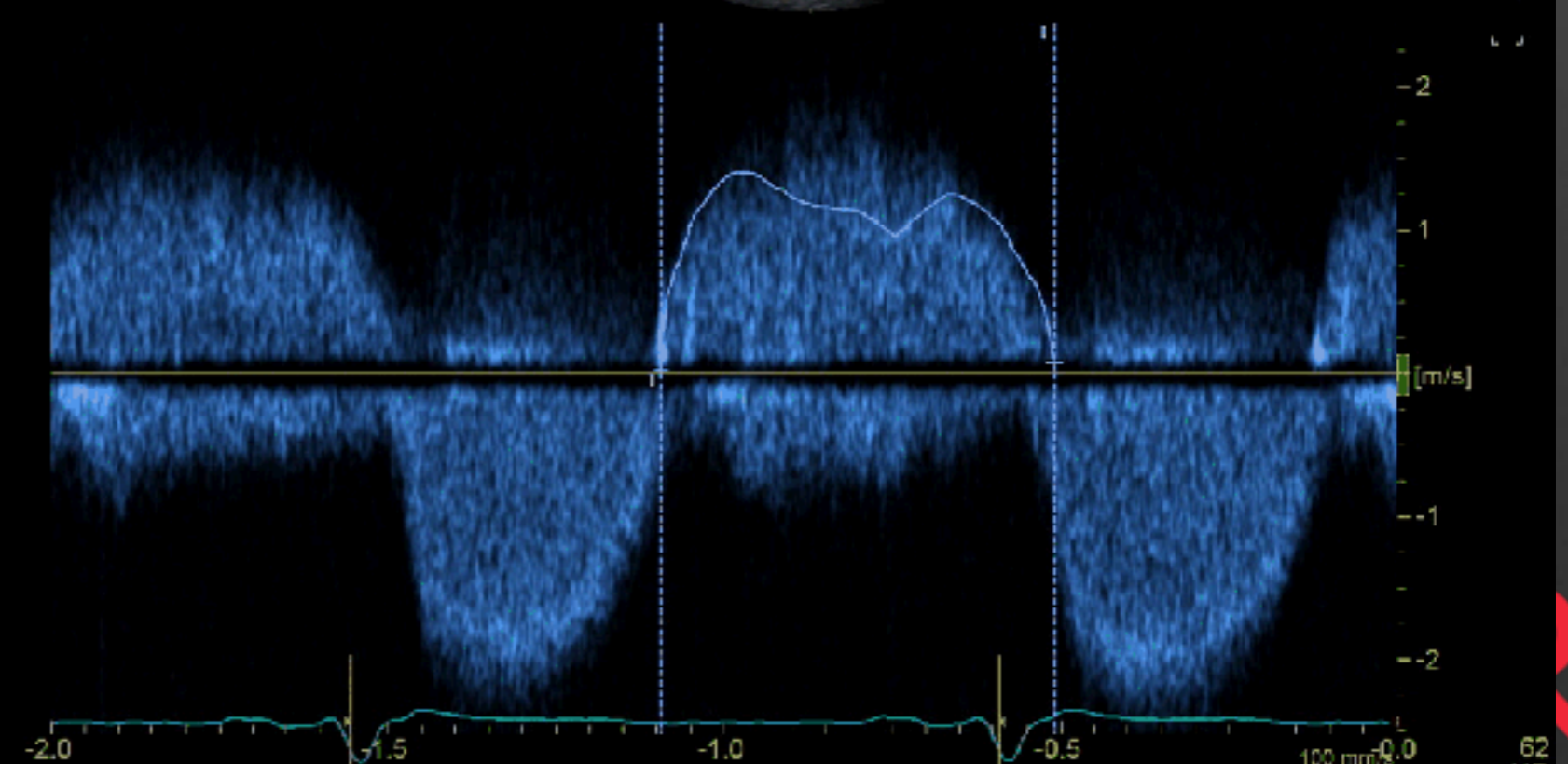
D: 20.0 cm





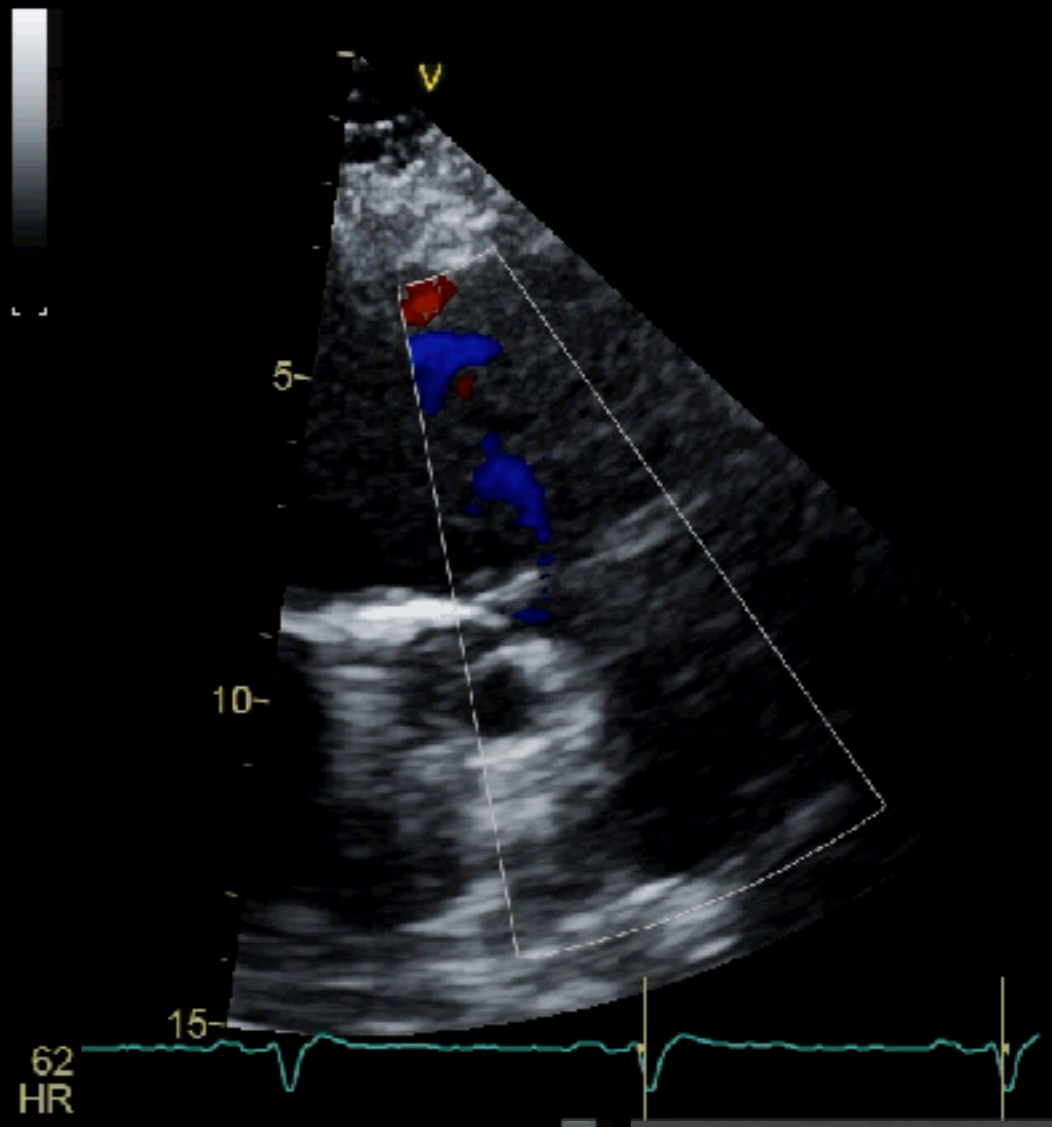
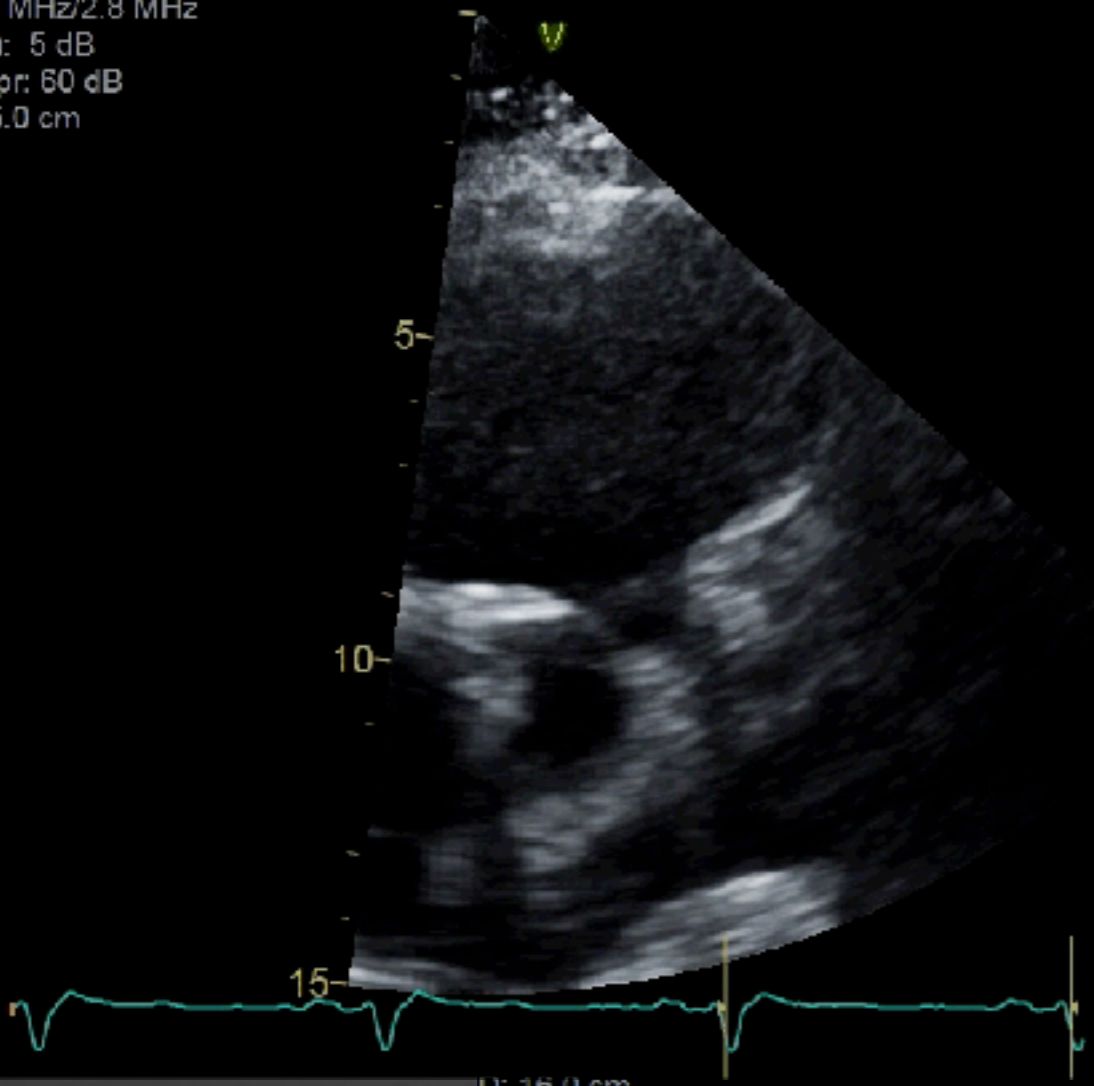


TV Vmean	1.08 m/s
TV maxPG	7.99 mmHg
TV meanPG	5.04 mmHg
TV VTI	63.8 cm
HR	103 BPM

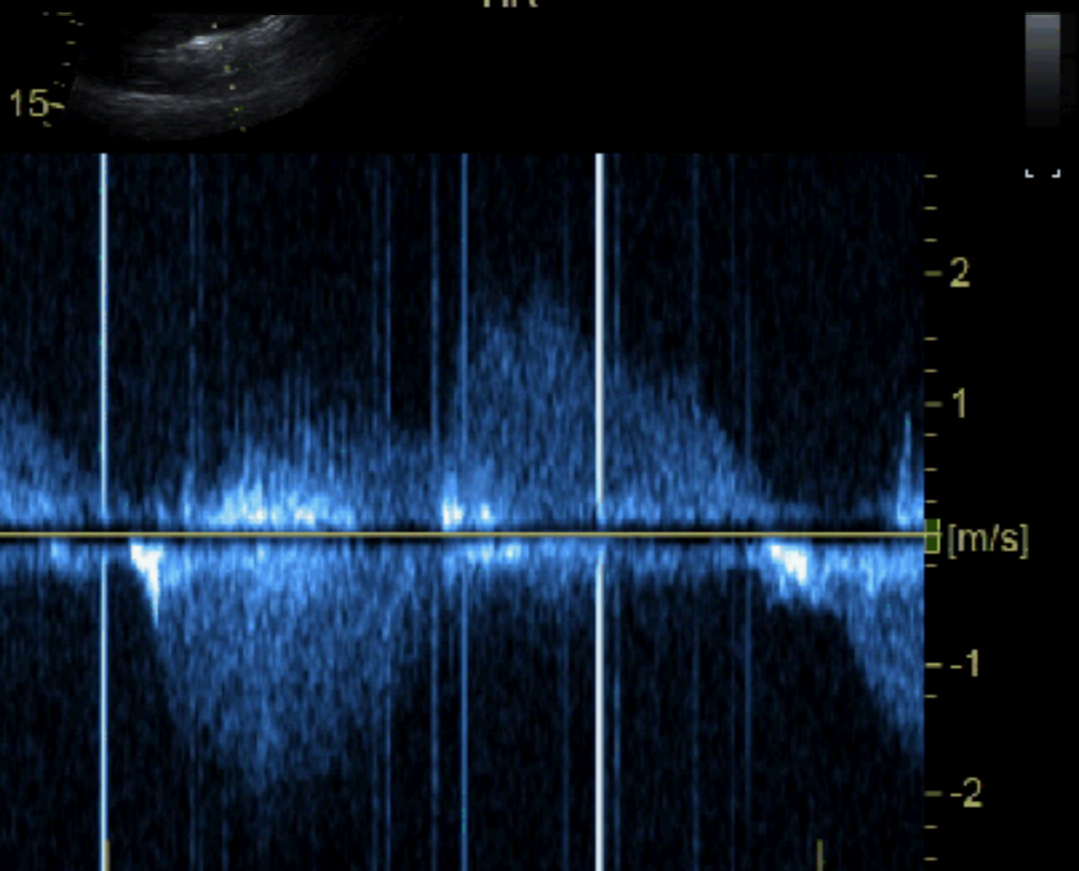


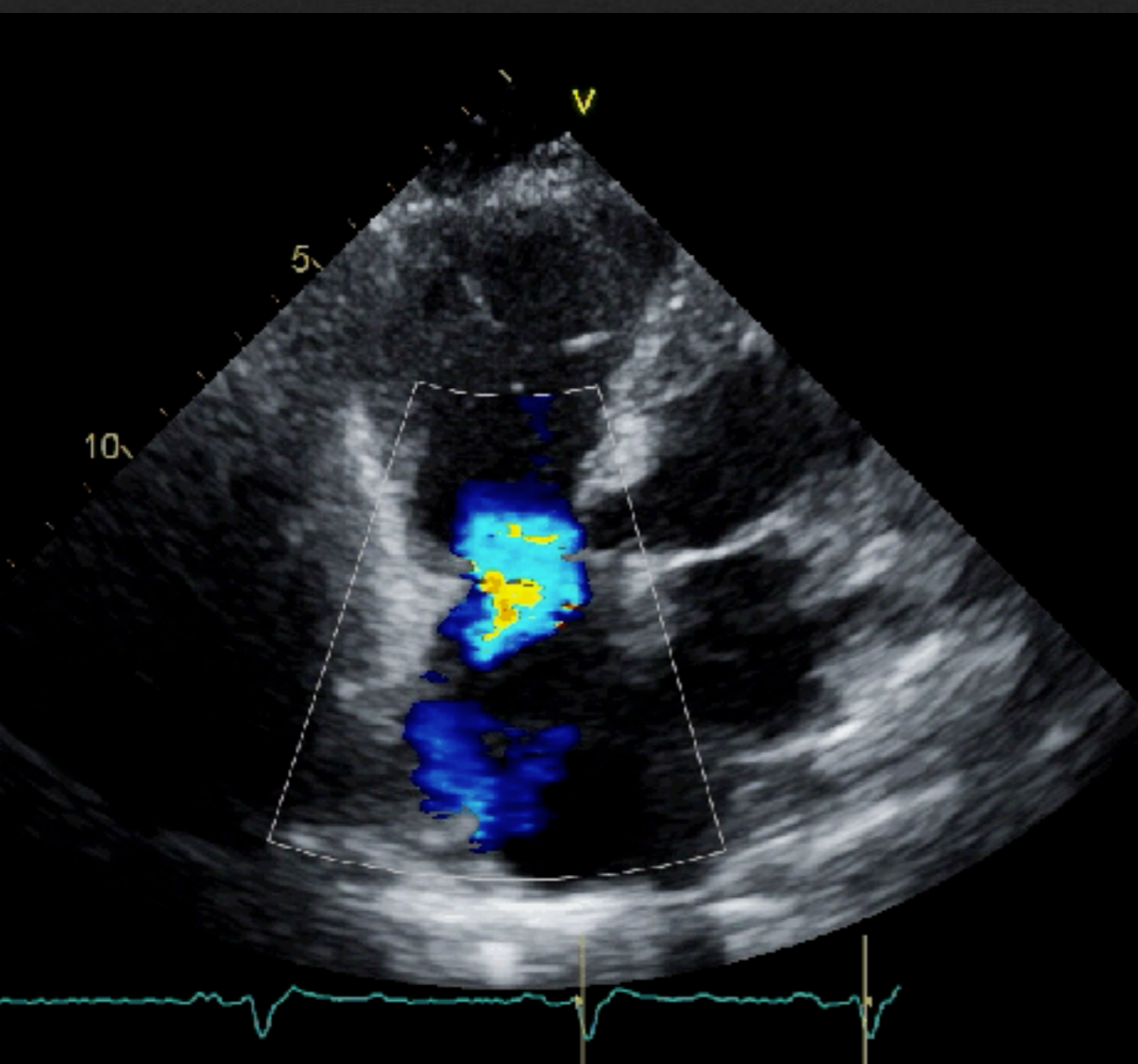
PS: 82  
1.4 MHz/2.8 MHz  
G(t): 5 dB  
Compr: 60 dB  
15.0 cm

Soft

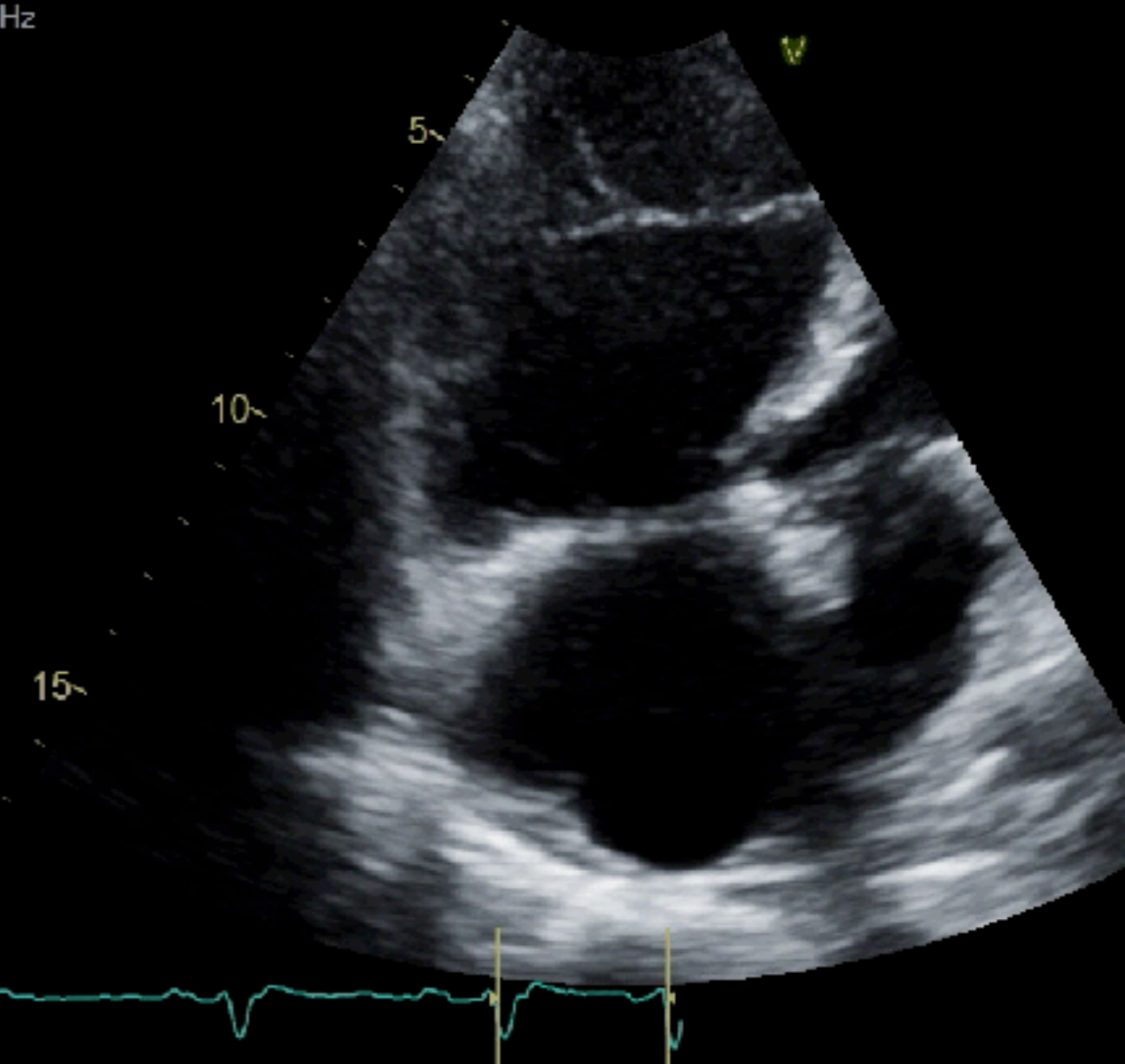


D: 16.0 cm  
G(d): 2 dB  
Scale: 590.7 cm/s





FPS: 51  
f: 1.4 MHz/2.8 MHz  
AG(t): 8 dB  
Compr: 60 dB  
D: 17.0 cm



## Imaging Assessment of Tricuspid Regurgitation Severity

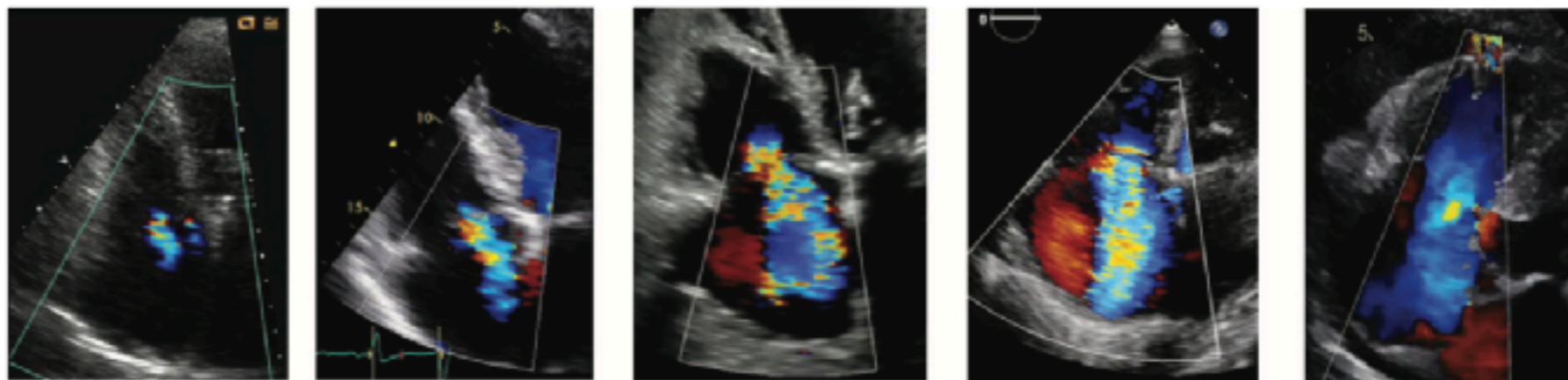


Rebecca T. Hahn, MD,<sup>a</sup> James D. Thomas, MD,<sup>b</sup> Omar K. Khalique, MD,<sup>a</sup> João L. Cavalcante, MD,<sup>c</sup>  
 Fabien Praz, MD,<sup>a,d</sup> William A. Zoghbi, MD<sup>e</sup>

**FIGURE 9** Proposed New Grading Scheme

Parameters	MILD	MODERATE	SEVERE	MASSIVE	TORRENTIAL
Vena Contracta width (biplane average)	<3 mm	3-6.9 mm	7 mm - 13 mm	14-20 mm	≥21 mm
EROA by PISA	<20 mm <sup>2</sup>	20-39 mm <sup>2</sup>	40-59 mm <sup>2</sup>	60-79 mm <sup>2</sup>	≥80 mm <sup>2</sup>
3D Vena Contracta Area or Quantitative Doppler EROA	-	-	75-94 mm <sup>2</sup>	95-114 mm <sup>2</sup>	≥115 mm <sup>2</sup>

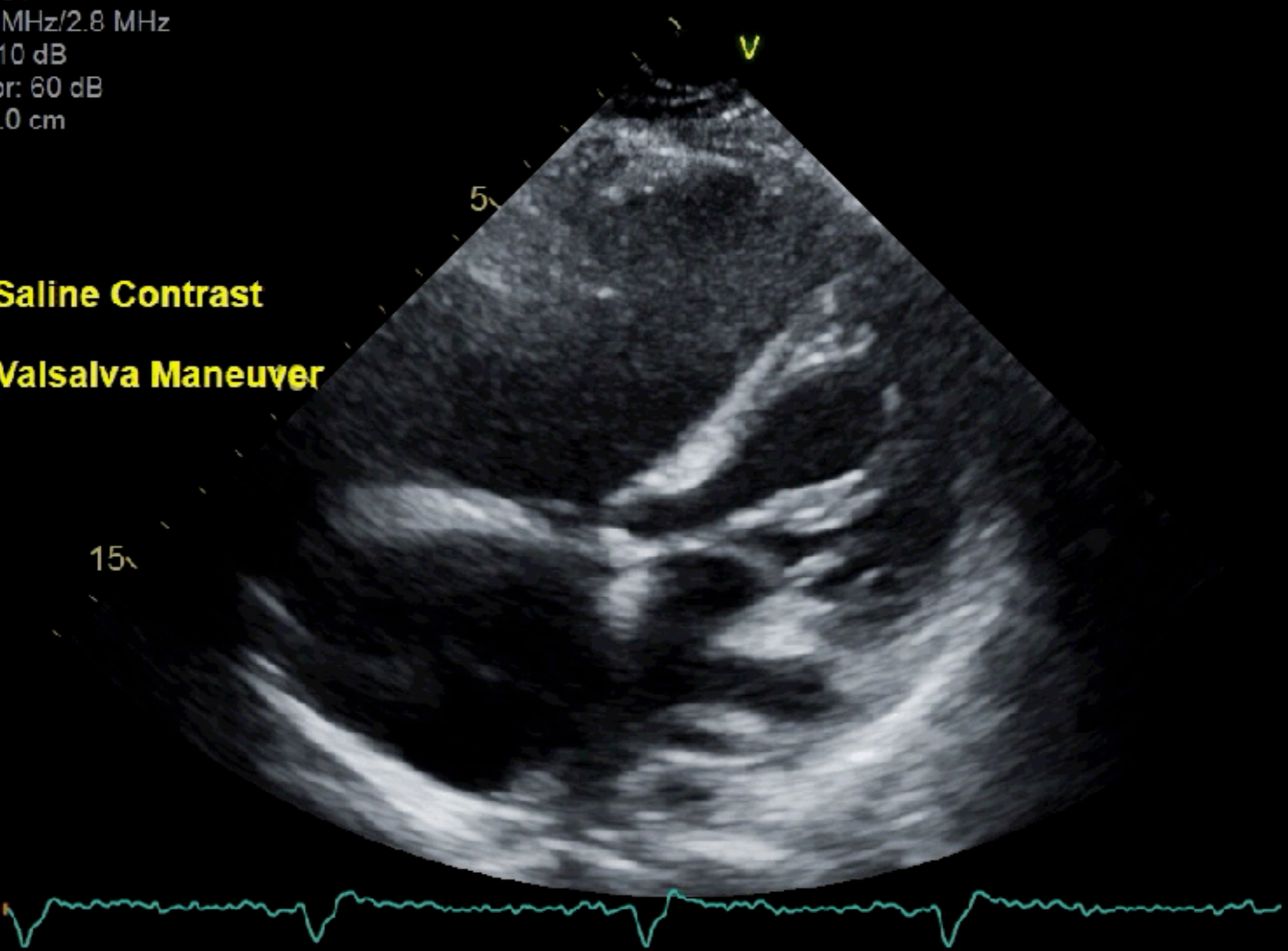
Example:



FD  
FPS: 51  
f: 1.4 MHz/2.8 MHz  
G(t): 10 dB  
Compr: 60 dB  
D: 17.0 cm



**Saline Contrast**  
**Valsalva Maneuver**



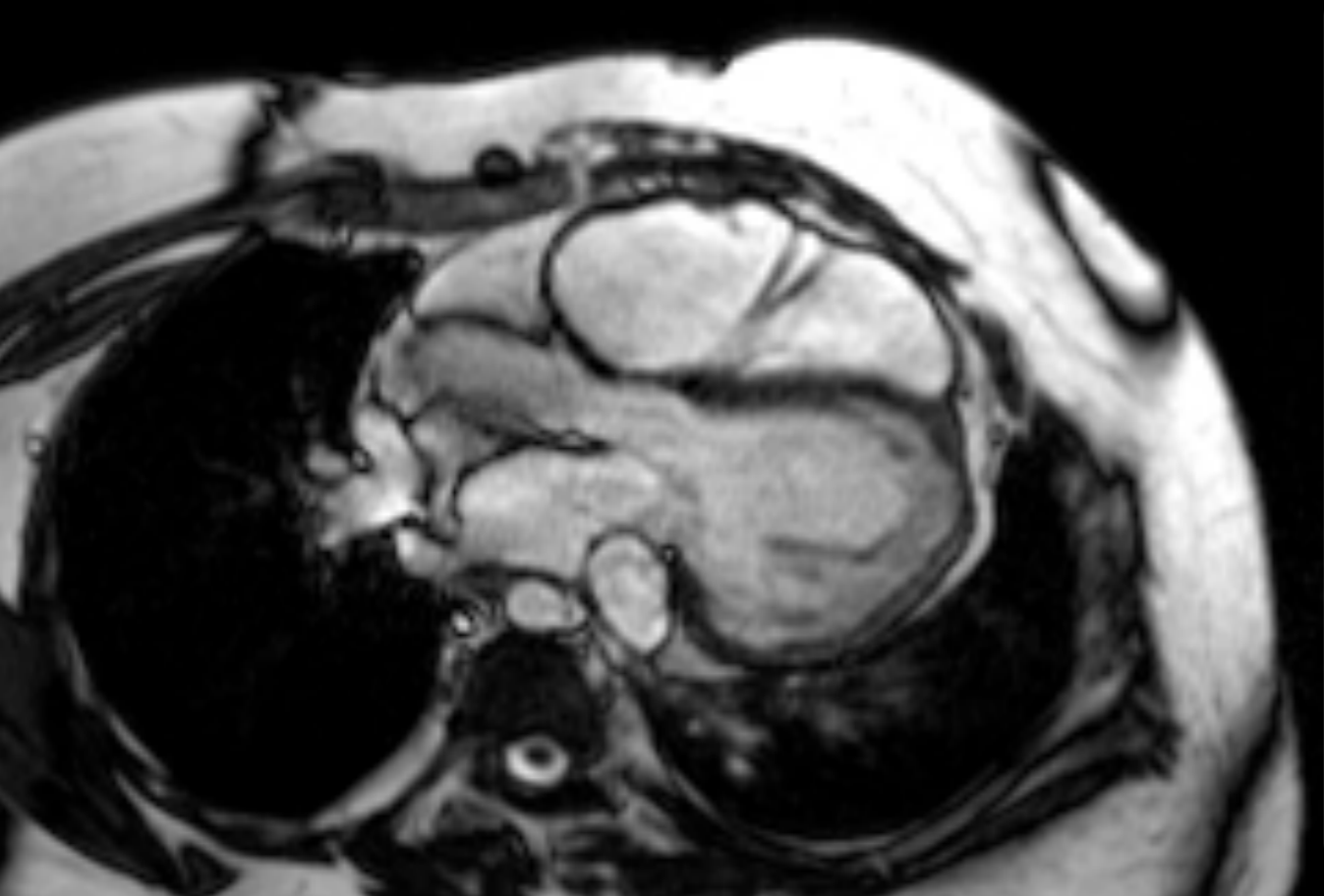
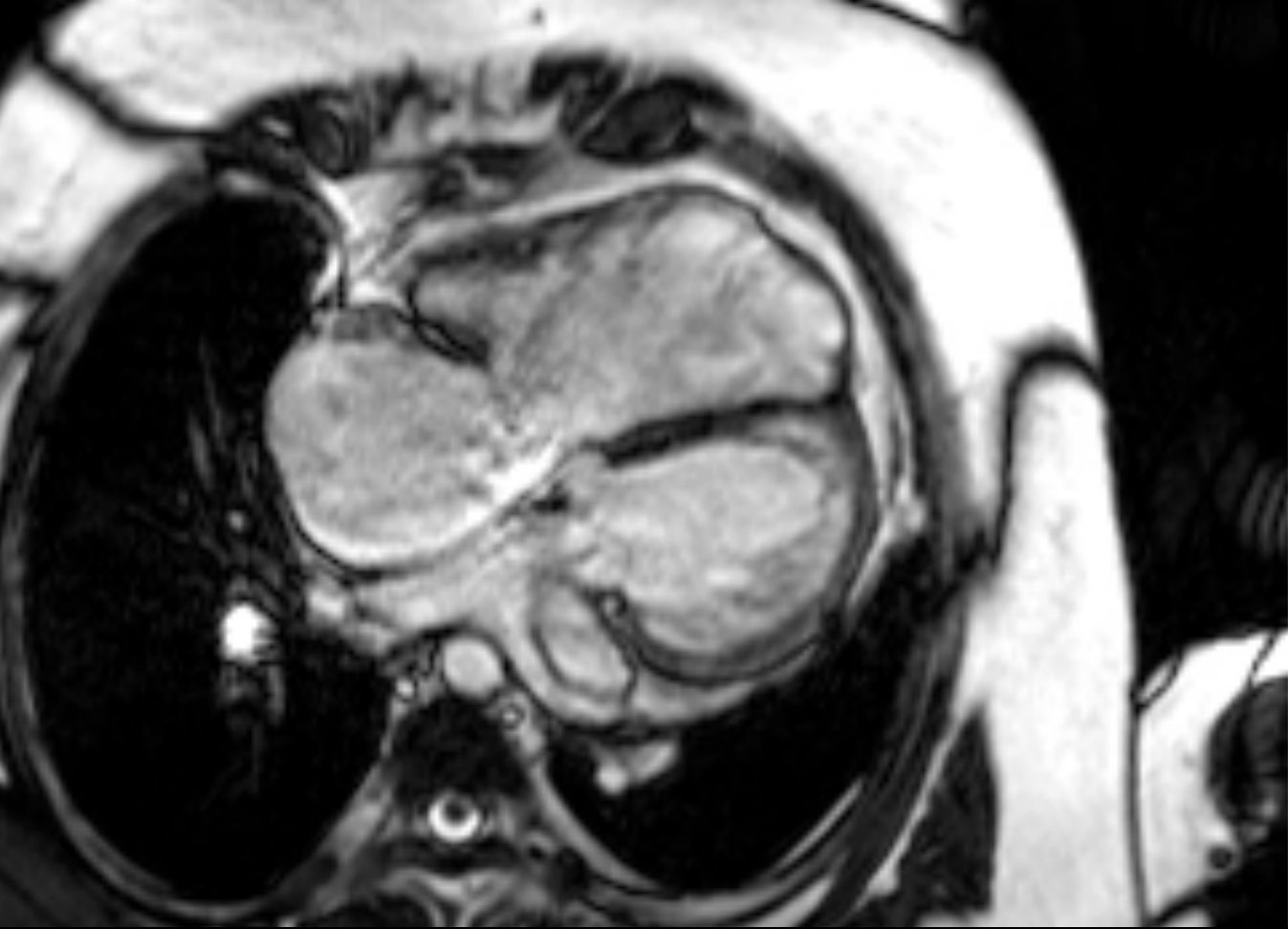
63  
HR

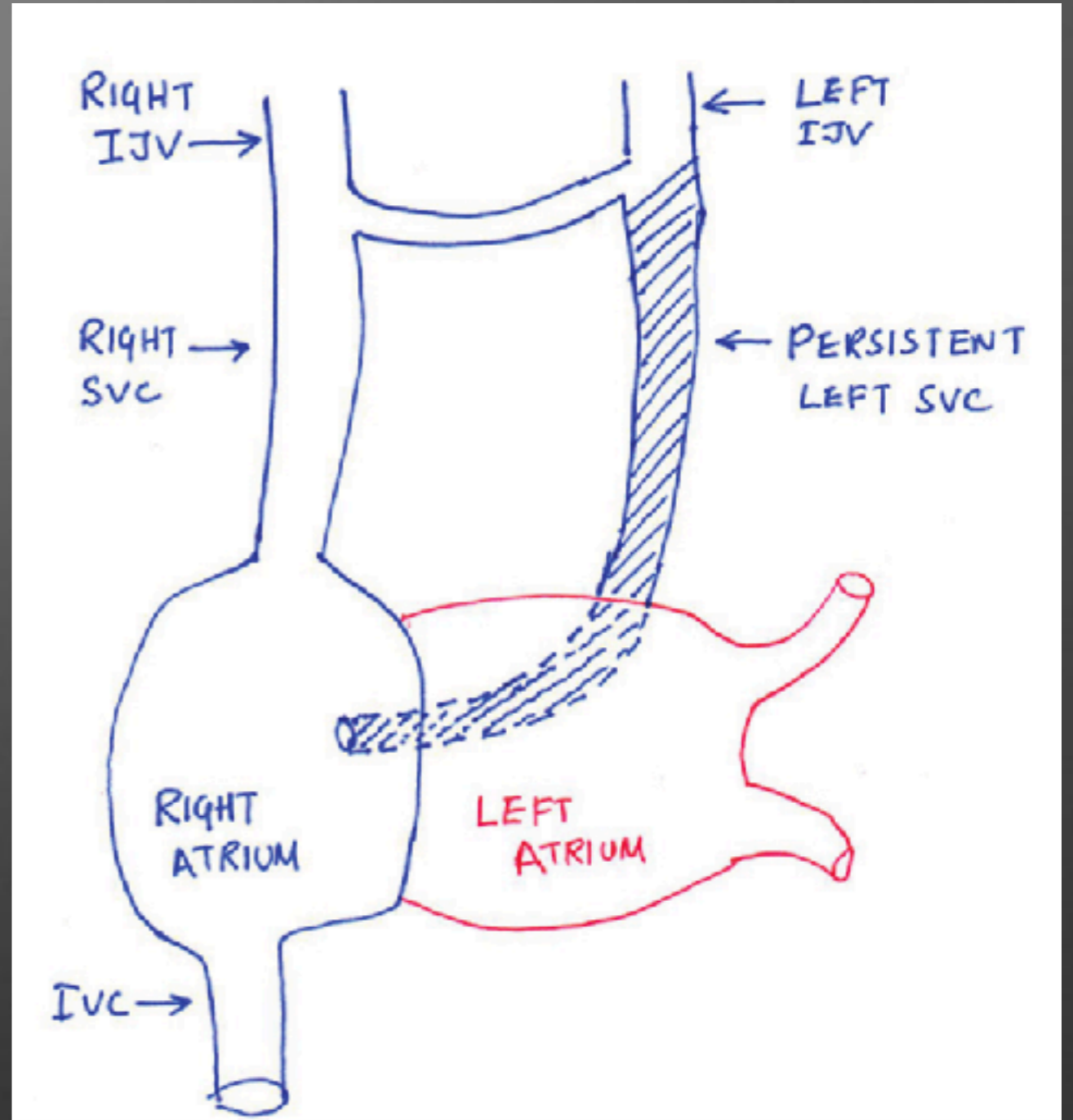


# Echo (Right heart findings)

- Dilated IVC with RAP > 15mmHg
- Severely dilated right heart with moderately reduced RV systolic function
- Torrential tricuspid regurgitation
- Mild pulmonary stenosis with moderate PI
- Very positive agitated saline injection









# MRI

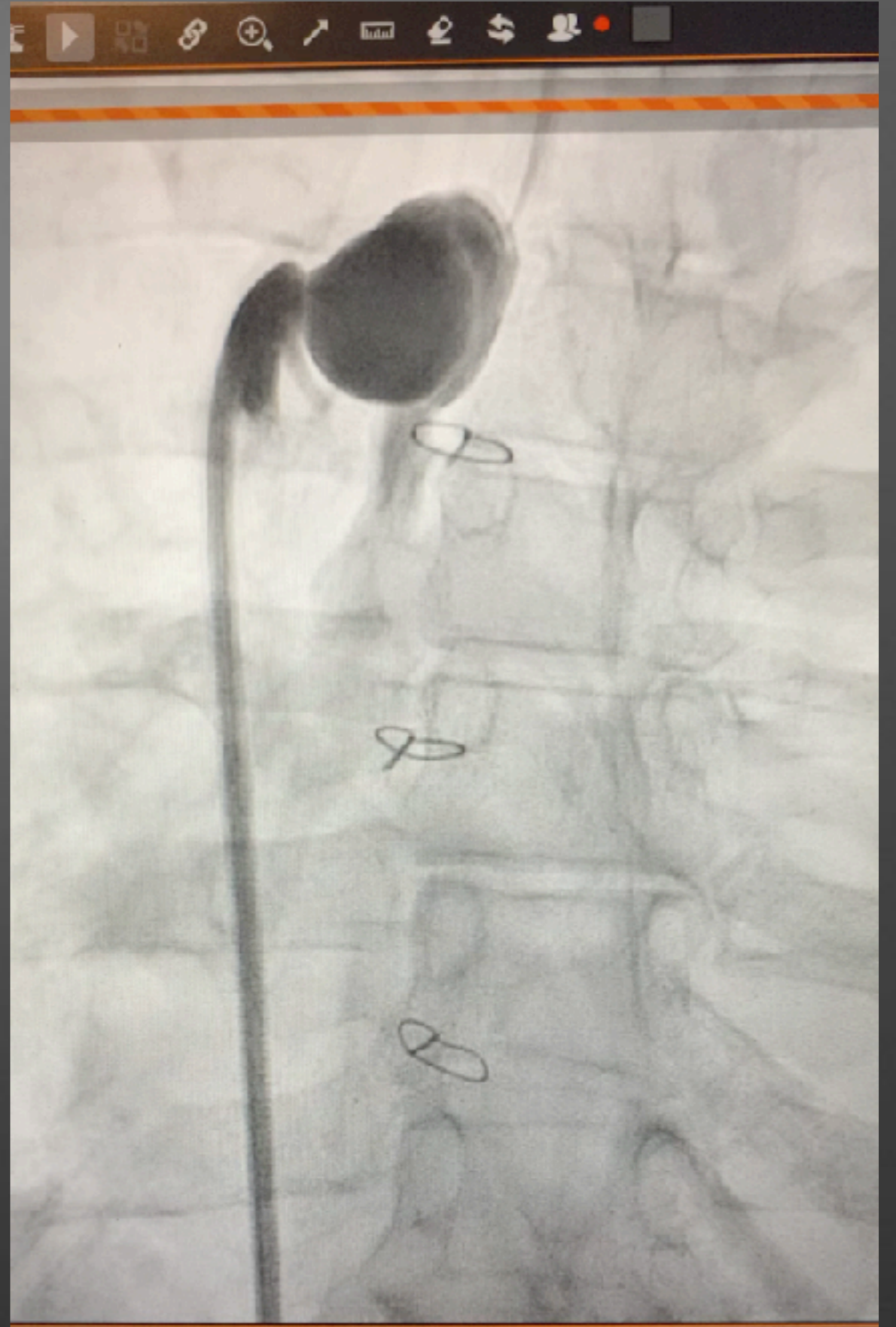
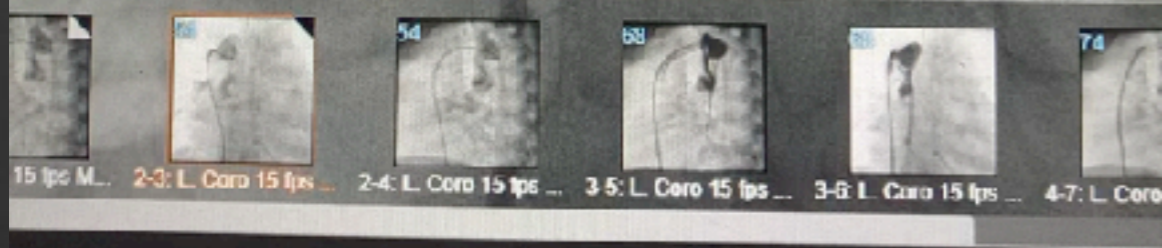
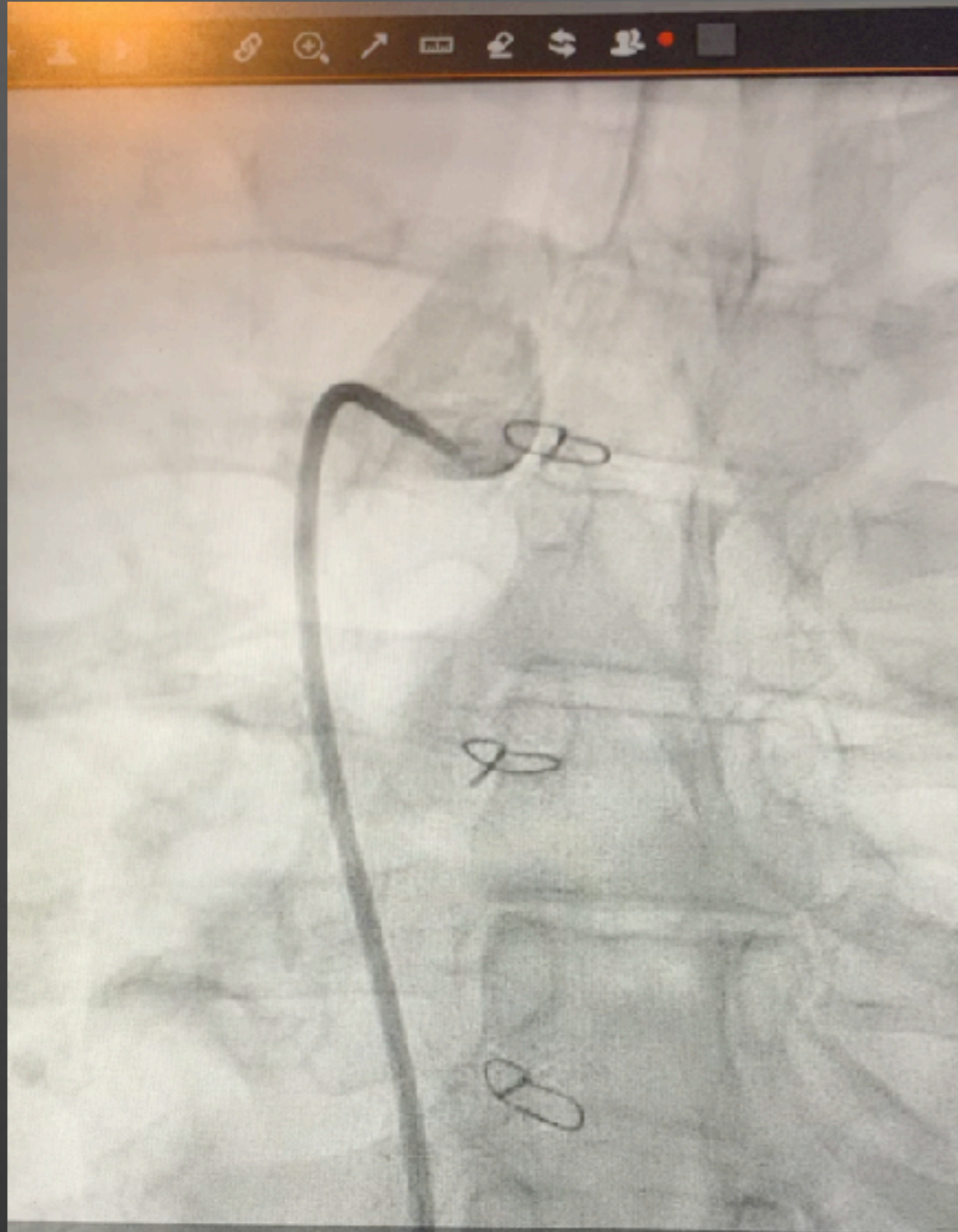
- Bilateral SVC with small branching vein
- Collaterals noted from the right SVC
- Increased RV volumes with RVEF of 47%
- Severe enlarged RA
- Tricuspid annular ring with severe TR (regurgitation volume of 59cc and fraction of 41%)
- Pulmonary regurgitation mild to moderate
- LVEF of 48%
- Qp:Qs not accurate



# CPET

- Peak  $\text{VO}_2$  17 mL/kg/min (RER 1.2)
- Baseline saturation of 98% which decreased to 90%
- Normal BP response





# Systemic Venous - Pulmonary Venous Collaterals

- Occur with elevated systemic venous pressures or systemic venous obstruction
  - DTGA with Mustard/Senning (atrial switch) obstruction
  - Single ventricles - Obstruction along total caval pulmonary shunt



# Indications for closure

- Collateral > 3mm
- Hypoxemia
- SVC syndrome
- Congestive hepatopathy/Ascites/LE edema

***When due to elevate pressures - treat underlying cause***



# Diagnosis and Plan

- Systemic venous to pulmonary venous collaterals plugged
- Surgery - Tricuspid valve replacement; Right atrial reduction; long discussion of pulmonary valve replacement

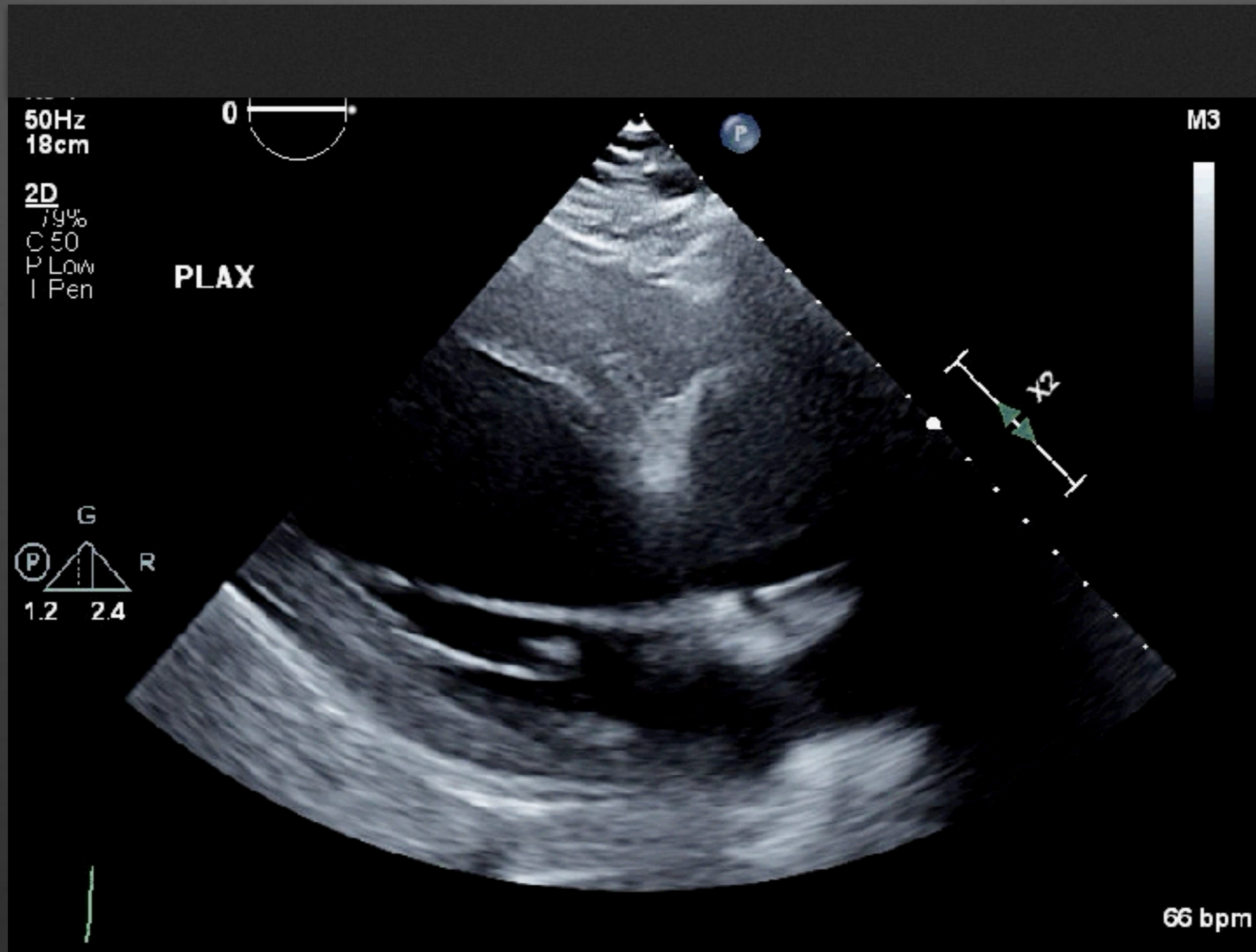


# Case 3

- 35 yo transferred to CTS for surgical evaluation of ascending aorta of 7 cm.
- Dyspnea on exertion started after his first COVID vaccine about 7 weeks prior
- Since his second vaccine orthopnea, LE edema, and dyspnea on exertion.
- Admission systolic BP 190/50's



# Echocardiogram Admission





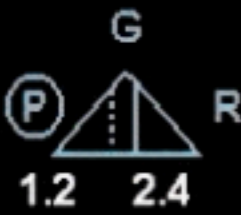
16Hz  
18cm



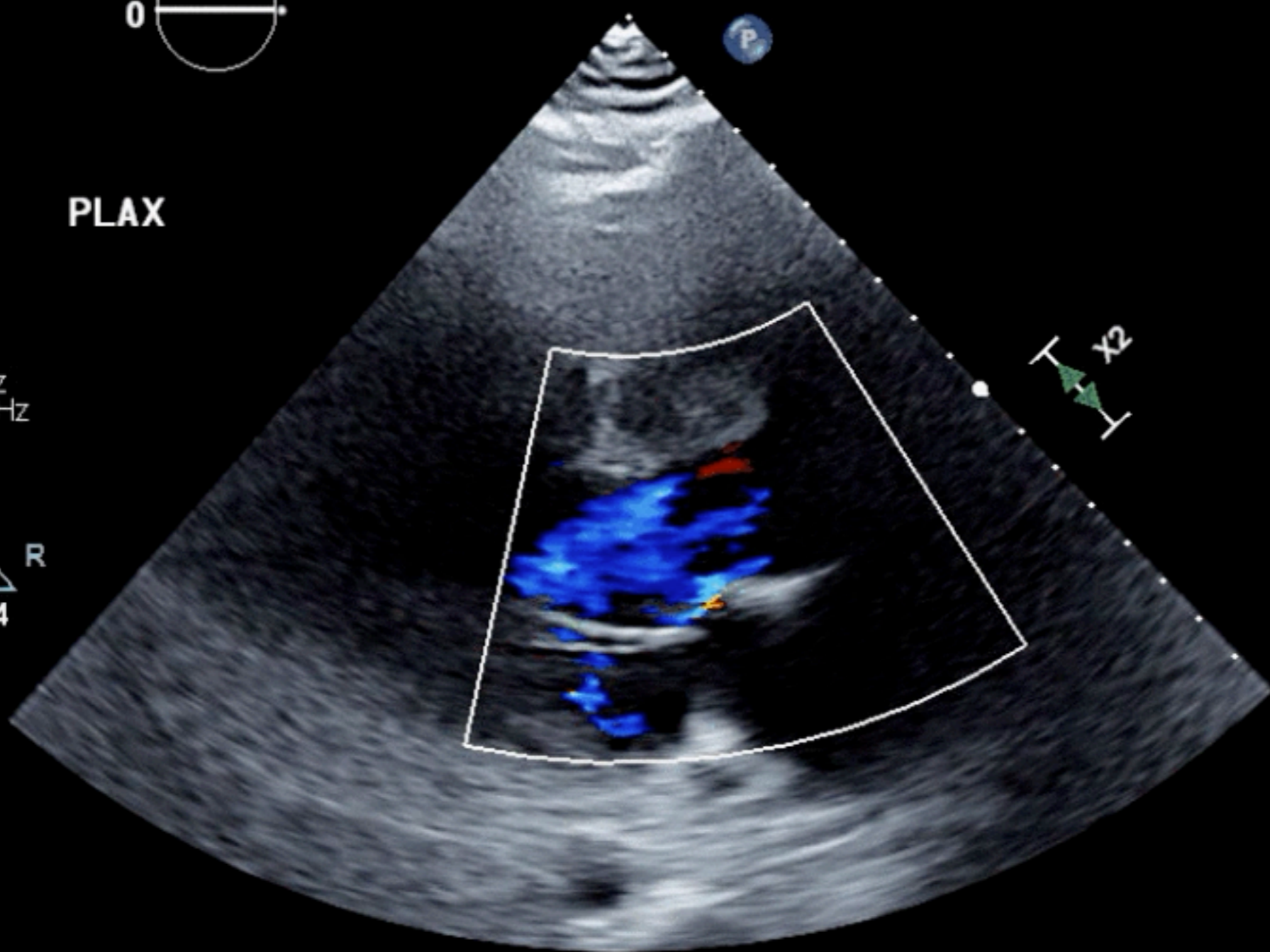
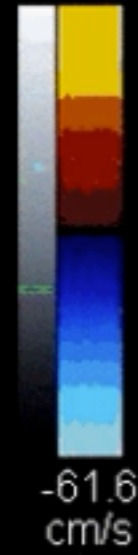
**2D**  
77%  
C 50  
P Low  
HPen

**PLAX**

**CF**  
50%  
4000Hz  
WF 399Hz  
2.5MHz



M3 M4  
+61.6



65 bpm



X5-1  
40Hz  
26cm



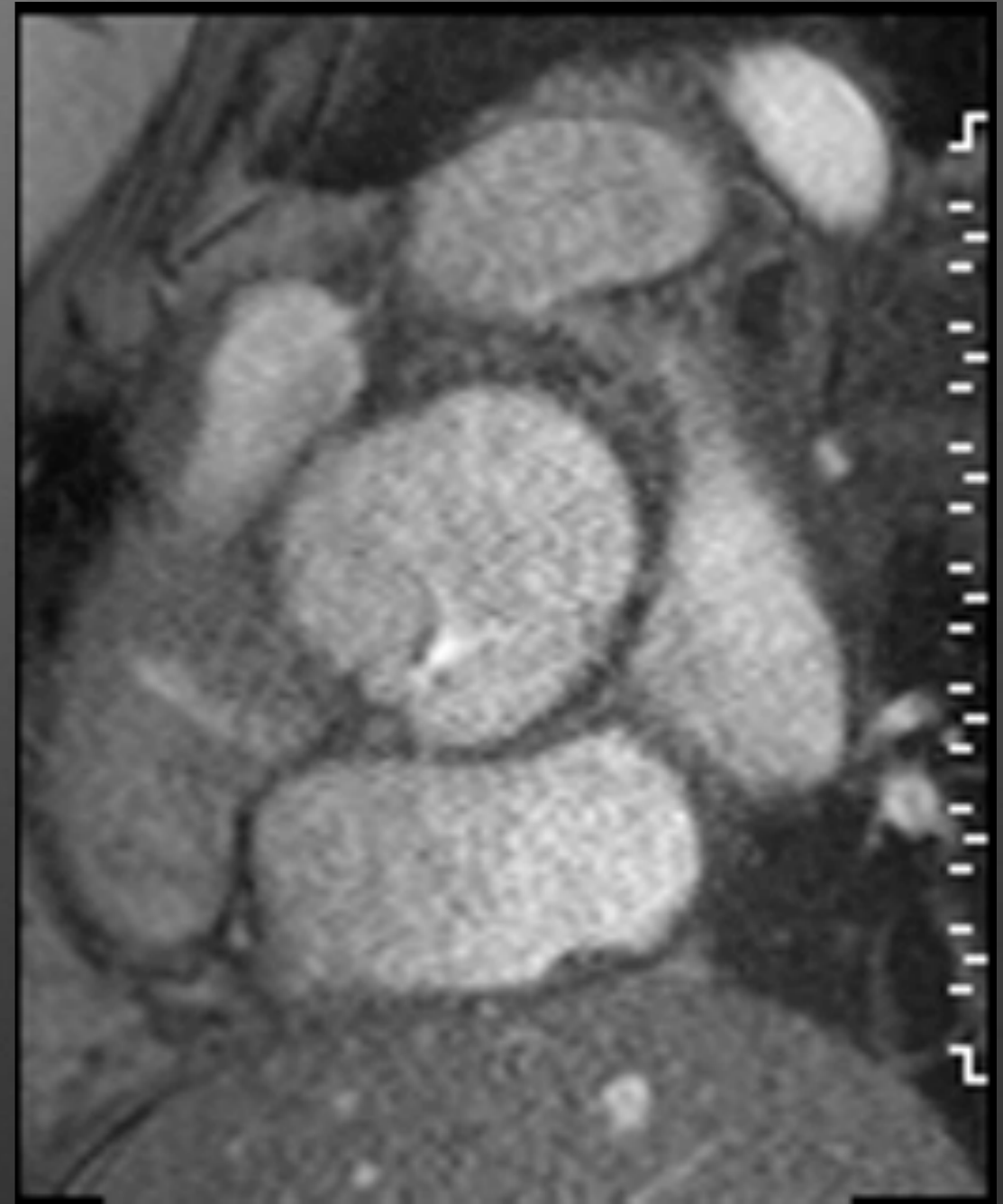
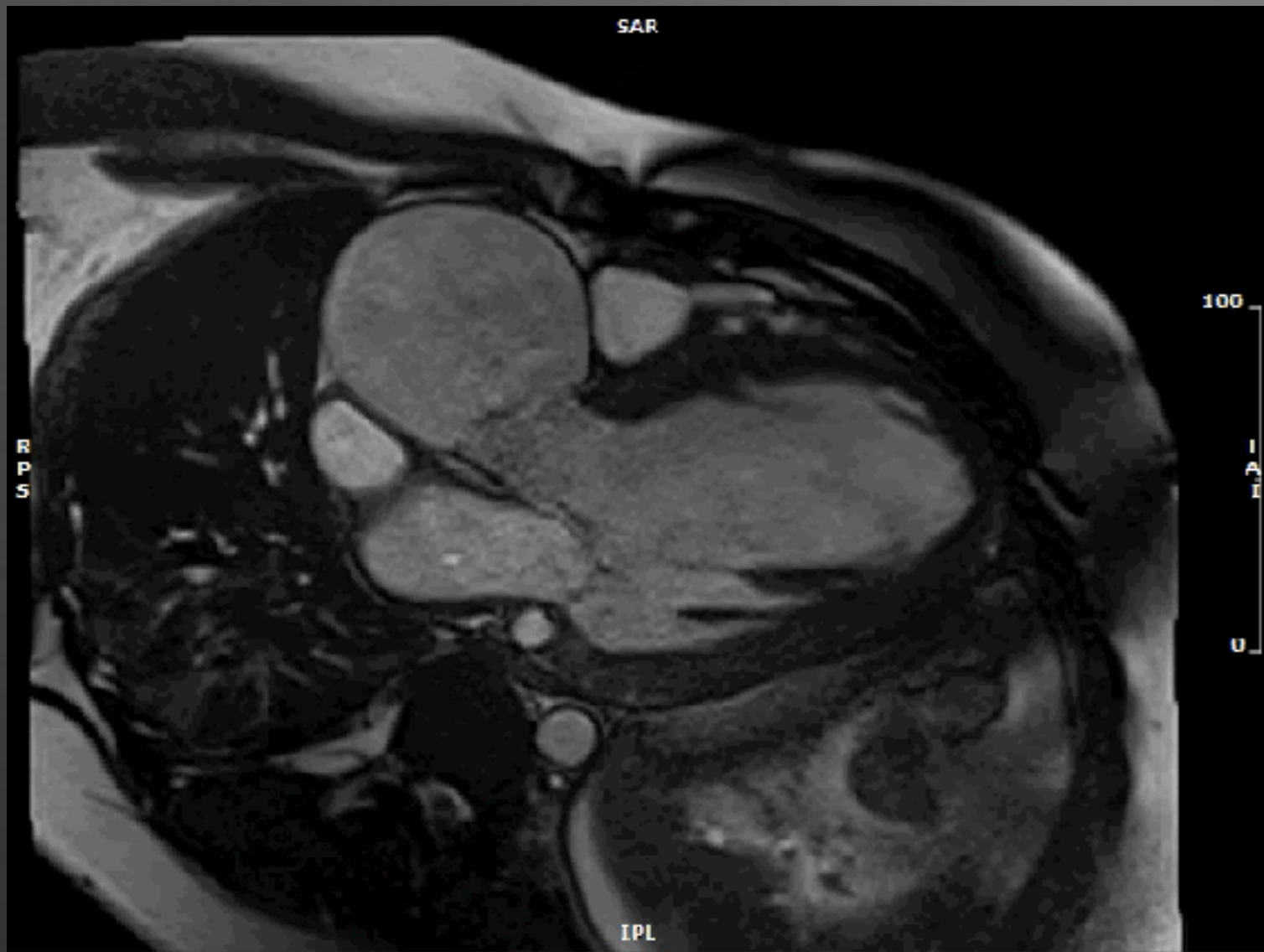
2D  
79%  
C 50  
P Low  
HPen

M3



65 bpm

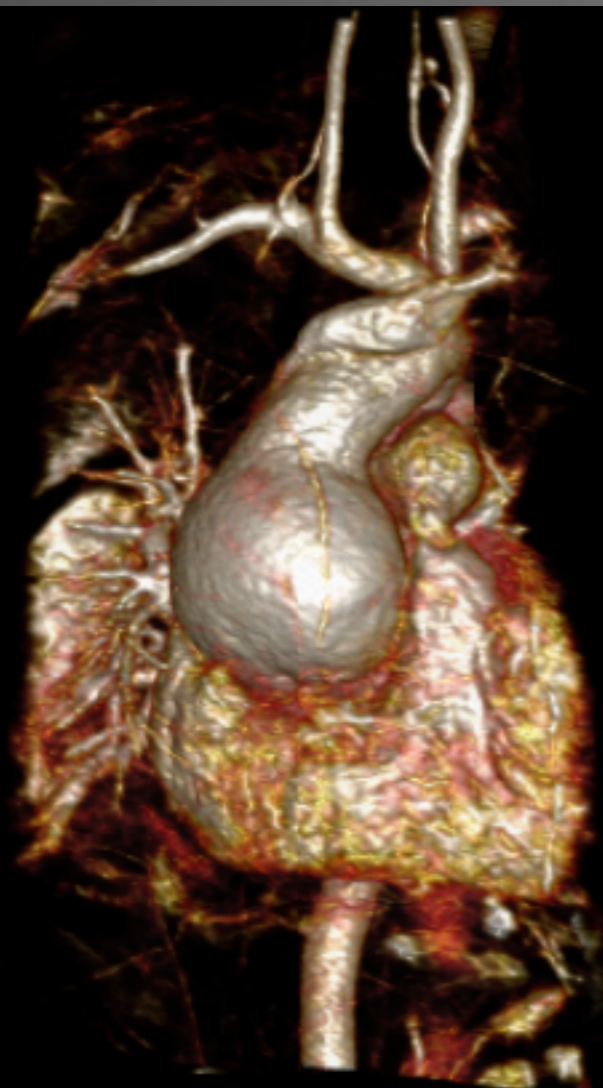




12  
10

my H

VCUHS GW 3



S  
A  
L  
I

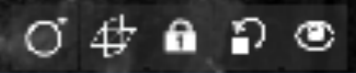
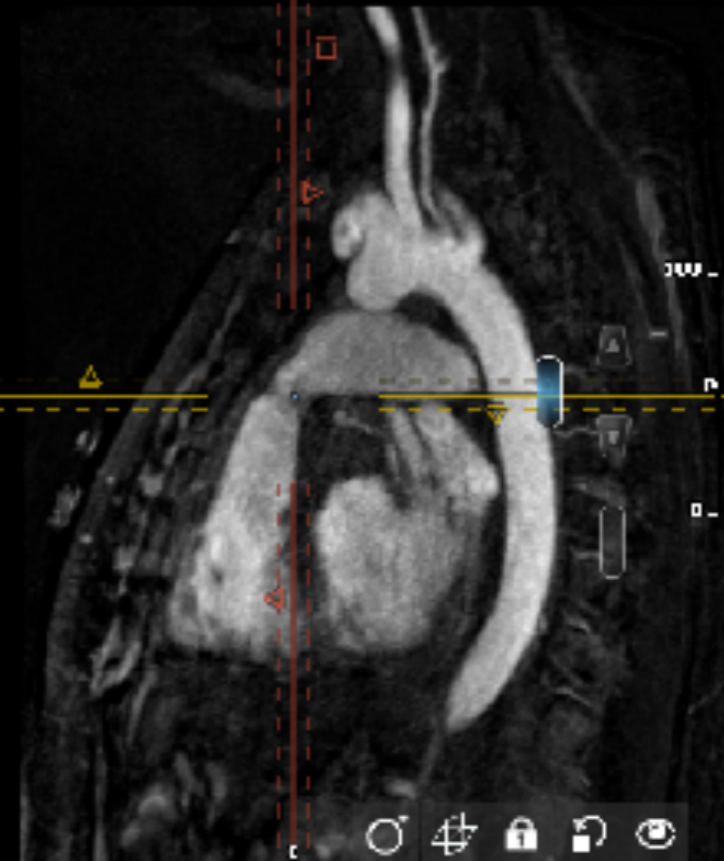
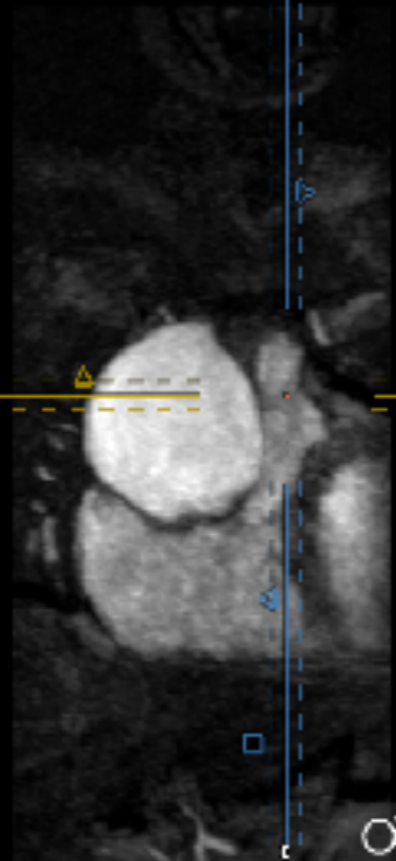
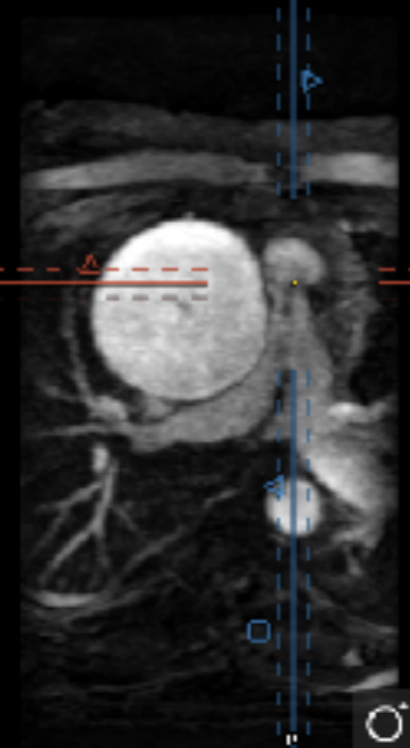
S  
A  
L  
I

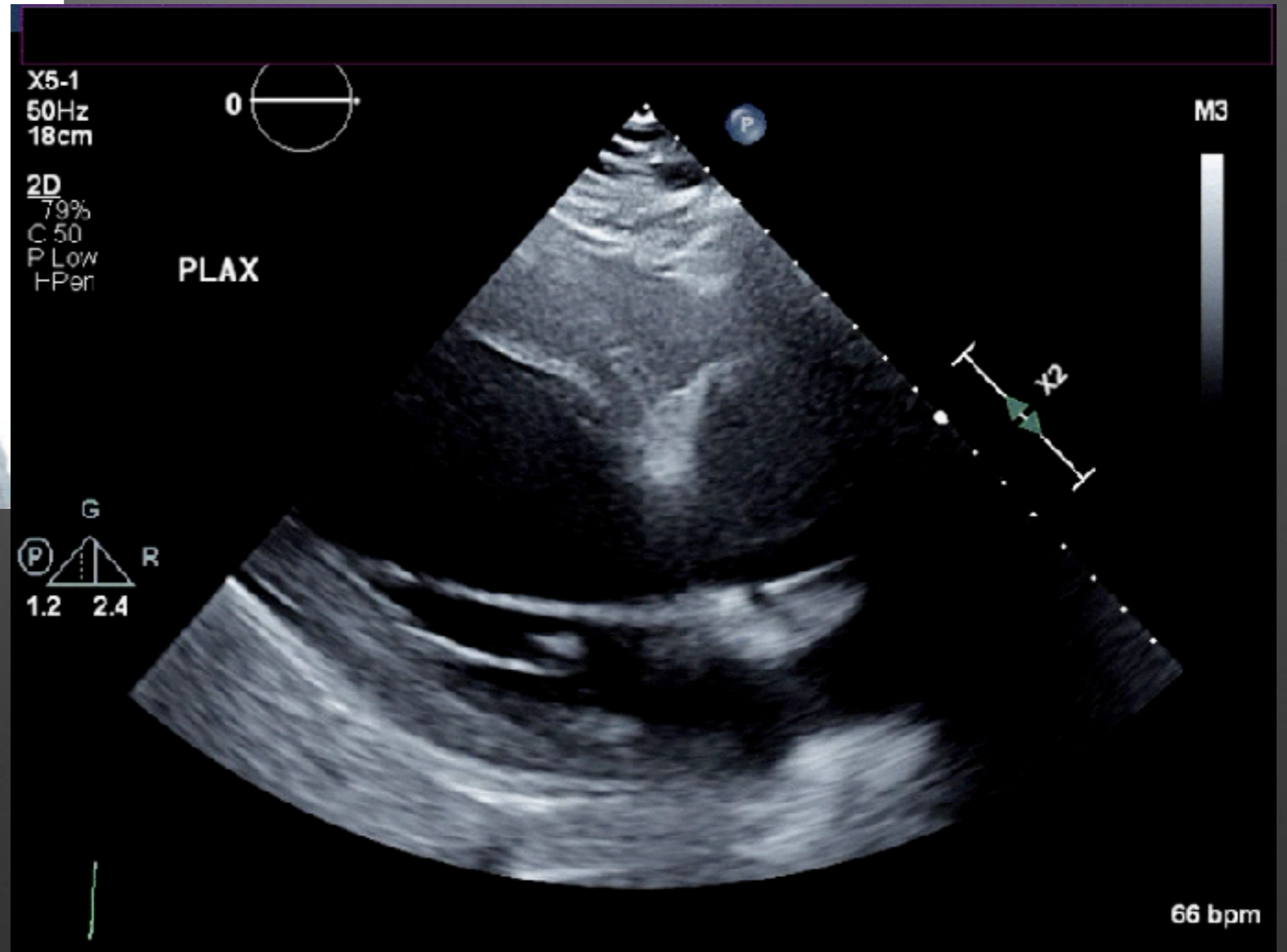


Juliusson Jimmy '91  
2121 11 20  
2217/1  
HR 0

RAU 12  
PRA 10

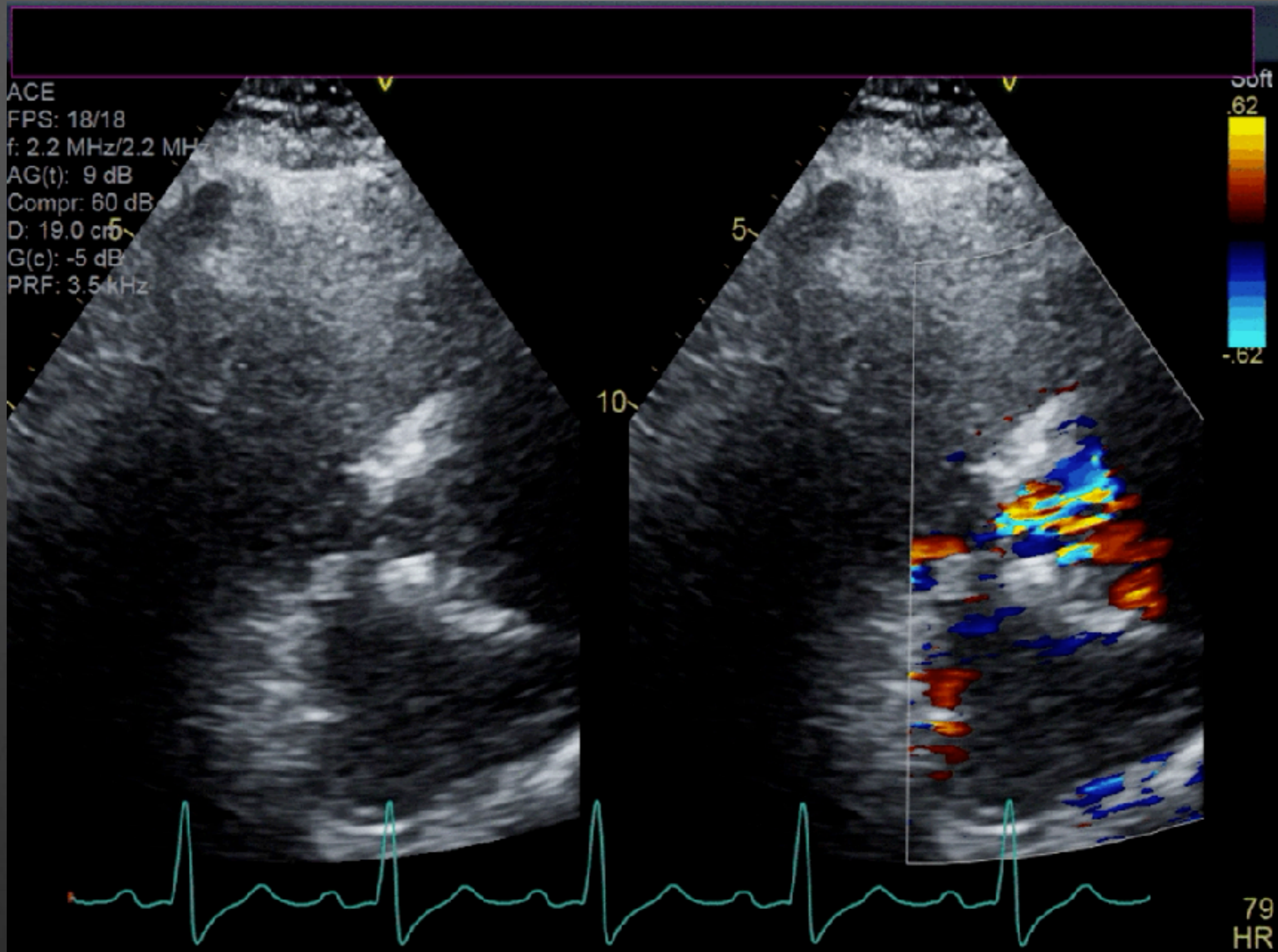
YLR 0.07 J





ACE  
FPS: 18/18  
f: 2.2 MHz/2.2 MHz  
AG(t): 9 dB  
Compr: 60 dB  
D: 19.0 cm  
G(c): -5 dB  
PRF: 3.5 kHz

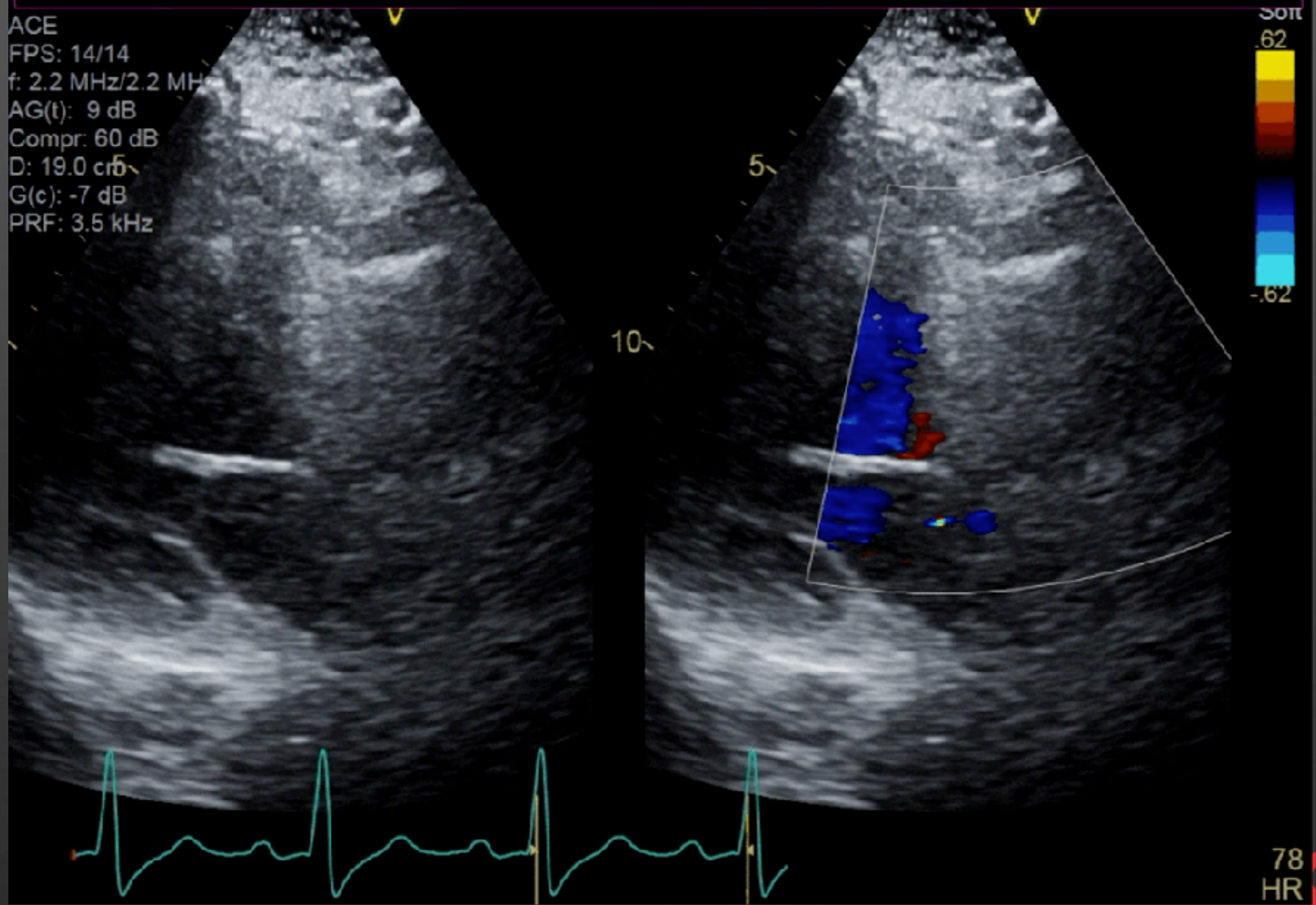
Soft  
.62  
-62



79  
HR

ACE  
FPS: 14/14  
f: 2.2 MHz/2.2 MHz  
AG(t): 9 dB  
Compr: 60 dB  
D: 19.0 cm  
G(c): -7 dB  
PRF: 3.5 kHz

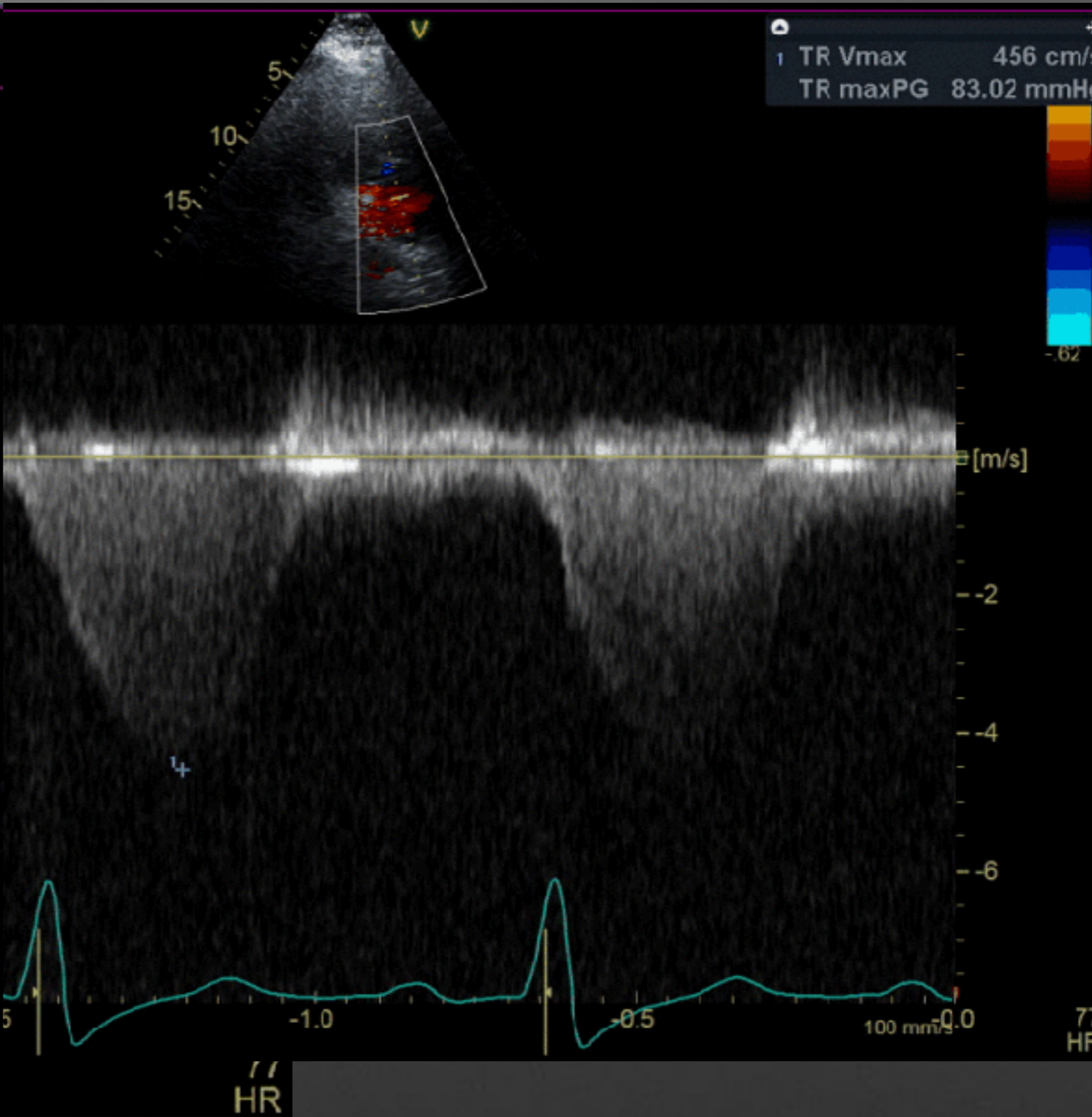
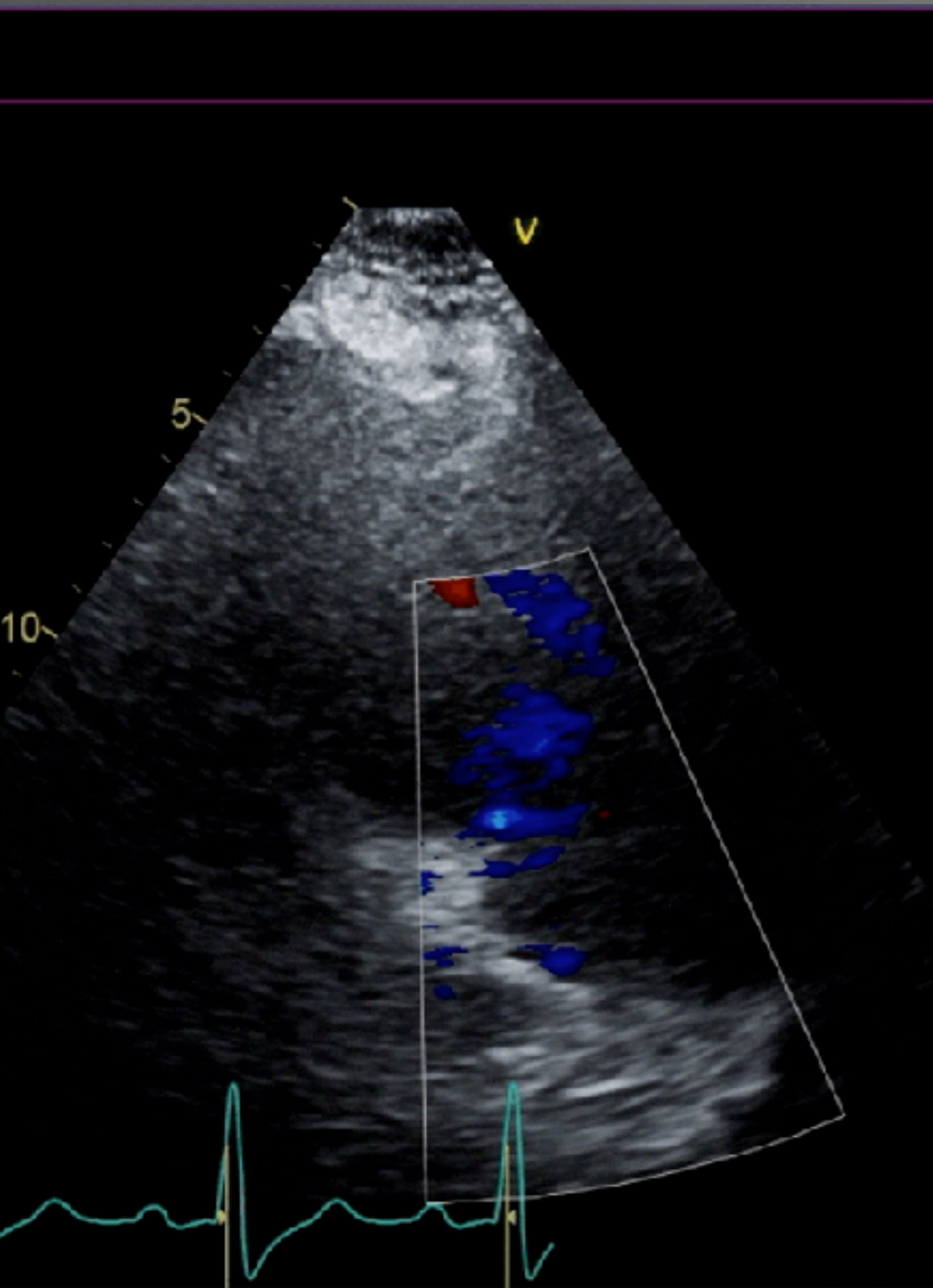
SOT  
.62  
-.62

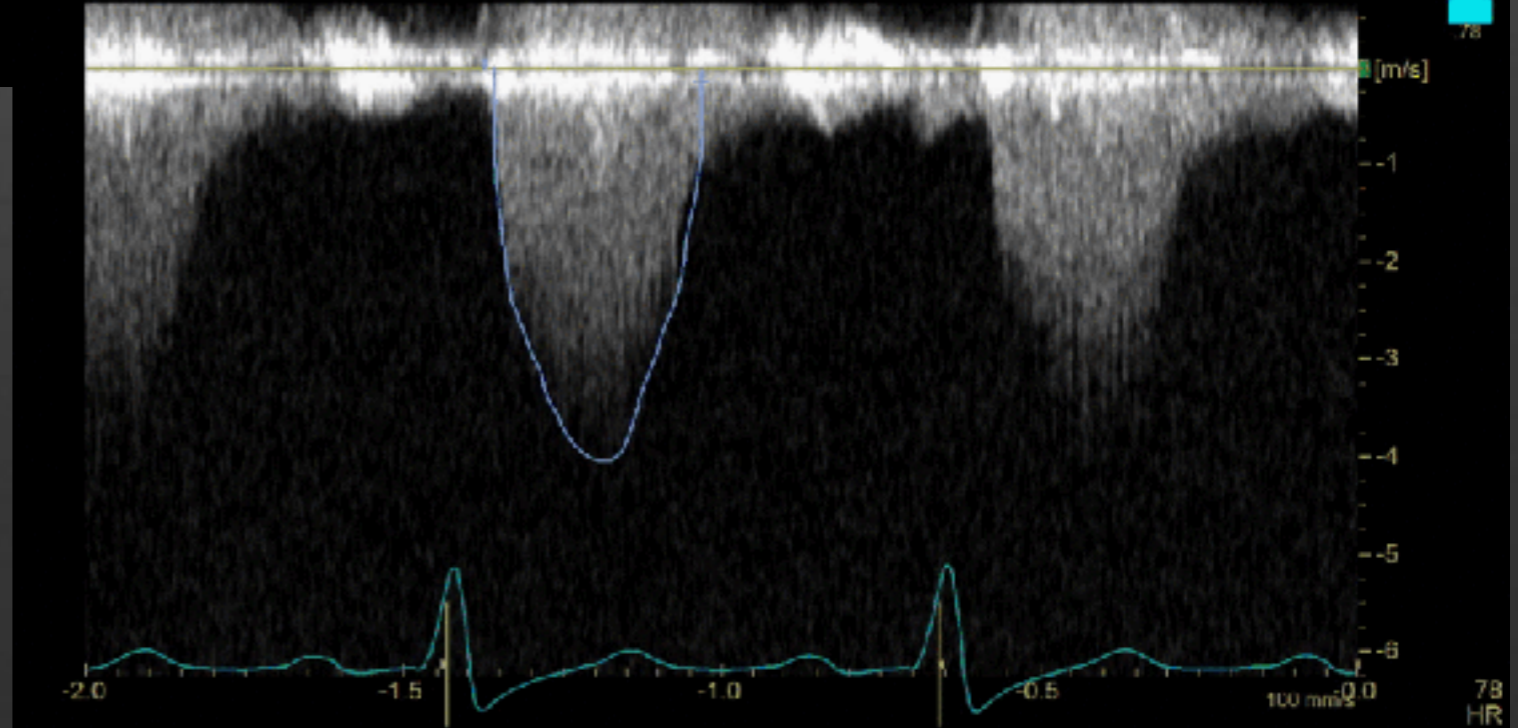
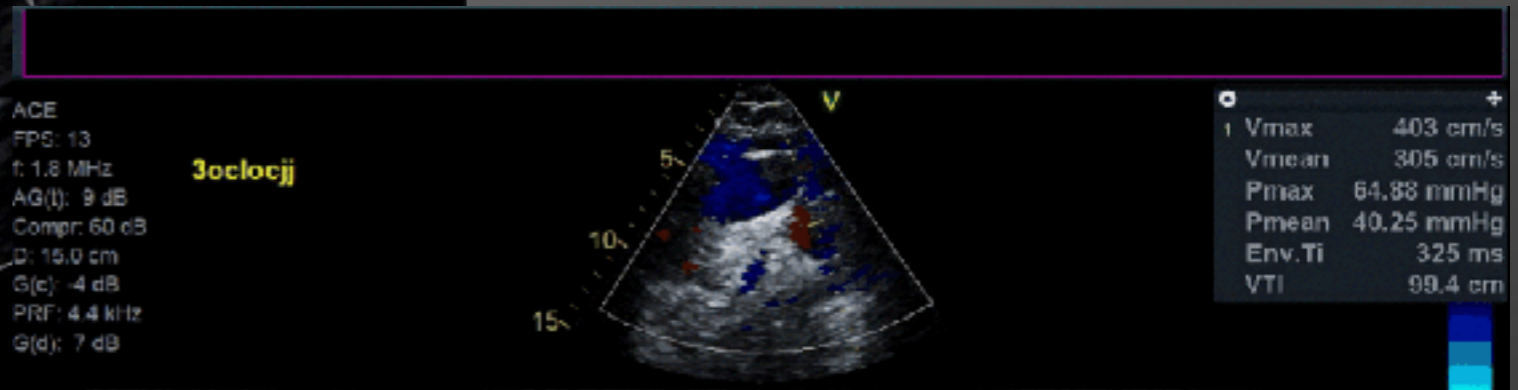
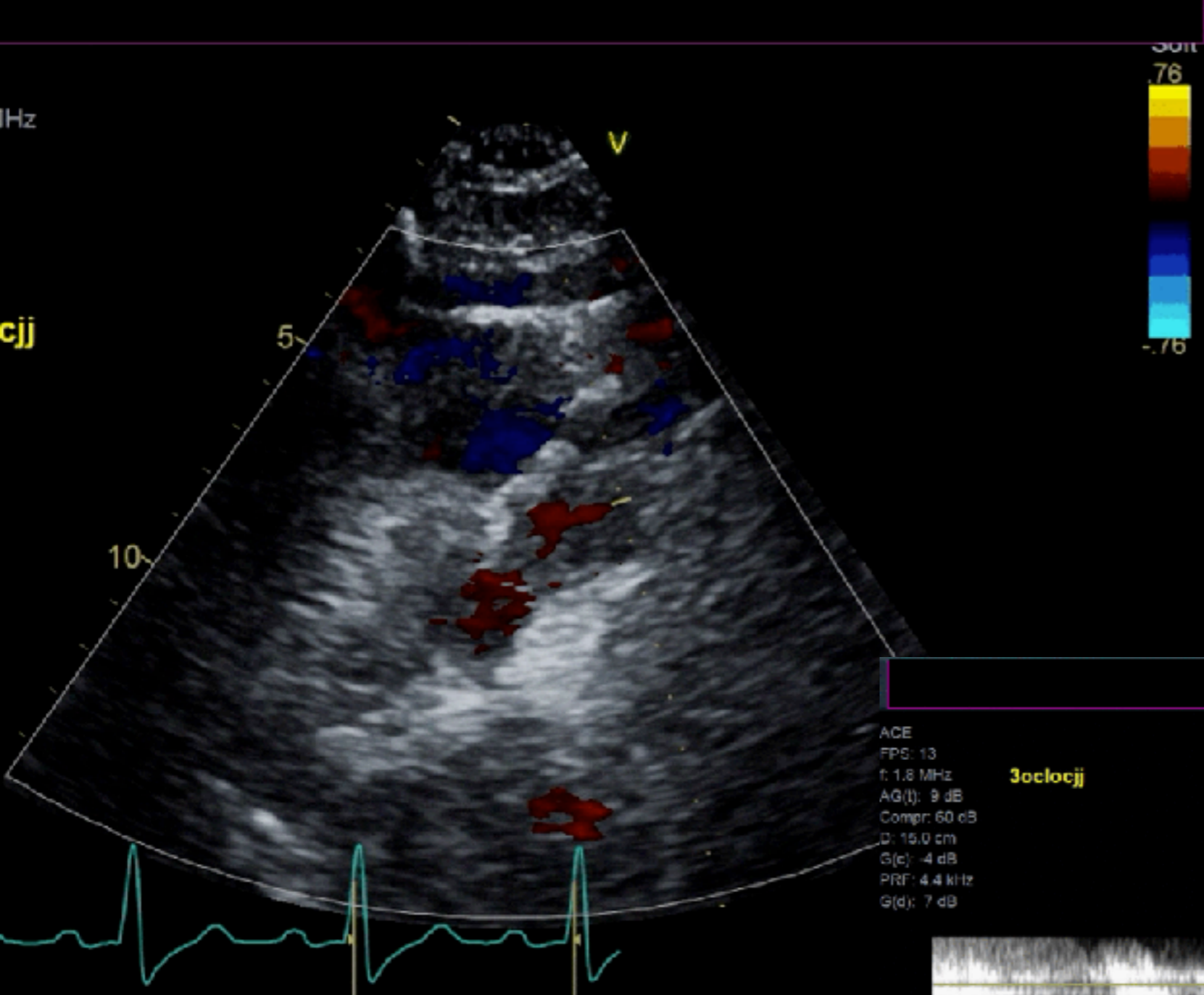


78  
HR

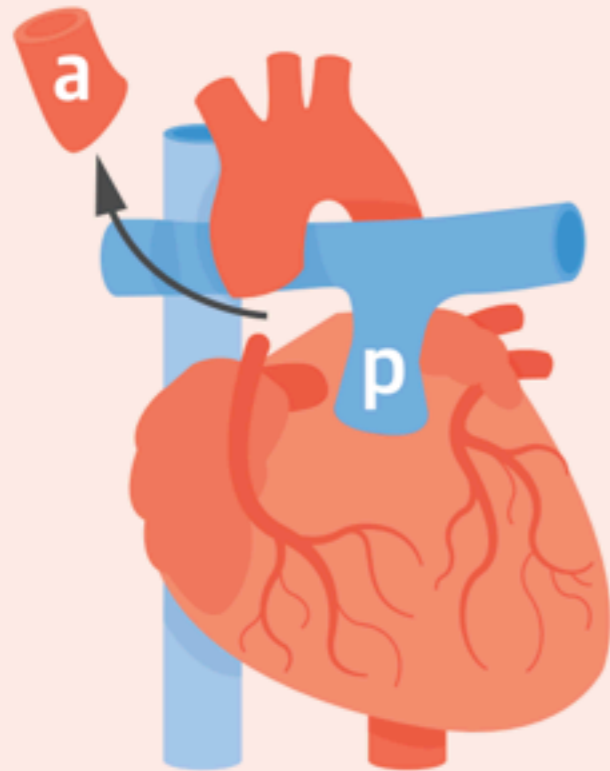




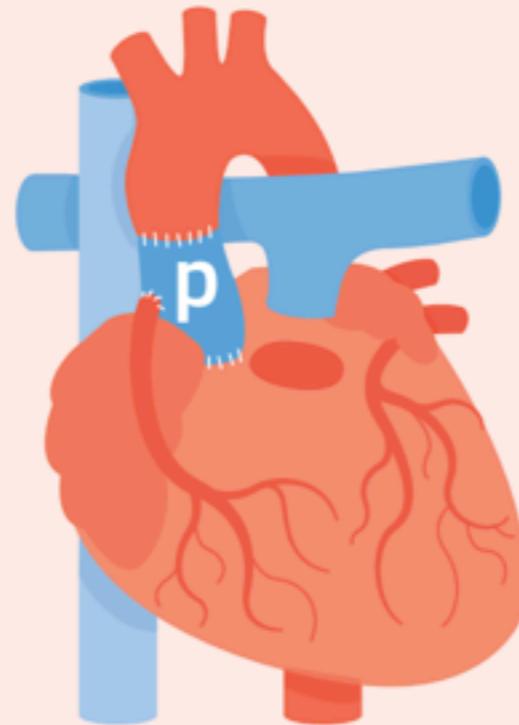




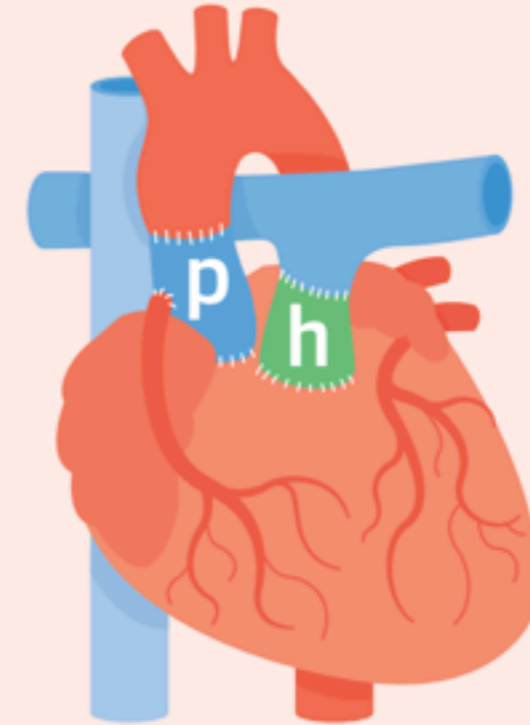
# Ross Procedure



The diseased aortic valve is removed



The pulmonic valve replaces the aortic valve



A homograft replaces the pulmonic valve

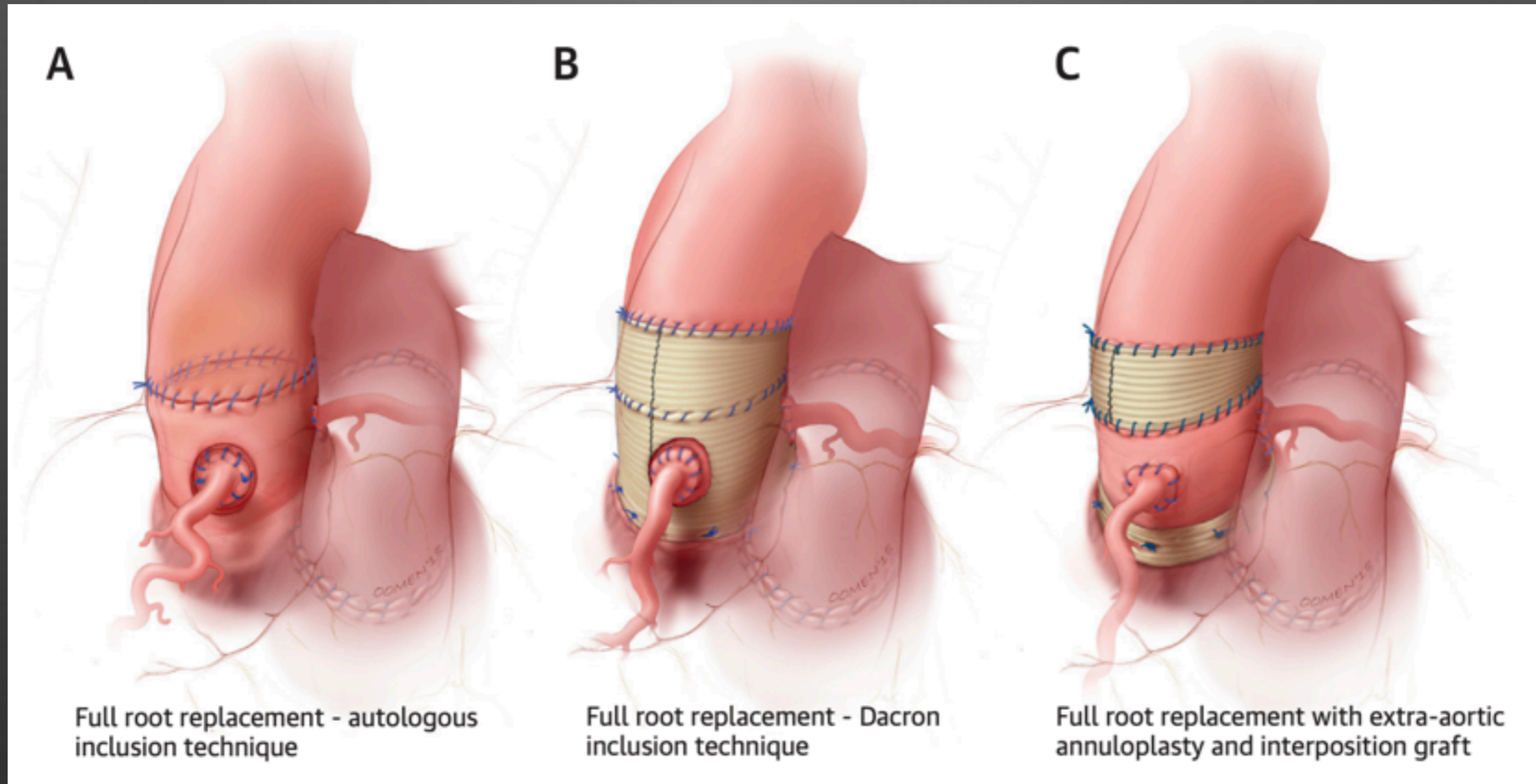


# Risk of Neo-aorta (autograft) Dilation

- Prior Aortic regurgitation
- Dilated aortic annulus
- Dilated ascending aorta
- aortic/pulmonary mismatch
- Uncontrolled hypertension (Goal <110 systolic)
- Unreinforced pulmonary autograft



# Reducing complication risk



# Risk of pulmonary homograft

- Stenosis- valvular and supra-valvular
- Regurgitation (prolapse)
- Size < 19mm
  - Goal >25mm- 93% reoperation at 20 years
- Younger age of placement
- Aortic homografts > pulmonary homografts



**TABLE 1** Summary of Contemporary Series Reporting Long-Term Outcomes ( $\geq 15$  Years) of the Ross Procedure in Adults

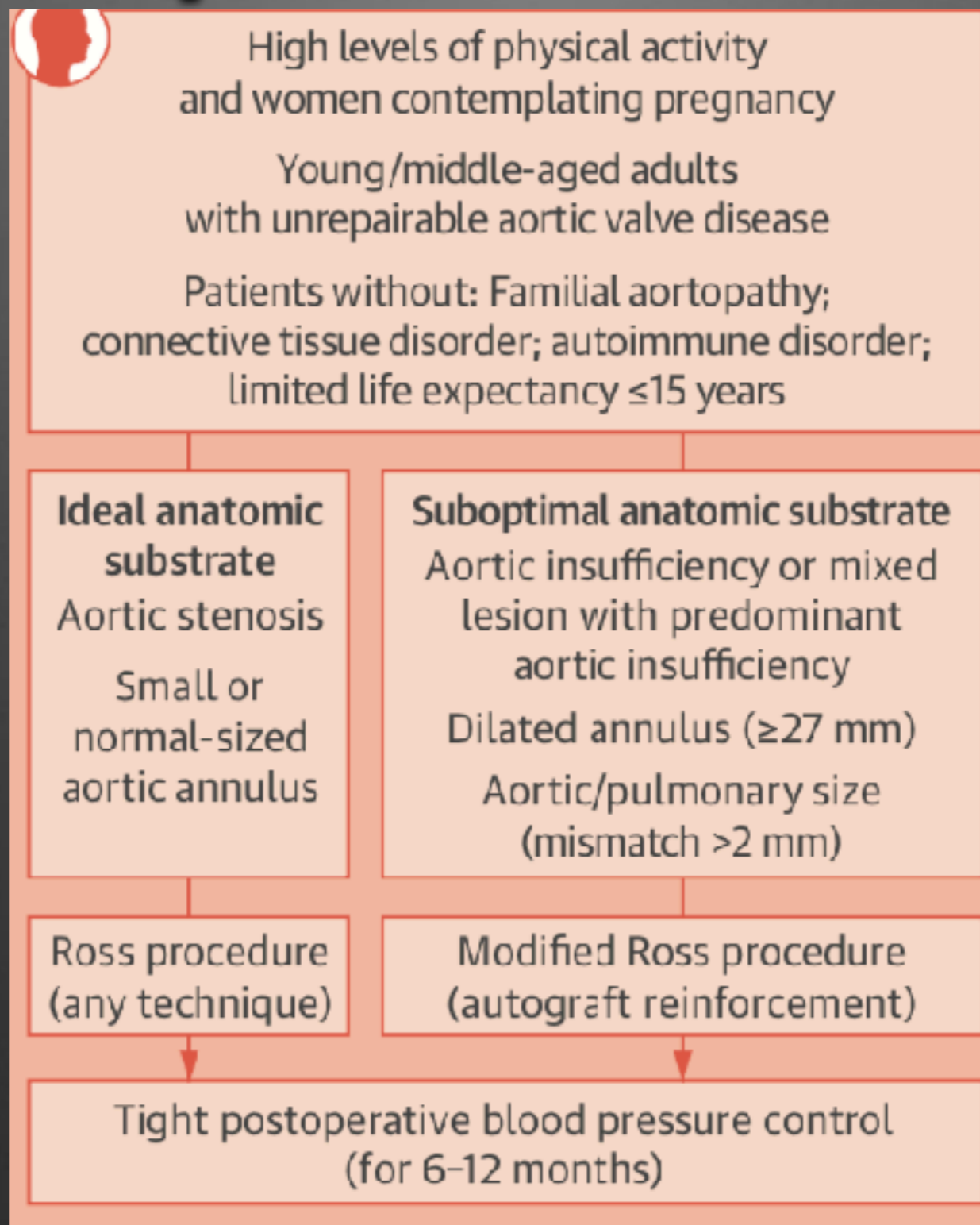
First Author (Year) (Ref. #)	Design	Patients, n	Mean Age, yrs	BAV, %	Pure AI/ Mixed AS-AI, %/%	Mean Follow-Up, yrs	Operative Mortality, %	10-yr Survival, %	15-yr Survival, %	20-yr Survival, %	10-yr Freedom From Reintervention, %*	15-yr Freedom From Reintervention, %*	20-yr Freedom From Reintervention, %*
El-Hamamsy et al. (2010) (9)	RCT	108	38	49	45/27	10.2	0.9	97	95†	—	95	94	—
David et al. (2014) (29)	Single-center	212	34	72	36/13	13.8‡	0.4	98	94	94‡	AG 97 HG 98	AG 93 HG 96	AG 82 HG 93
Da Costa et al. (2014) (101)	Single-center	414	31	50	39/31	8.2	2.7	92	89†	—	90	81	—
Andreas et al. (2014) (100)	Single-center	246	29	75	40/31	10.0‡	1.6	95	91†	—	88	81	—
Skillington et al. (2015) (33)	Single-center	322	39	92	32/22	9.8	0.3	98	97	97†	94	93	—
Mastrobuoni et al. (2016) (31)	Single-center	306	42	59	31/0	10.6‡	2.3	97	88	—	—	75	—
Sievers et al. (2016) (32)	Multicenter (prospective)	1,779	45	65	22/52	8.3	1.1	96	90†	—	91	83	—
Martin et al. (2017) (30)	Single-center	310	41	73	19/7	15.1‡	1.3	94	92	84	93	86	70
Sievers et al. (2018) (128)	Single-center	630	45	78	24/—	12.5‡	0.3	95	87	73†	AG 96 HG 97	AG 94 HG 94	AG 90 HG 91

\*Includes any reintervention on the pulmonary autograft and/or pulmonary homograft. †Survival equivalent to age- and sex-matched general population. ‡Median (rather than mean) follow-up.

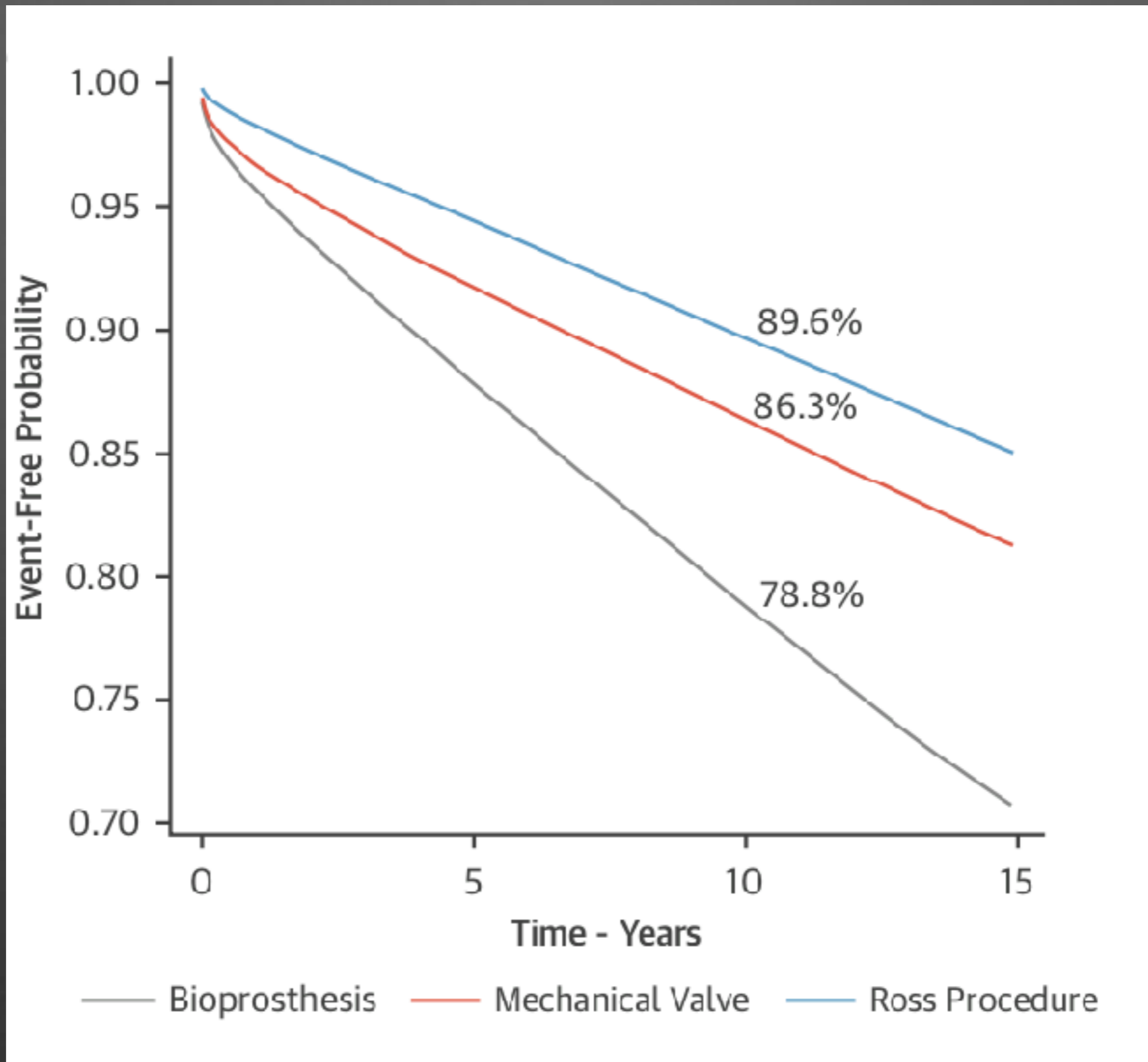
AG — autograft; AI — aortic insufficiency; AS — aortic stenosis; BAV — bicuspid aortic valve; HG — homograft; RCT — randomized controlled trial.



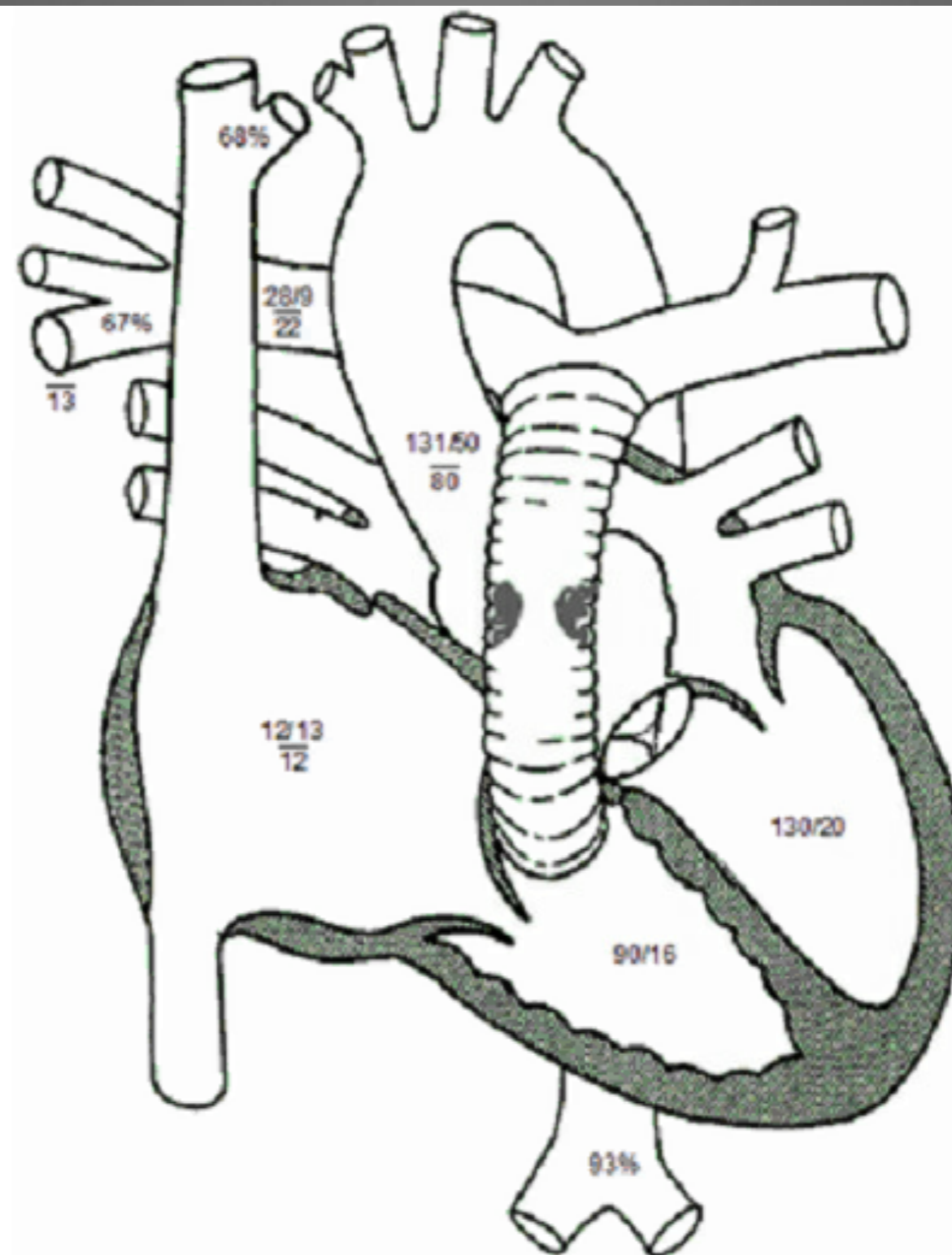
# Considering the Ross procedure?



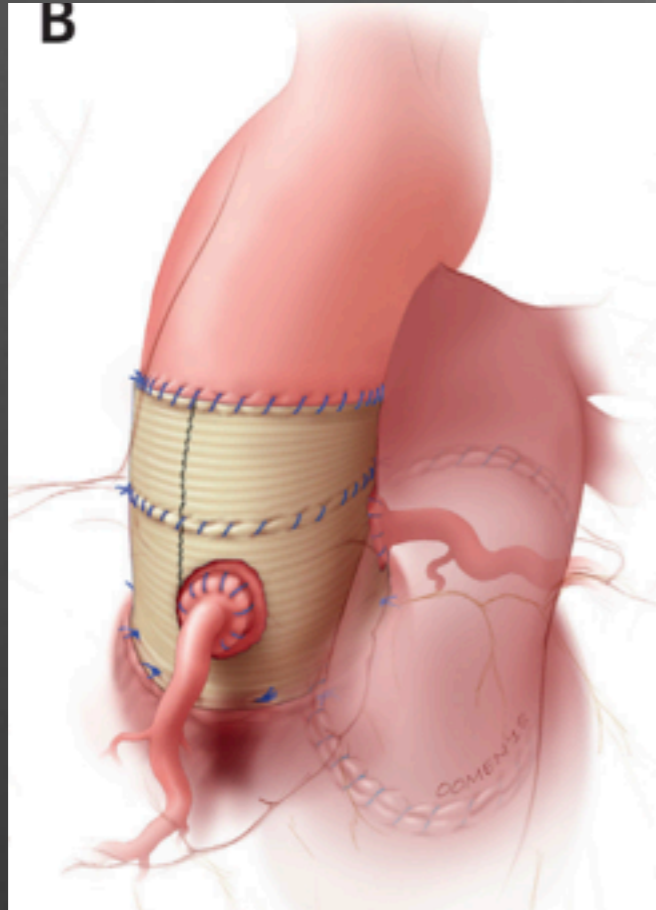




# Cath findings

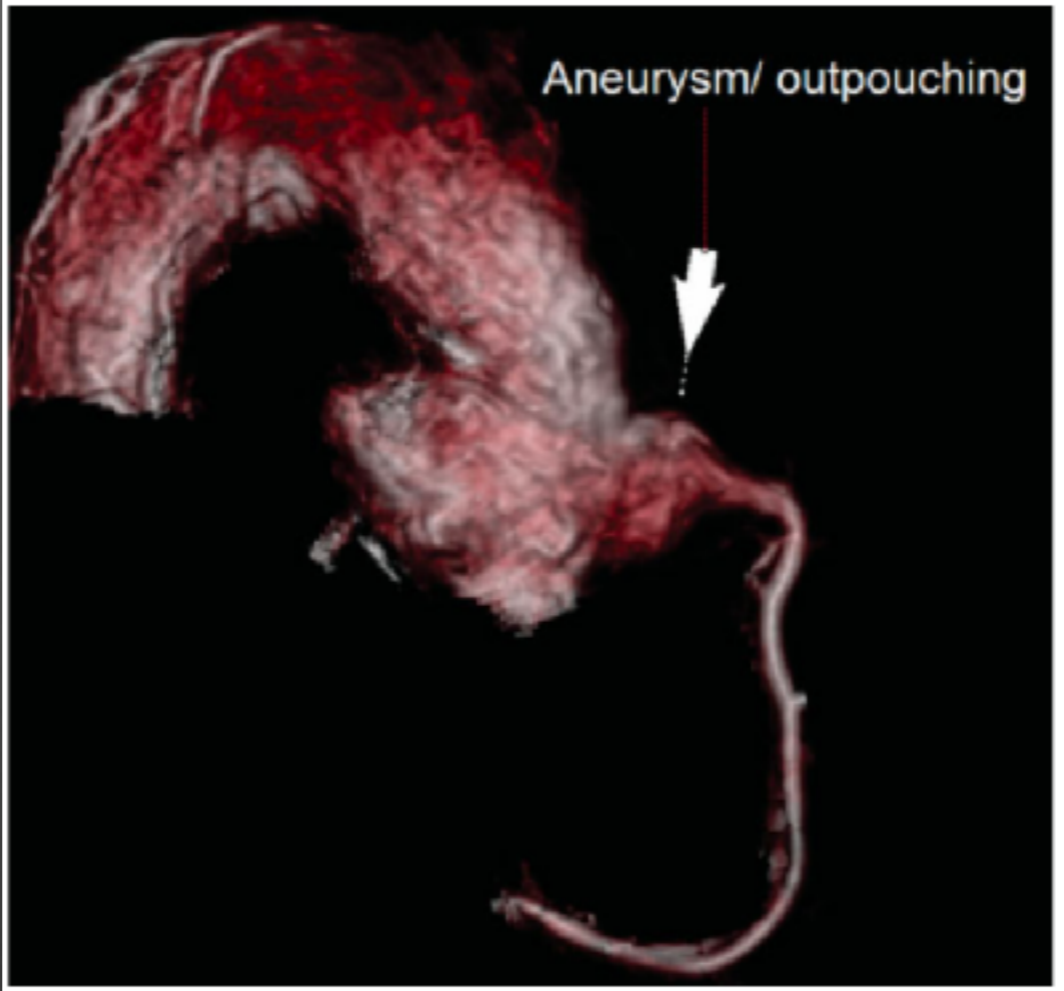


# Uncommon Complications



- 19 y.o.
- Aortic stenosis — BAV 3 months
- AI and age 12 underwent ROSS procedure
- 24mm Pulmonary homograft
- Dacron graft at the aortic annulus
- 28 mm Hemishield aortic graft around the pulmonary autograft





# BMJ Open Ross for Valve replacement In Adults (REVIVAL) pilot trial: rationale and design of a randomised controlled trial

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# Diagnosis and Plan

- Severe Neo-aortic root dilation with severe AI
- Severe pulmonary homograft stenosis
- Bentall procedure with On-X valve for aorta and bioprosthetic valve for pulmonary position with augmentation of the main PA (goal >27mm)

**Look beyond the aorta in the young with prior scar**



## THE PRESENT AND FUTURE

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### JACC STATE-OF-THE-ART REVIEW

# Ross Procedure in Adults for Cardiologists and Cardiac Surgeons

## JACC State-of-the-Art Review

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