



# When do Adults with Congenital Heart Disease Need an Intervention? Role of Echo (Multimodal Imaging)

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# Congenital Heart Disease

- Babies with congenital heart disease (~0.8% live births)
  - 1940's survival < 20%
  - 1970's survival >65%
  - 2000's survival >90%



## Adult Congenital Patients

- 13 million world wide
- 3000 per million world wide
- 1.4 million in the US

Fetal Diagnosis

Advances in  
NICU

Decreased  
Perioperative  
mortality

Improved  
Surgical  
Techniques

Advances in care  
of ACHD



# Congenital Heart Disease

- Tetralogy of Fallot
- D-Transposition of the Great Arteries
  - Atrial Switch
  - Arterial Switch
- Ebstein Anomaly



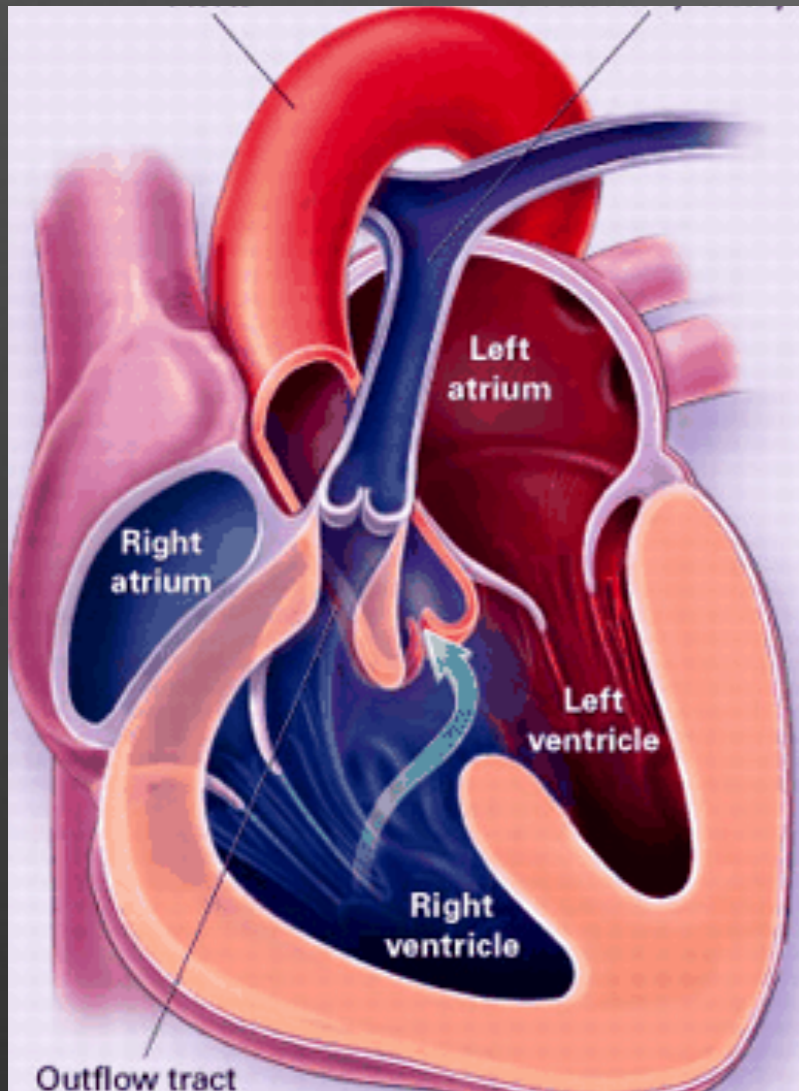
# Adult Congenital Heart Disease

Anatomy, Surgeries, and  
Complications

CV Testing

Percutaneous/Surgical

# Tetralogy of Fallot



## GUIDELINES AND STANDARDS

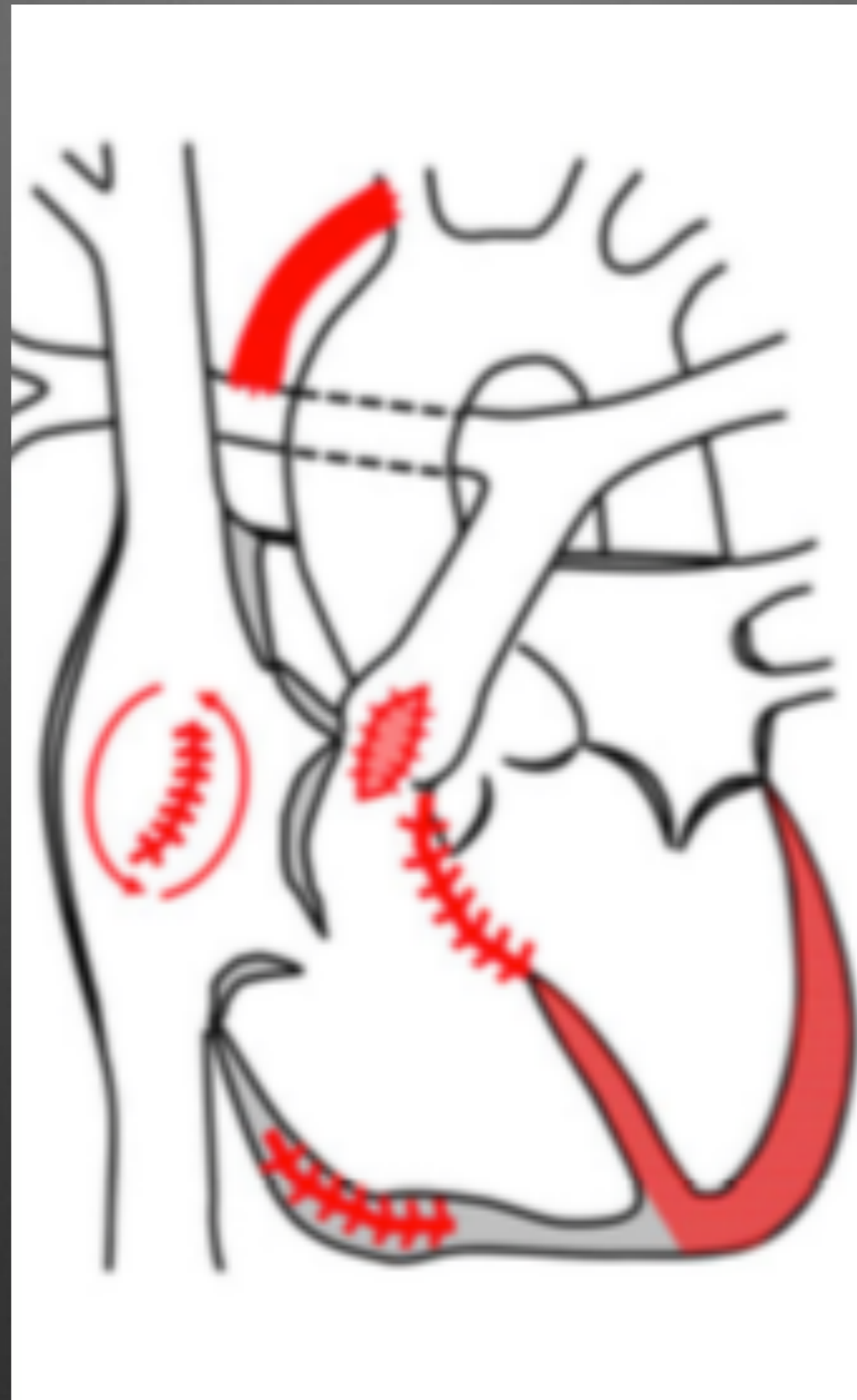
### Multimodality Imaging Guidelines for Patients with Repaired Tetralogy of Fallot: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society for Pediatric Radiology

Anne Marie Valente, MD, FASE, Co Chair, Stephen Cook, MD, Pierluigi Festa, MD, H. Helen Ko, BS, RDMS, RDCS, FASE, Rajesh Krishnamurthy, MD, Andrew M. Taylor, MD, Carole A. Warnes, MD, Jacqueline Kreutzer, MD, and Tal Geva, MD, FASE, Co Chair, *Boston, Massachusetts; Pittsburgh, Pennsylvania; Massa, Italy; New York, New York; Houston, Texas; London, United Kingdom; Rochester, Minnesota*

(J Am Soc Echocardiogr 2014;27:111-41.)



# Tetralogy of Fallot



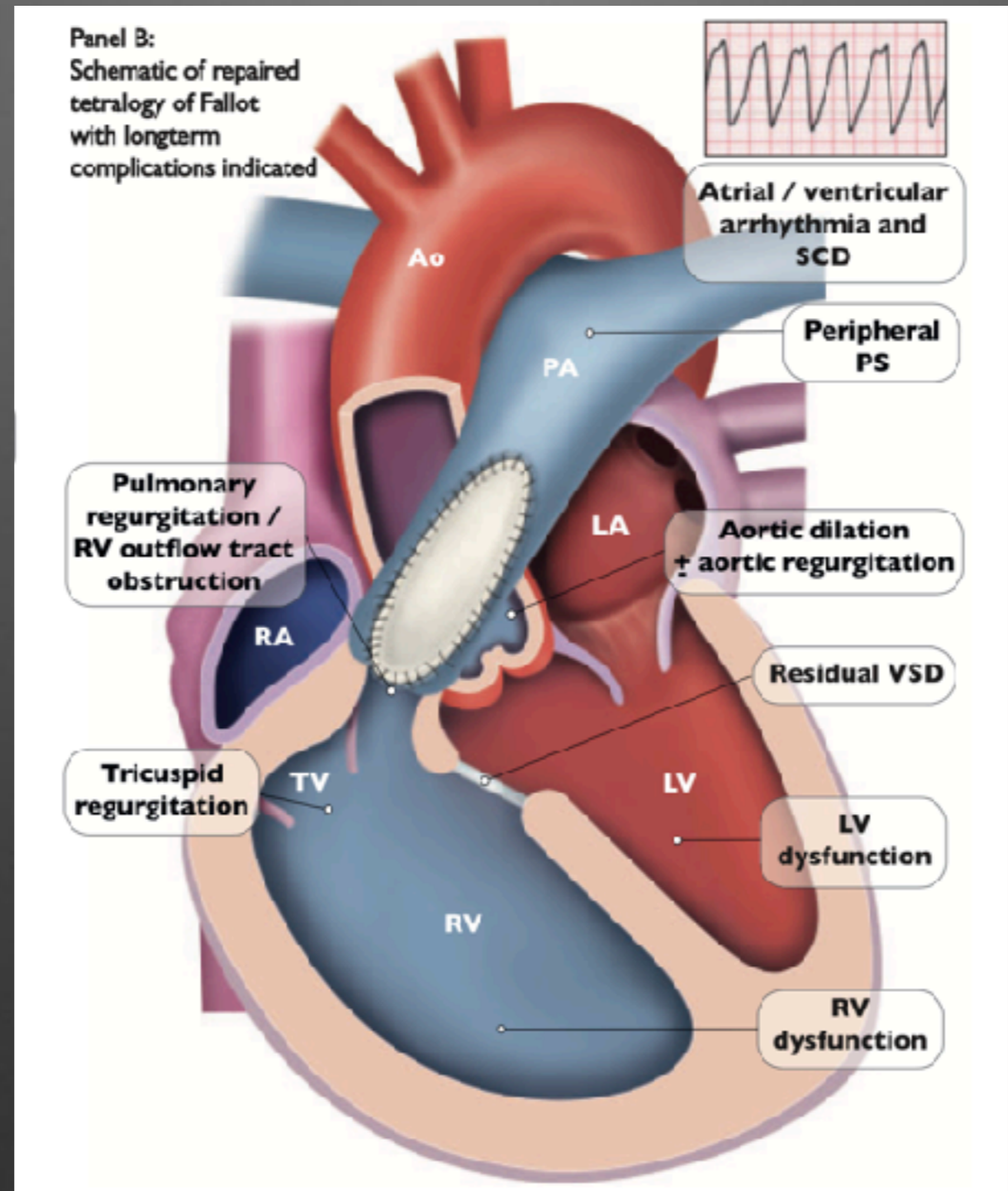
# Complications

Echocardiography

CMR

CT

CPET





# ToF PVR for > Moderate PR

		CLASS I	CLASS IIA (ANY 2 OF THE FOLLOWING)
ACC 2018	Symptoms		<ul style="list-style-type: none"> <li>• RV/LV dysfunction</li> <li>• RVEDVi <math>\geq</math> 160 mL/m<sup>2</sup>, RVESVi <math>\geq</math> 80mL/m<sup>2</sup> or RVEDVi <math>\geq</math> 2 x LVEDVi</li> <li>• RVSP <math>\geq</math> 2/3 systemic</li> <li>• Objective reduction in exercise tolerance</li> </ul>
EURO 2020	Symptoms		<p style="text-align: center;"><b>Severe PR and/or RVOTO</b></p> <ul style="list-style-type: none"> <li>• Progressive RV dysfunction</li> <li>• Progressive dilation RVEDVi &gt; 160 mL/m<sup>2</sup>, RVESVi &gt; 80mL/m<sup>2</sup> or progressive TR at least moderate</li> <li>• RVSP &gt; 80 mmHg</li> <li>• Objective reduction in exercise tolerance</li> </ul>



# Tetralogy of Fallot

<Moderate PR

> Moderate PR

RVOTO >2/3  
systemic

Symptoms

No symptoms

No symptoms

- Severe PR
  - Progressive RV dysfunction (<47%)
  - RVEDVi/RVESVi >160/80 mL/m<sup>2</sup>
  - RVEDV > 2x LVEDV
  - Objective exercise intolerance

- Identify area of obstruction
- Progressive exercise intolerance
  - Arrhythmias

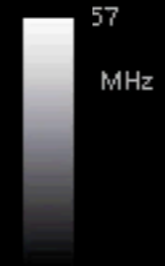
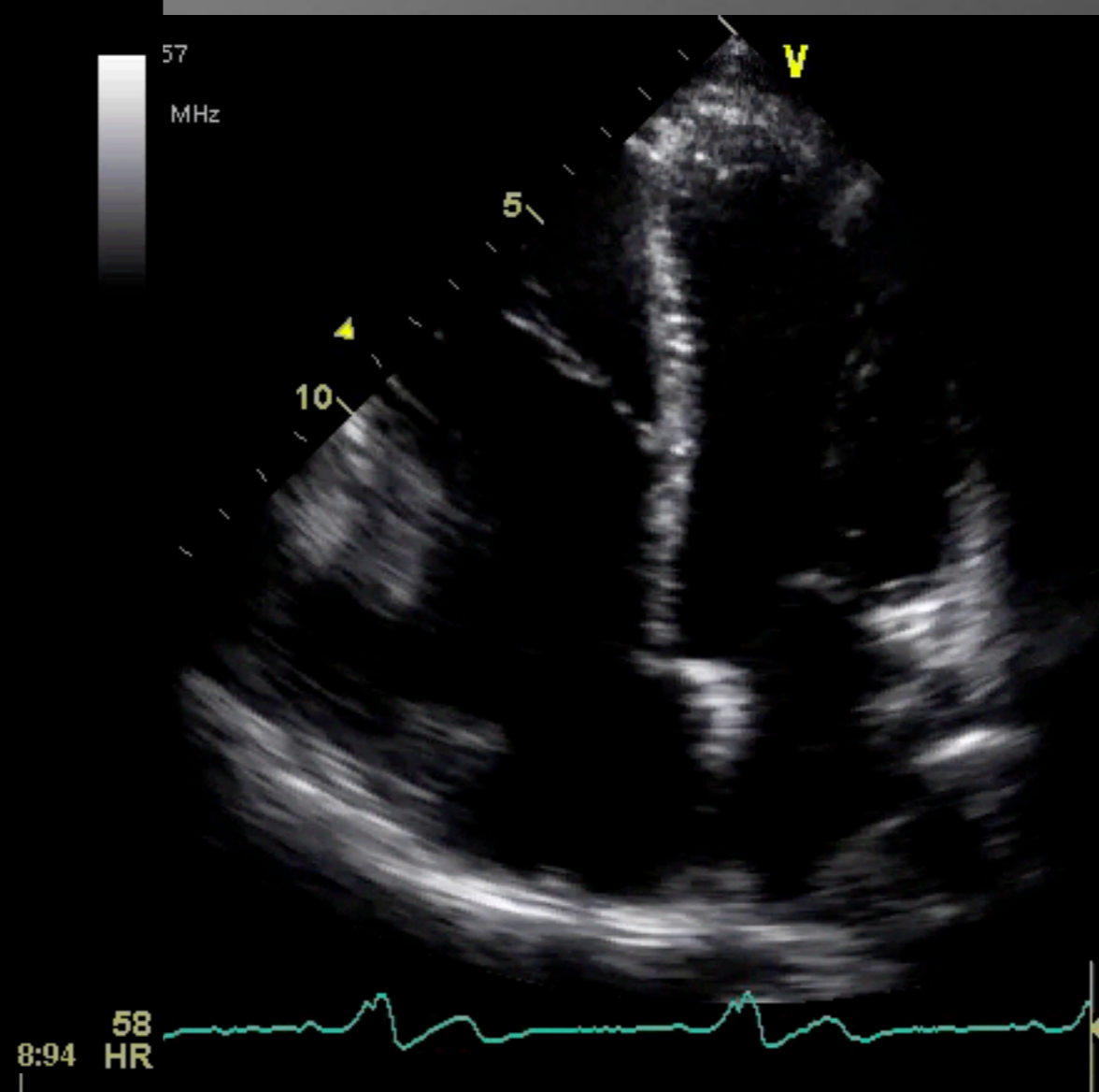
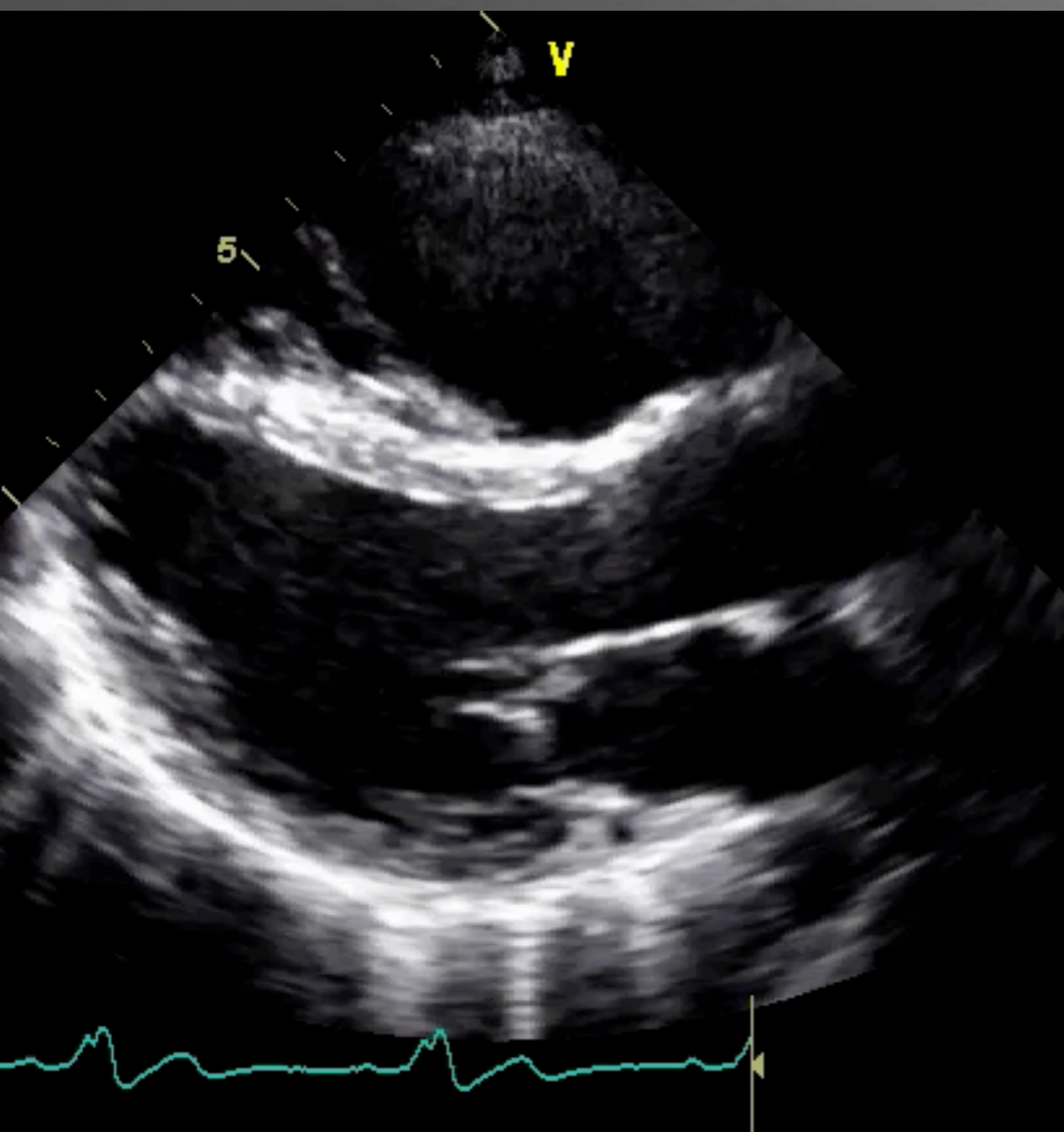
PVR



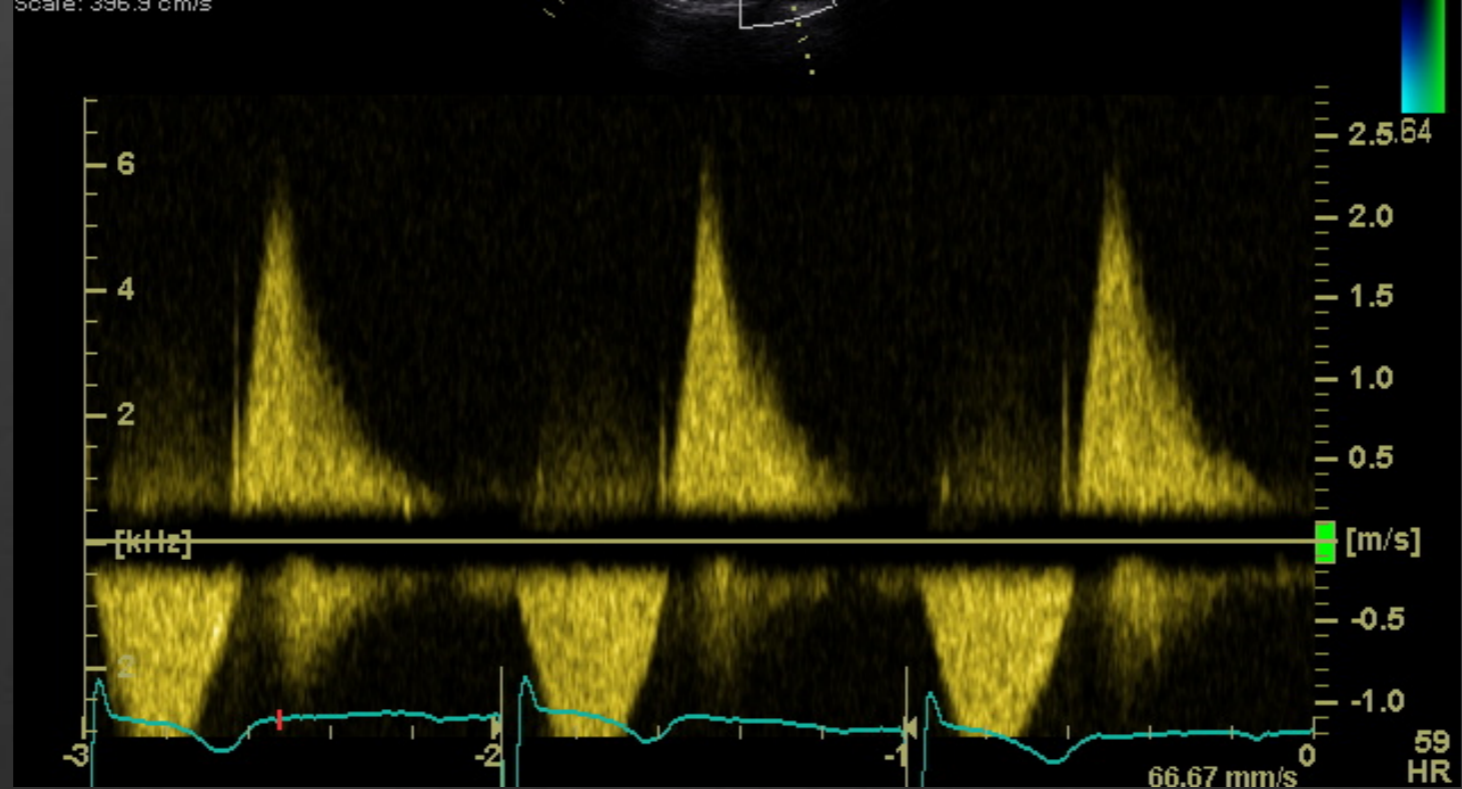
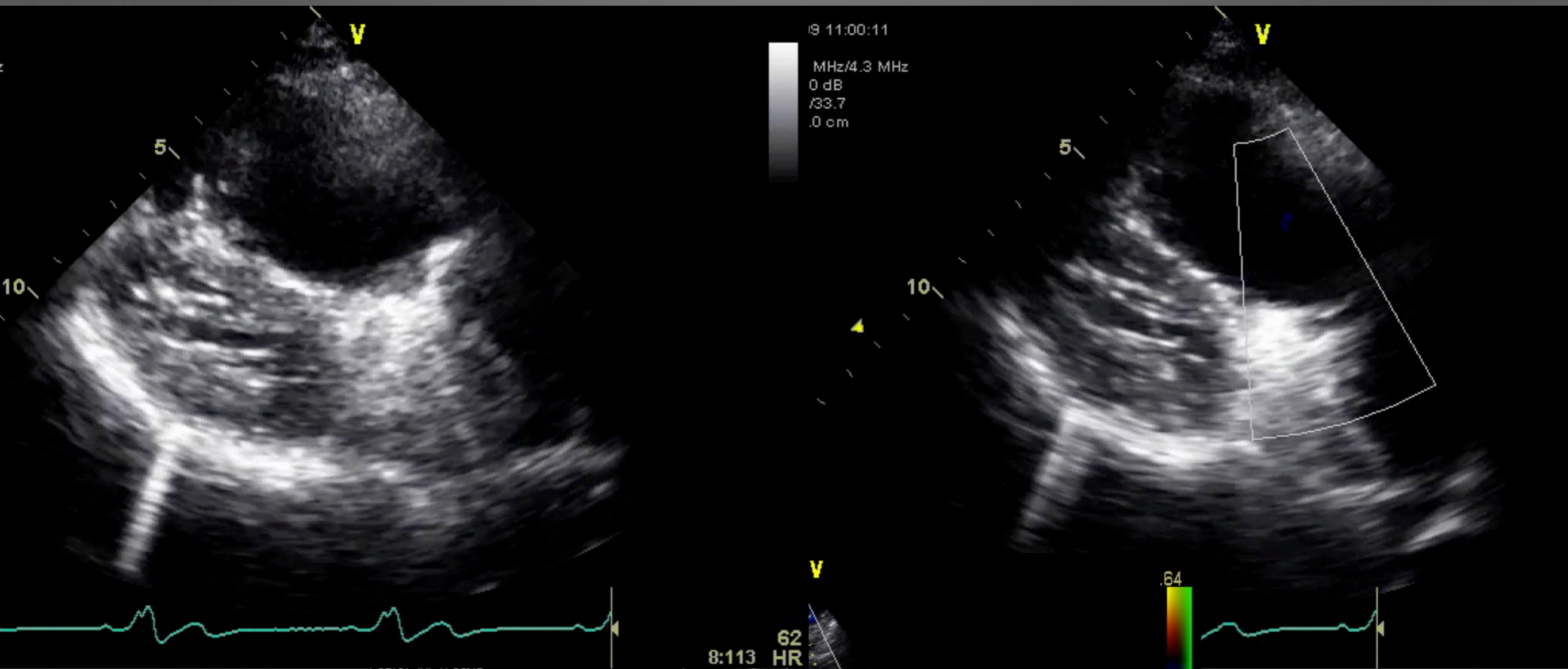
# Case 1

- 40 y.o. male who states over the last year has noted worsening fatigue and palpitations
- Congenital Heart History: ToF with two surgeries - Thoractomy and then sternotomy. Last surgery at age 5.

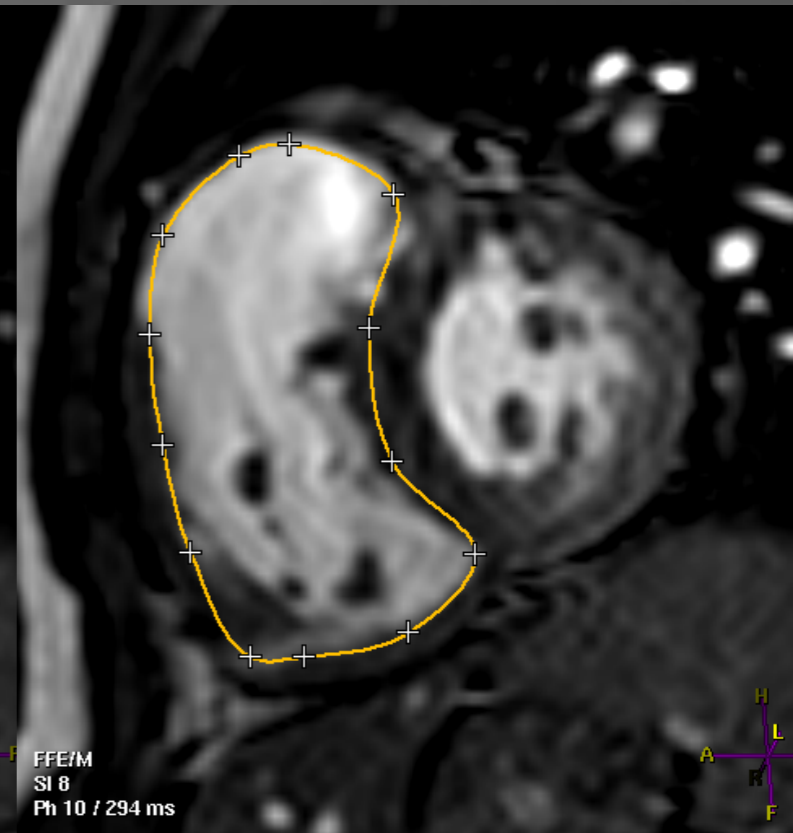
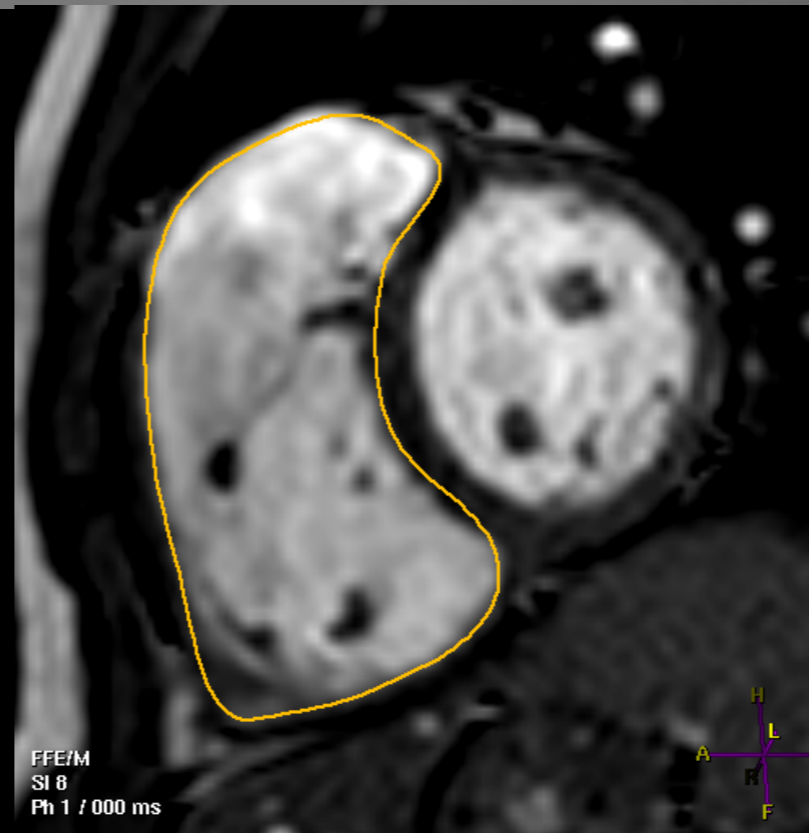
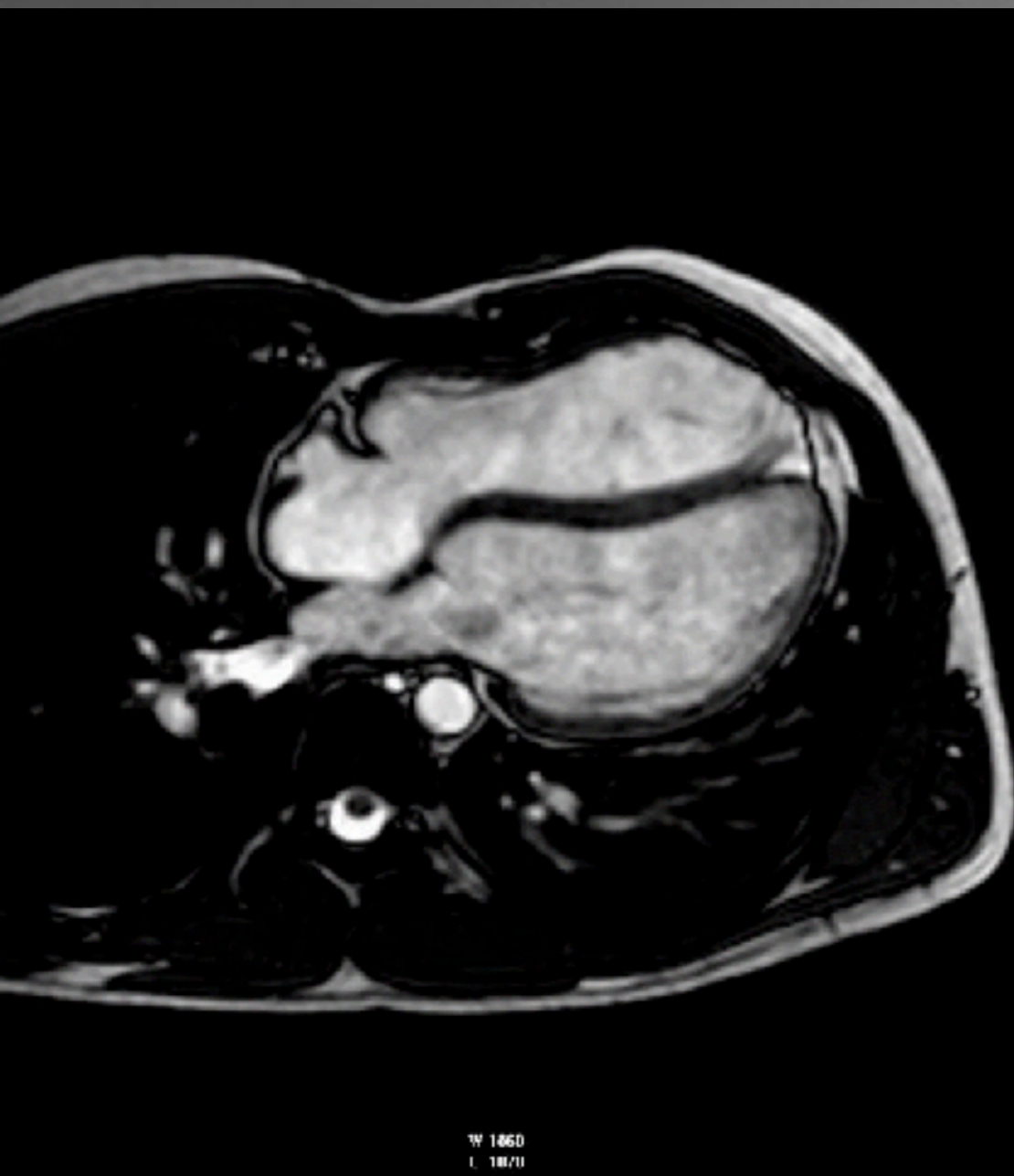




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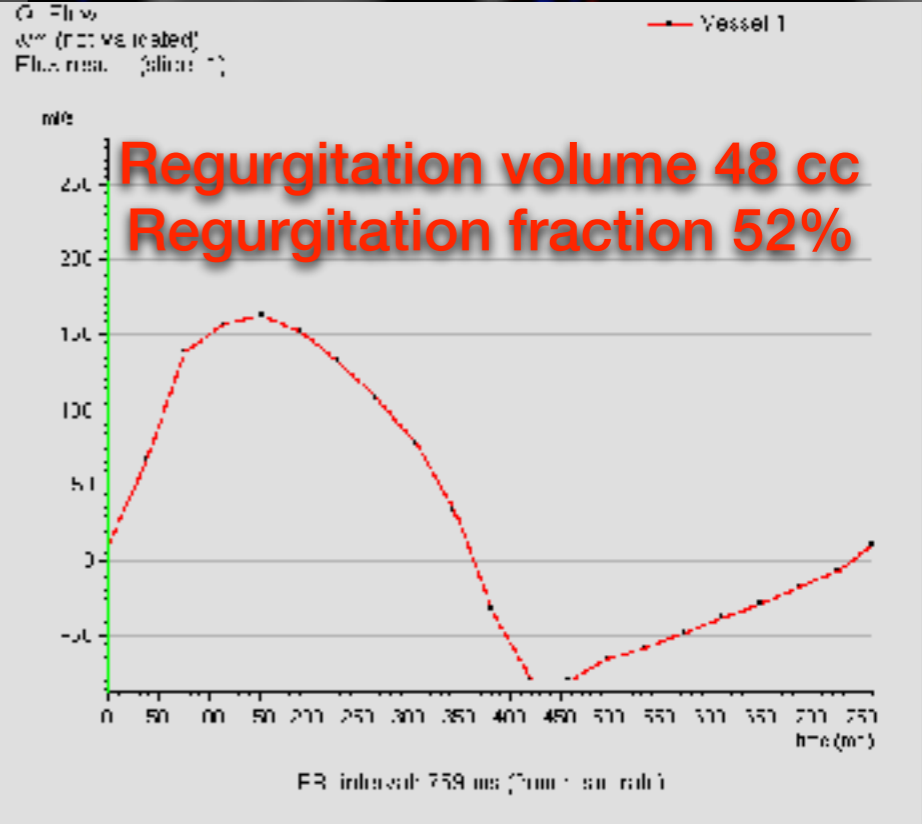
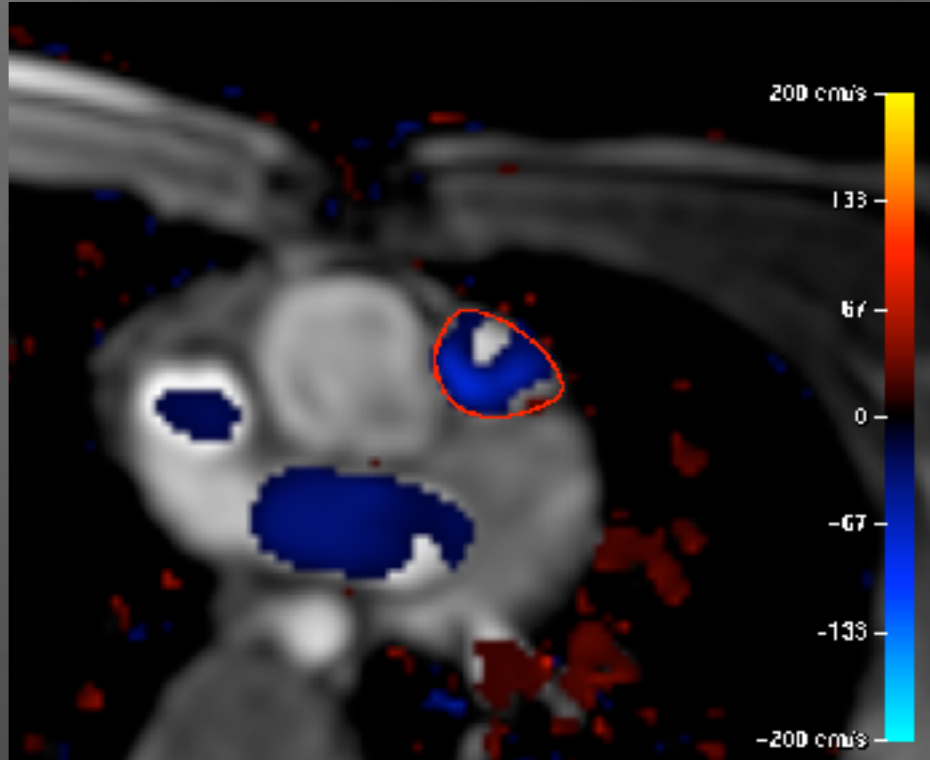
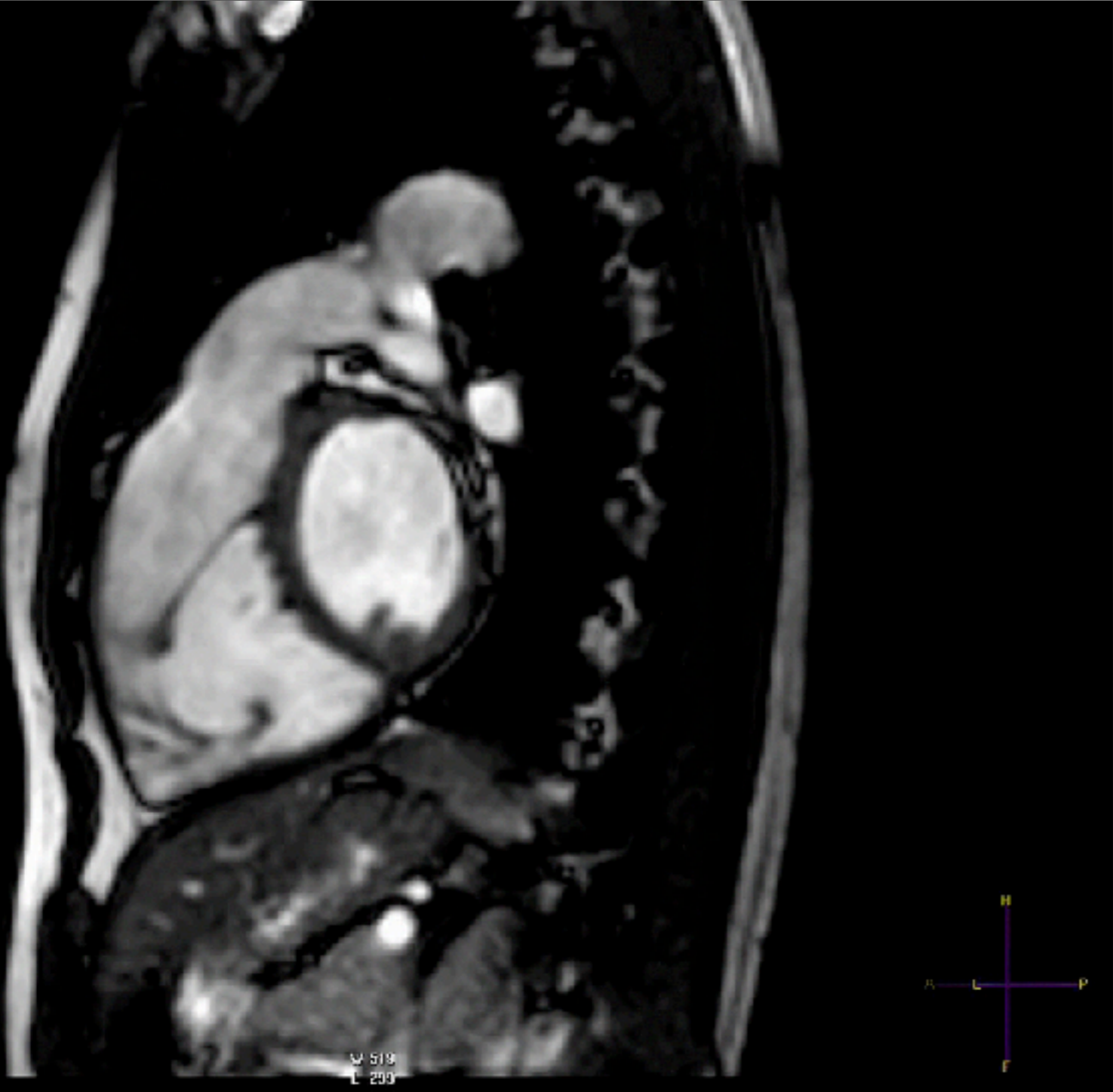


# RV dilation & dysfunction



- RV EF of 39%
- RVEDV: 167cc/m<sup>2</sup>
- RVESV: 102cc/m<sup>2</sup>

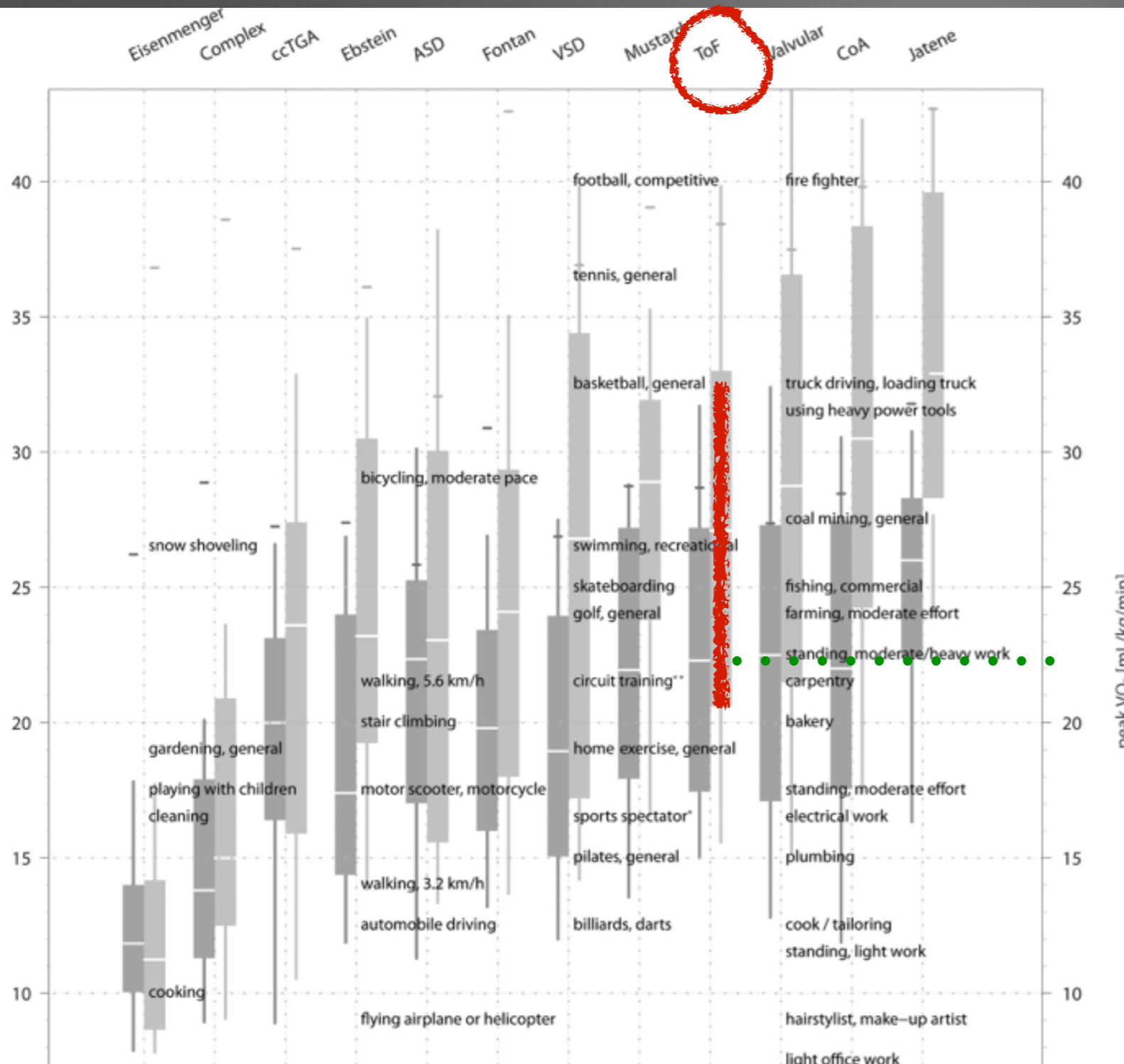
# Cardiac MRI







# Cardiopulmonary Stress



peak  $VO_2$ , [mL/kg/min]

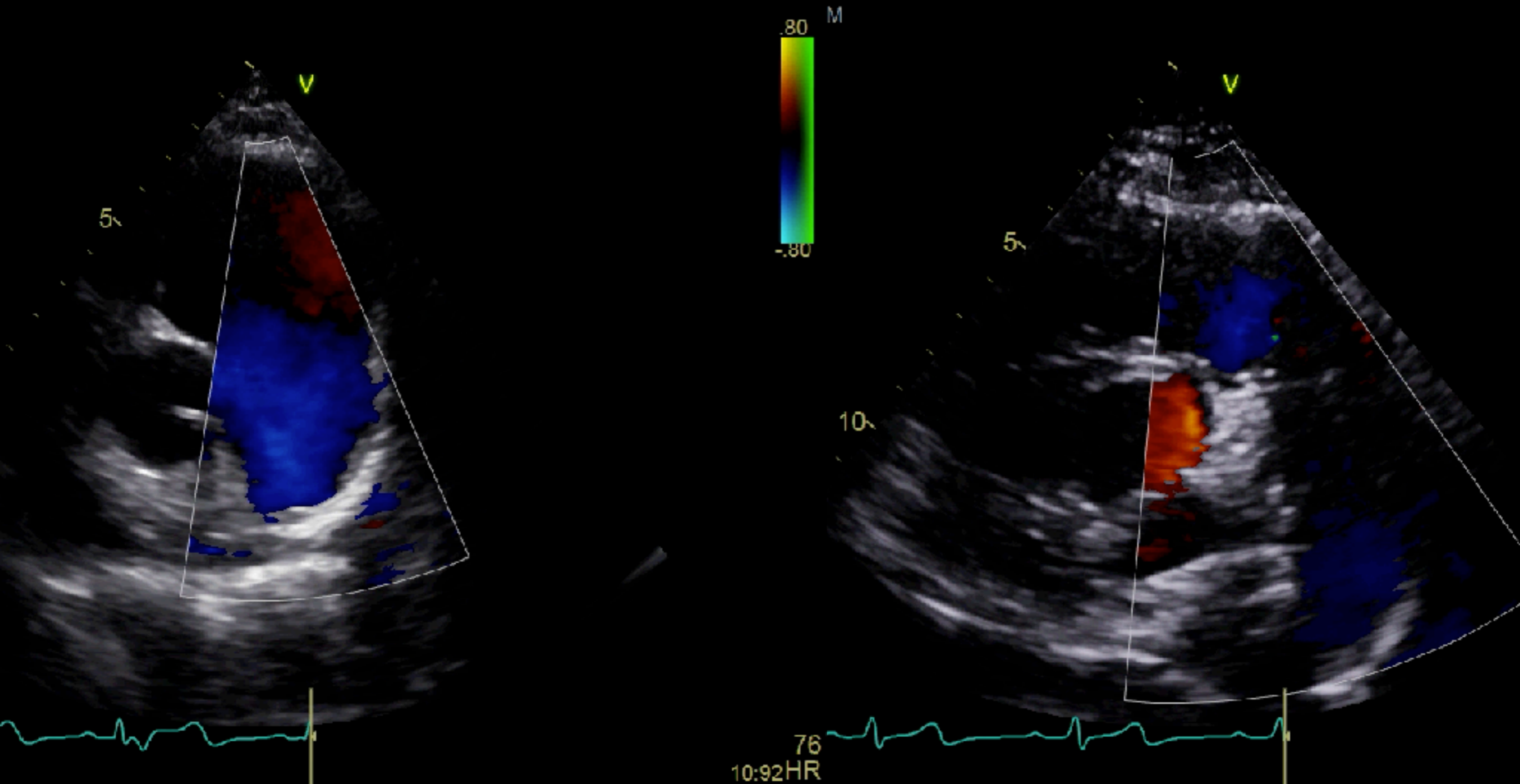
# Management: Surgery

- Therapeutic Decision:
  1. Surgical pulmonary valve replacement
  2. reconstruction of RVOT aneurysm
  3. patch augmentation of left PA

# After PVR

- RV improvement in volumes & function
- LV improvement in function
- QRS duration decreases
- Symptoms improve

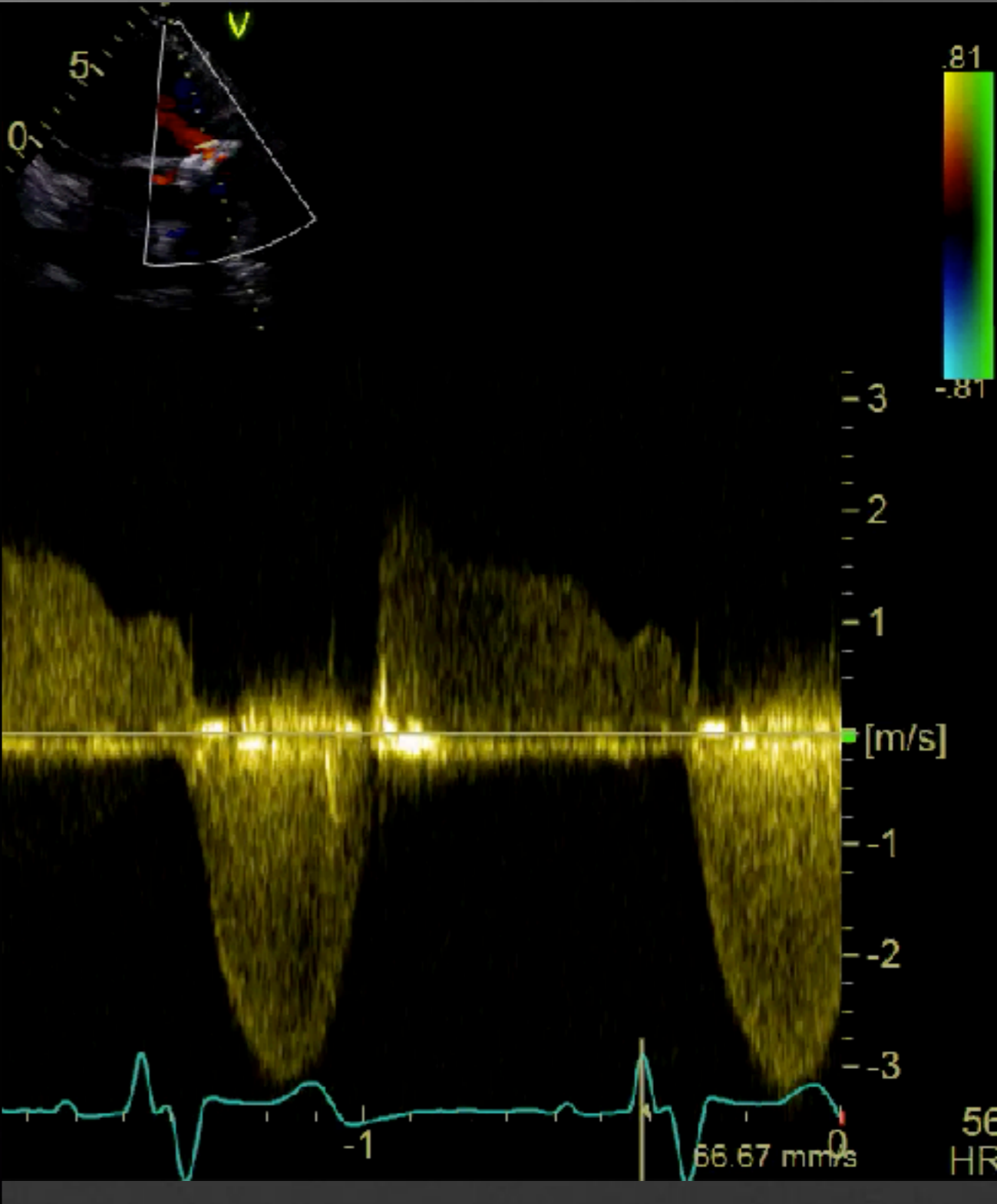
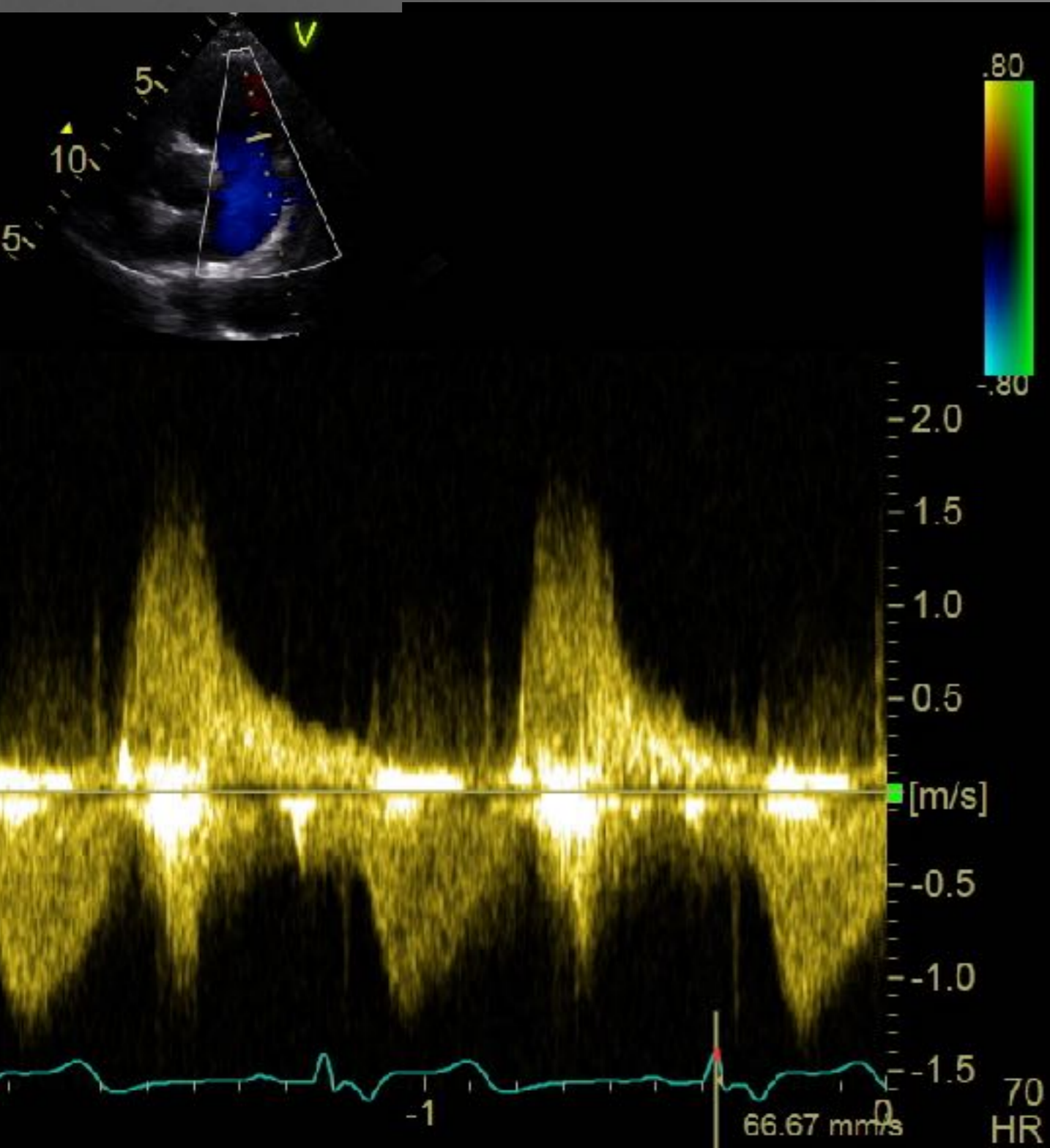
# Pulmonary regurgitation



Severe PR

Mild PR





Severe PR

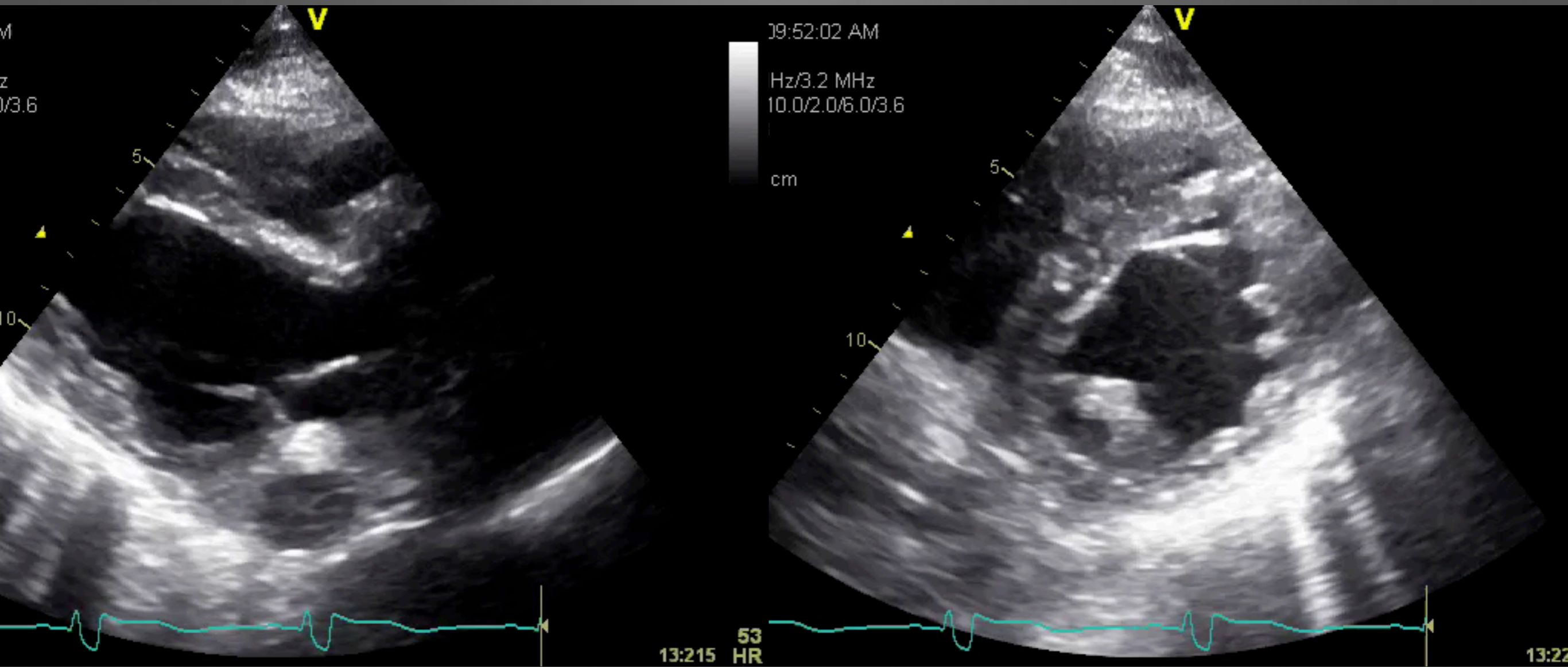
Mild PR



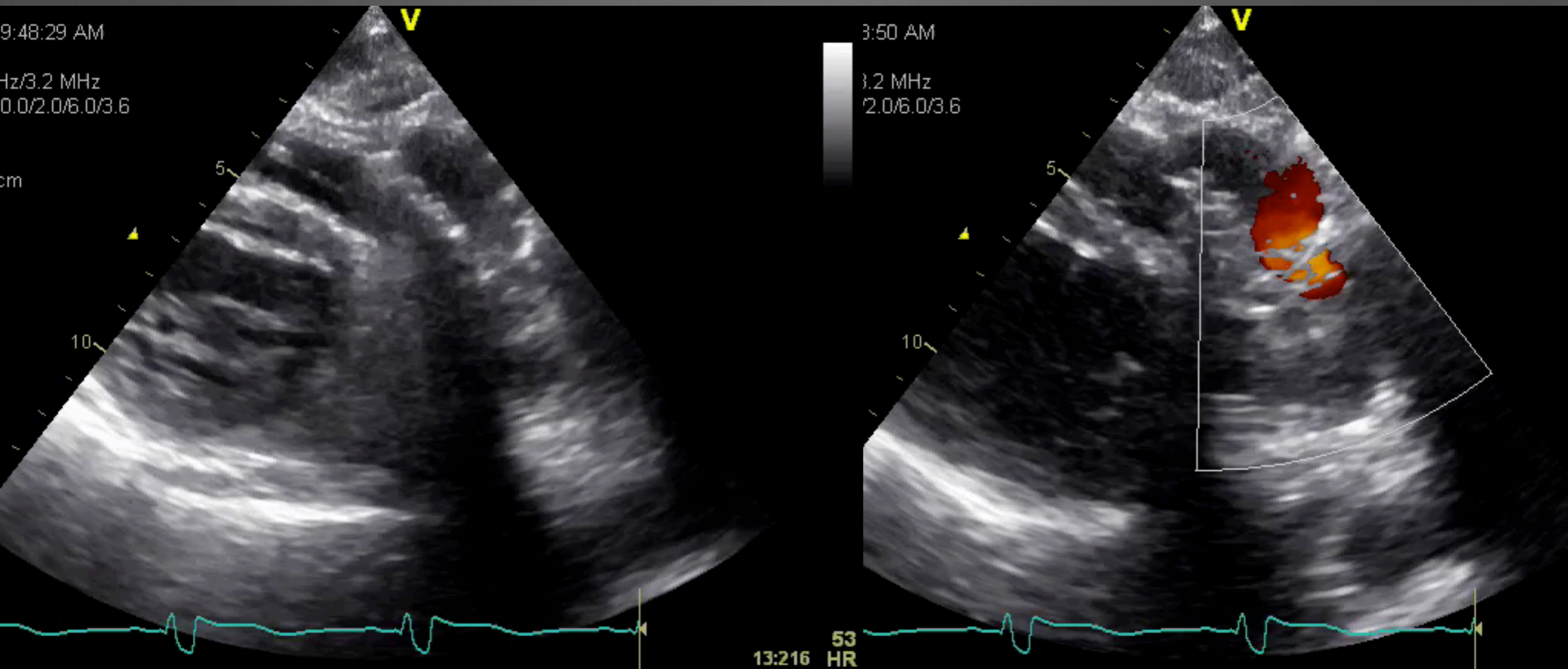
# Case 2

- 32 y.o. male with worsening symptoms of exercise intolerance
- PE:BP 110/70 with HR of 77 bpm; height 70in; weight 200 lbs; BSA 2.1 m<sup>2</sup>
  - Right thoracotomy scar and mid sternal scar
  - 3/6 systolic EM at RUSB with a 1/6 early peaking diastolic murmur

# Tetralogy of Fallot

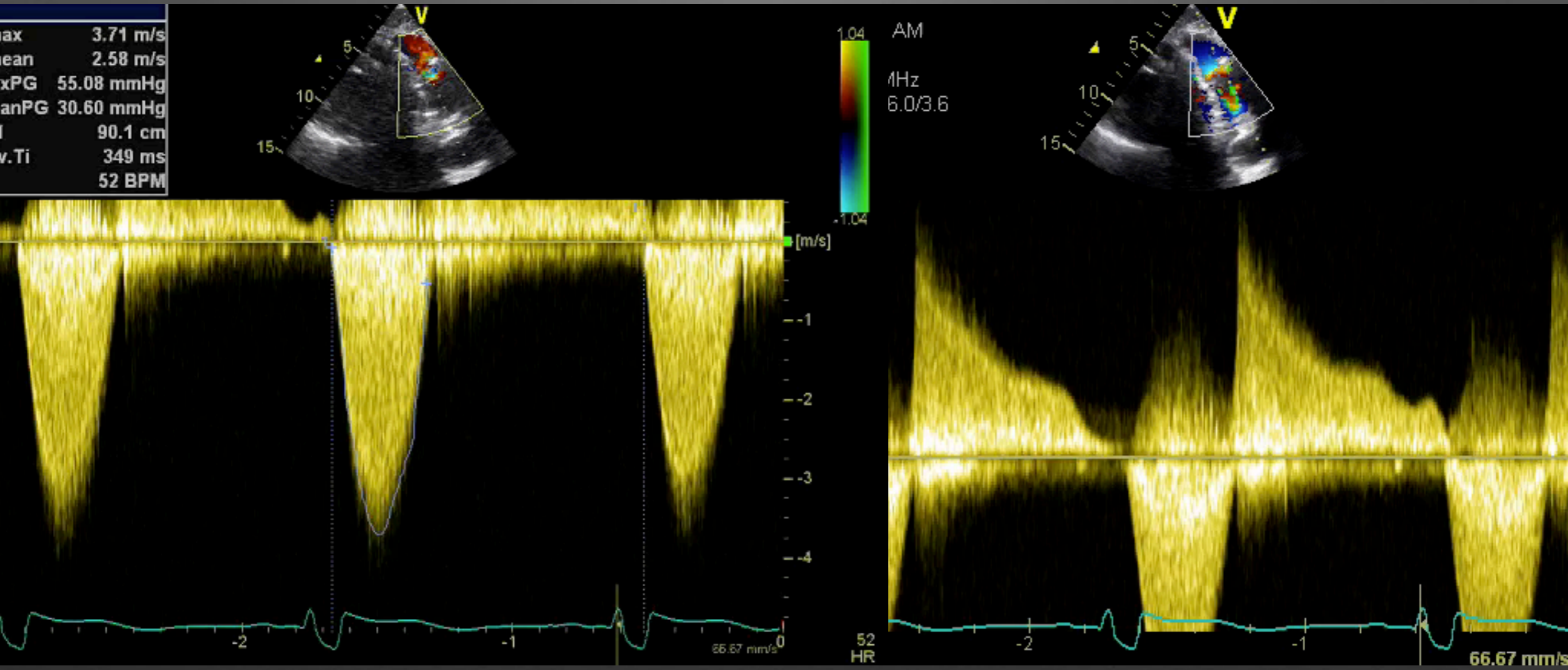


# RVOT View- Conduit





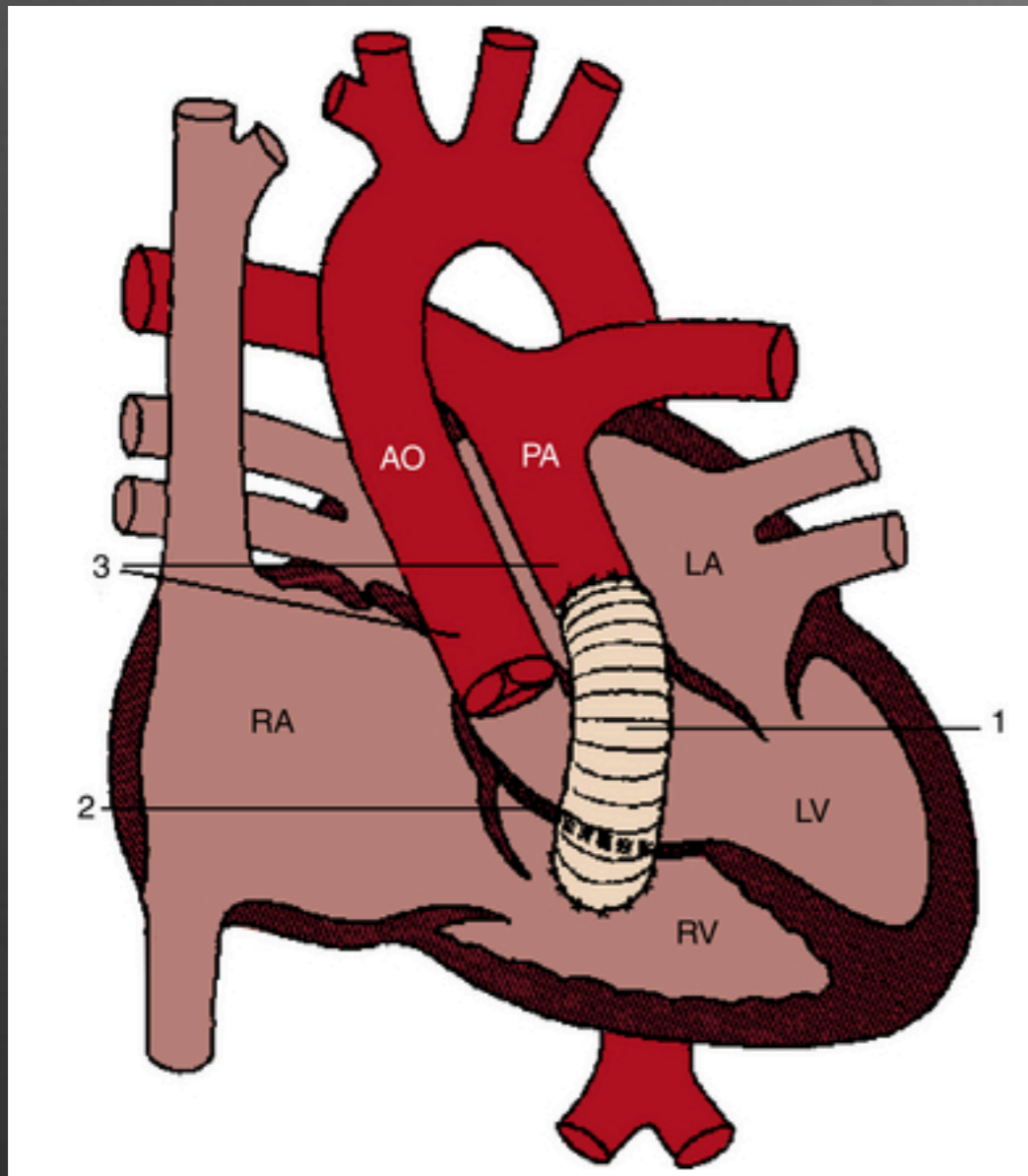
# CW Doppler



3.7 m/sec; mean 31mmHg;  
peak 55mmHg

Moderate-severe PI

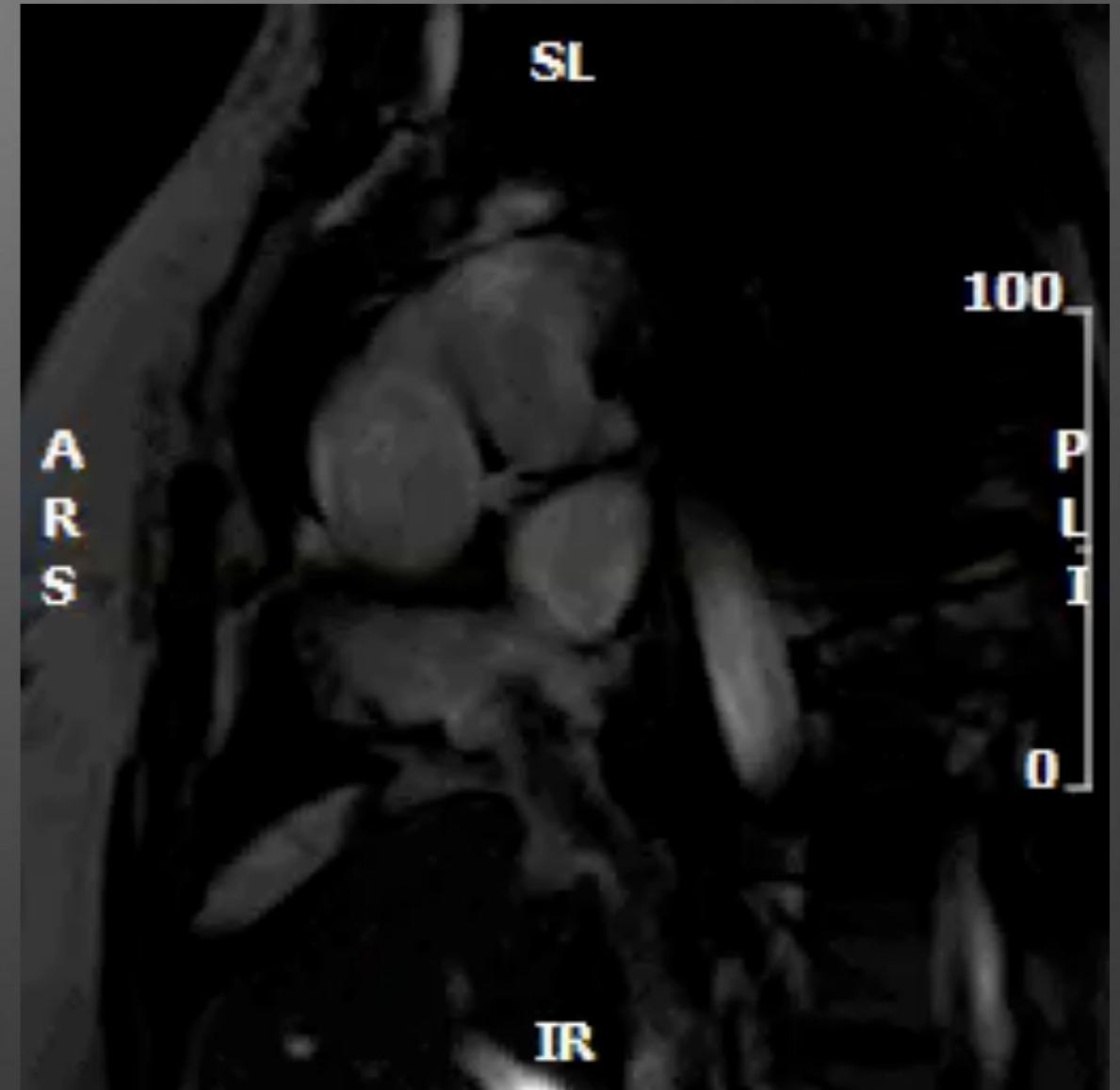
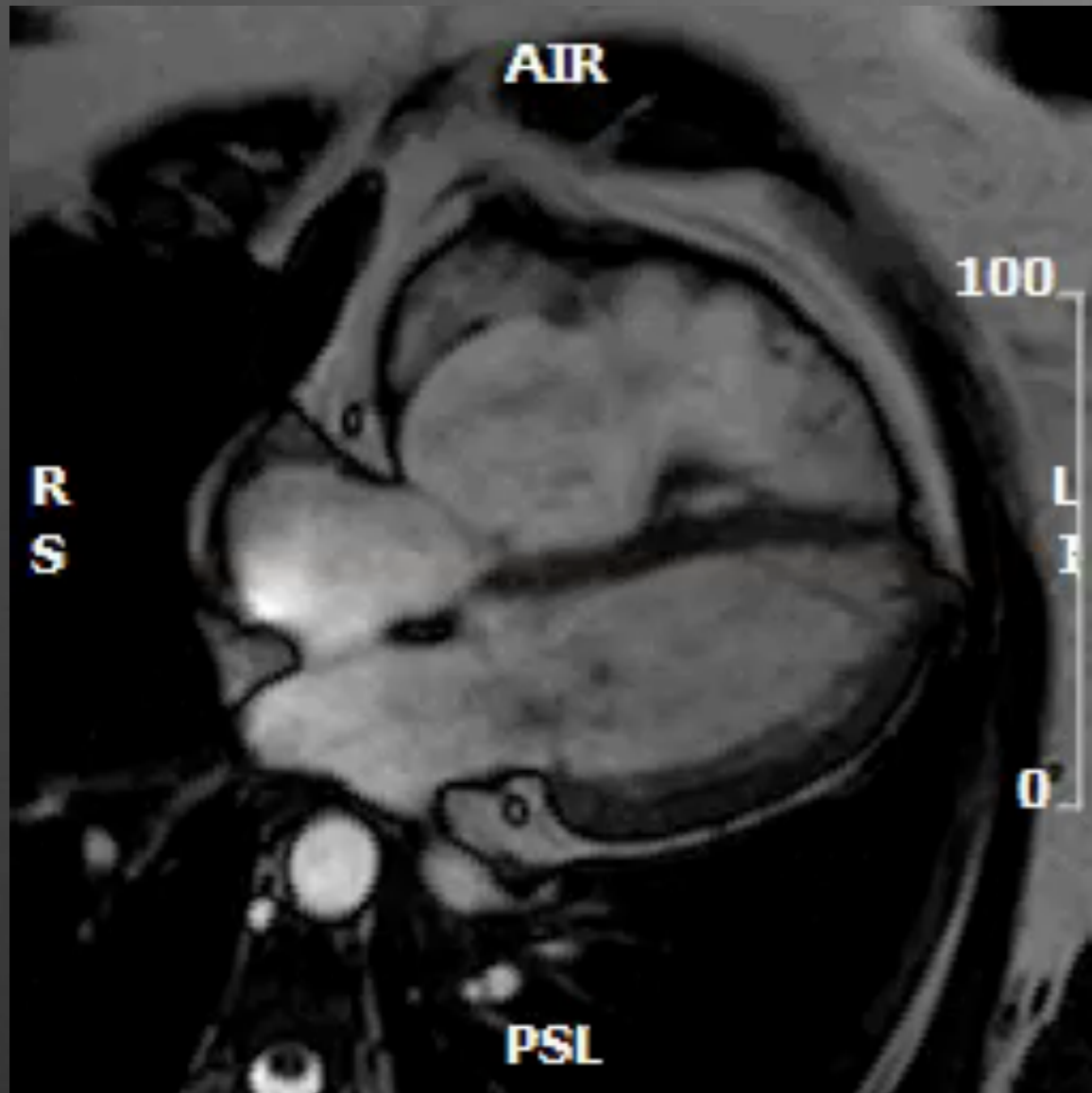
# Case 2



- Thoractomy: Ao to PA shunt
- Repair of ToF: VSD patch closure, take down of shunt, surgical valvulotomy
- Redo: RV to PA conduit

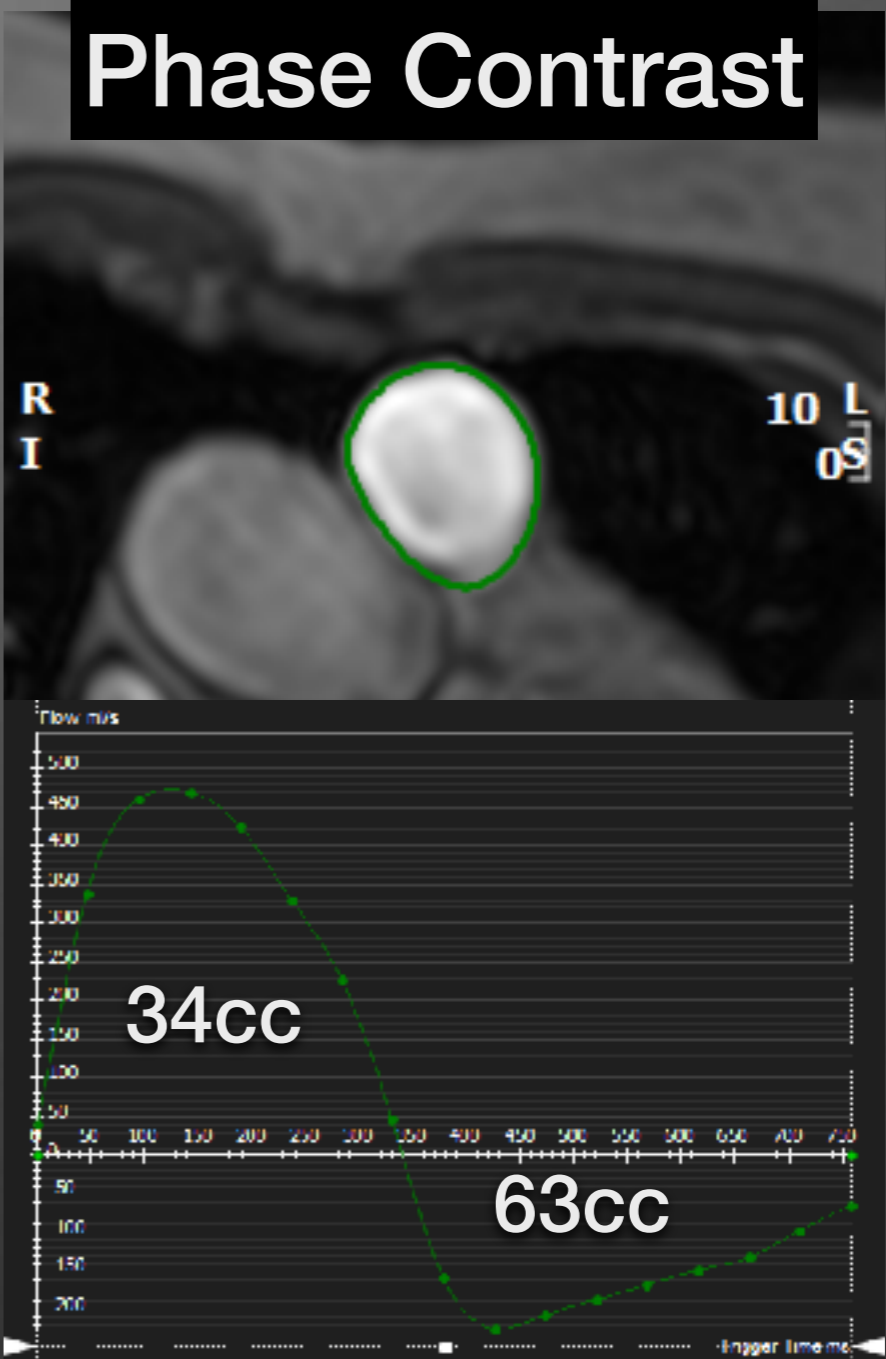
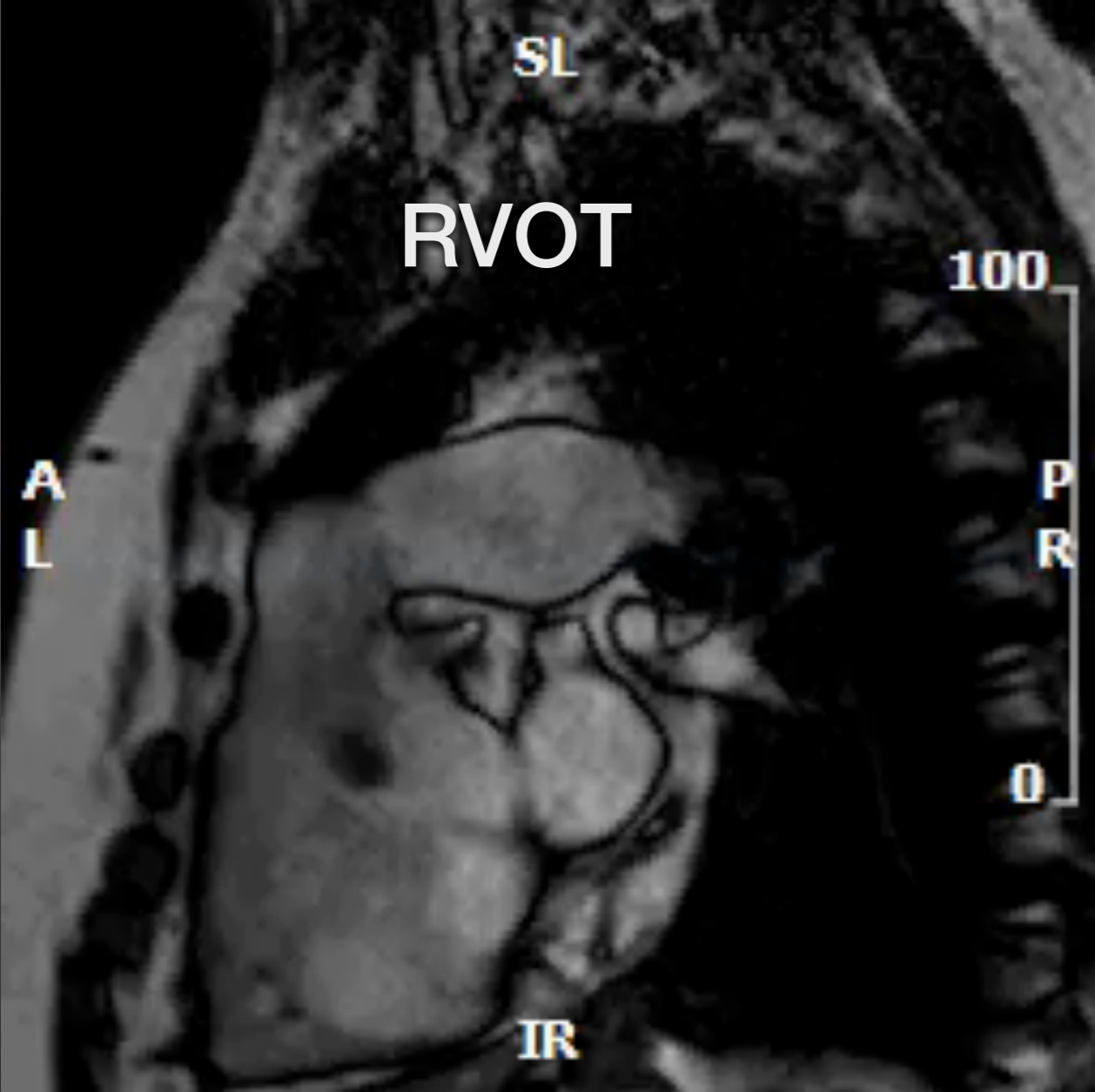


# Cardiac MRI



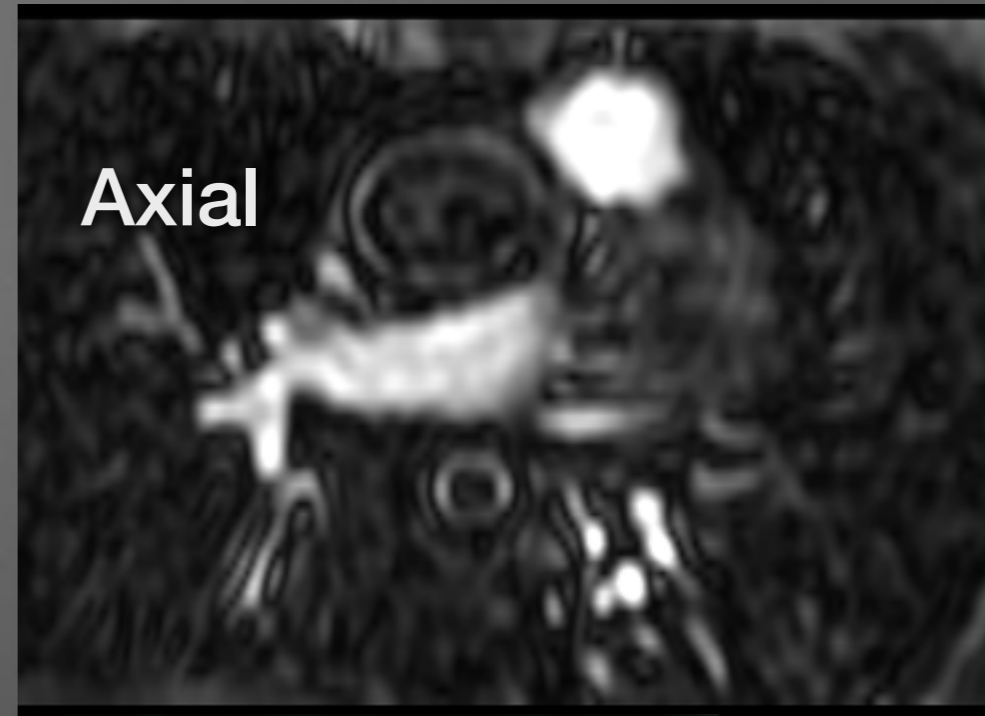
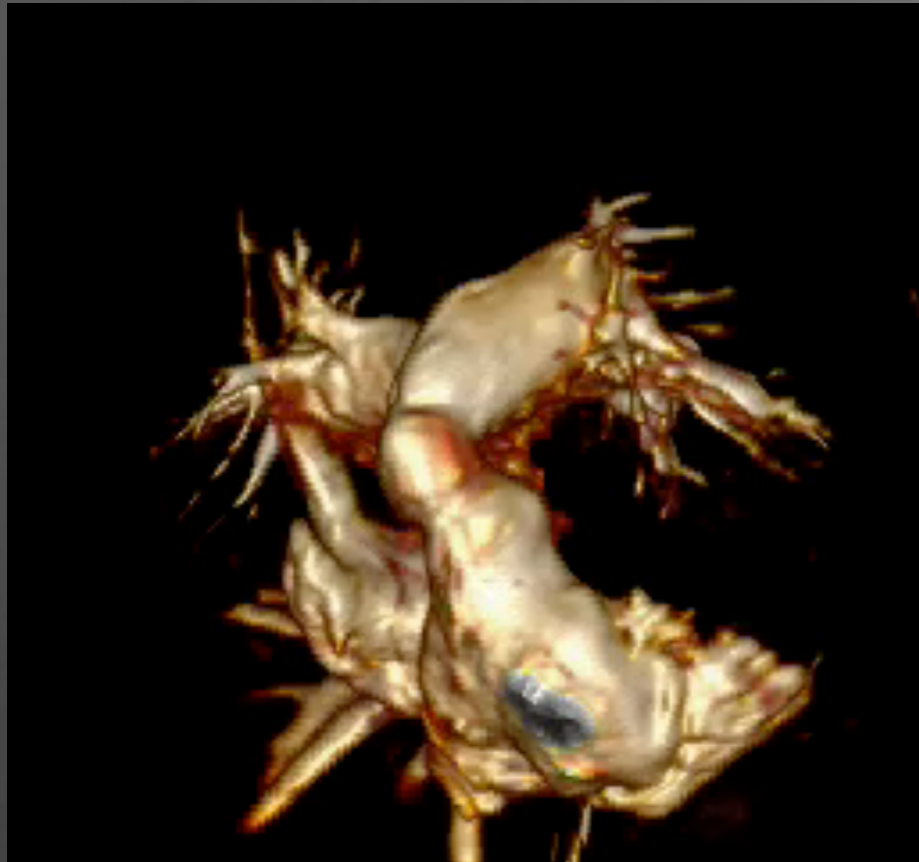
- Ventricular function
- LVEDV 54cc/m<sup>2</sup> LVESV 24cc/m<sup>2</sup>; EF 56%
- RVEDV 121cc/m<sup>2</sup> RVESV 68cc/m<sup>2</sup> EF 44%

# Cardiac MRI



Peak velocity 3.2m/sec

# MRA



# Pulmonary regurgitation

<Moderate PR

> Moderate PR

RVOTO >2/3  
systemic

Symptoms

No symptoms

No symptoms

- Severe PR
  - Progressive RV dysfunction (<47%)
  - RVEDVi/RVESVi >160/80 mL/m<sup>2</sup>
  - RVEDV > 2x LVEDV
  - Objective exercise intolerance

- Identify area of obstruction
- Progressive exercise intolerance
  - Arrhythmias

PVR



# Percutaneous



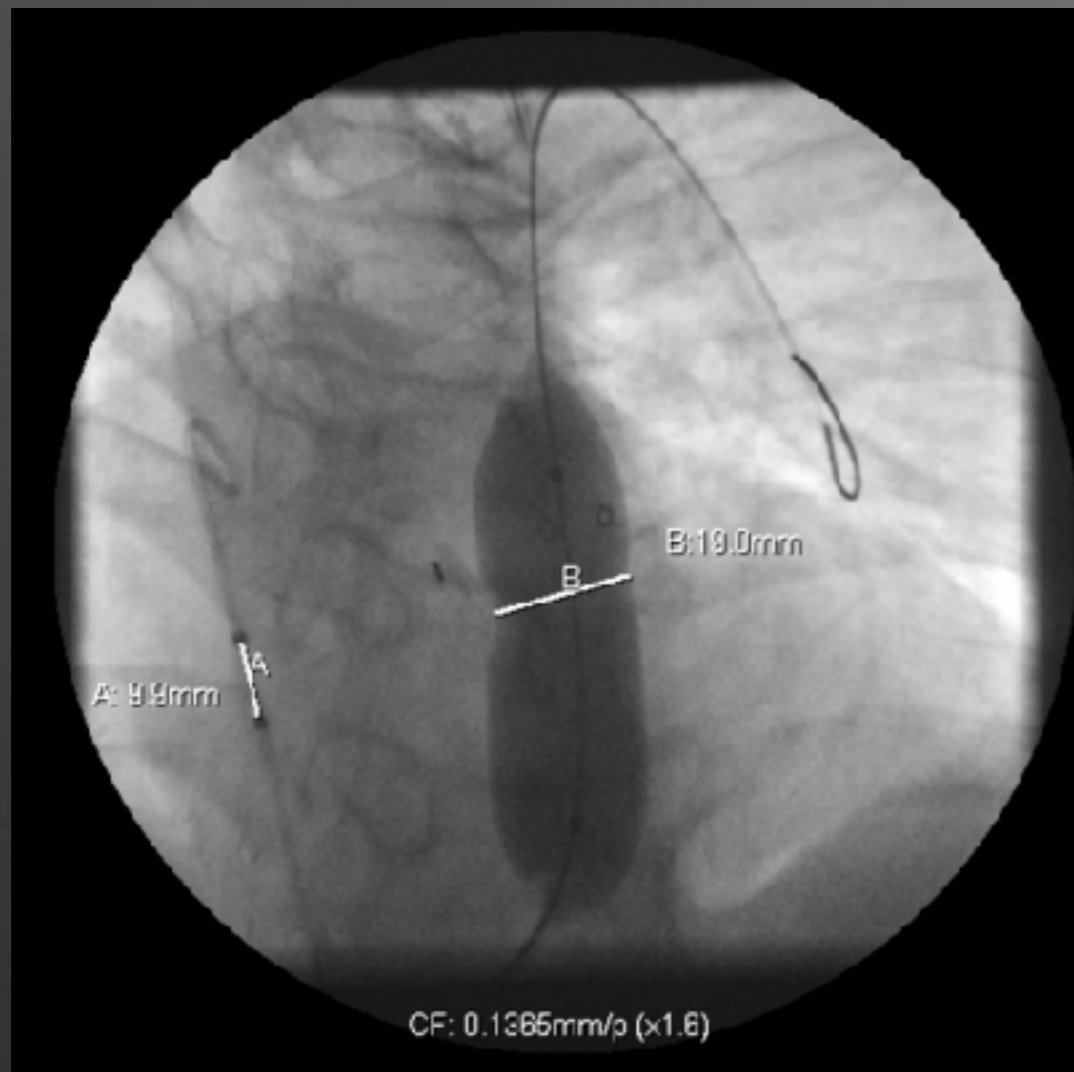
VALVE	TYPE	APPROVED USE	EXPANDABLE DIAMETER
MELODY	Bovine jugular venous valve in covered stent	RVOT conduits >16mm	20,22
SAPIEN	Bovine pericardial valve on stainless steel stent	Conduit >21mm	23,26
SAPIEN XT	Bovine pericardial valve on stainless steel stent	Aortic, mitral Native RVOT with Alterra anchoring adaptor	20.23.26.29
HARMONY TPV	Porcine pericardial valve on nitinol stent	Native RVOT	
VENUS-P	Porcine pericardial valve in covered self expanding stent	Investigational	

# Diagnosis and Plan

- Tetralogy of Fallot with RV to PA conduit dysfunction
  - Peak systolic conduit gradient 55mmhg
  - Moderate to severe PI
  - No pulmonary artery stenosis
- TPVR

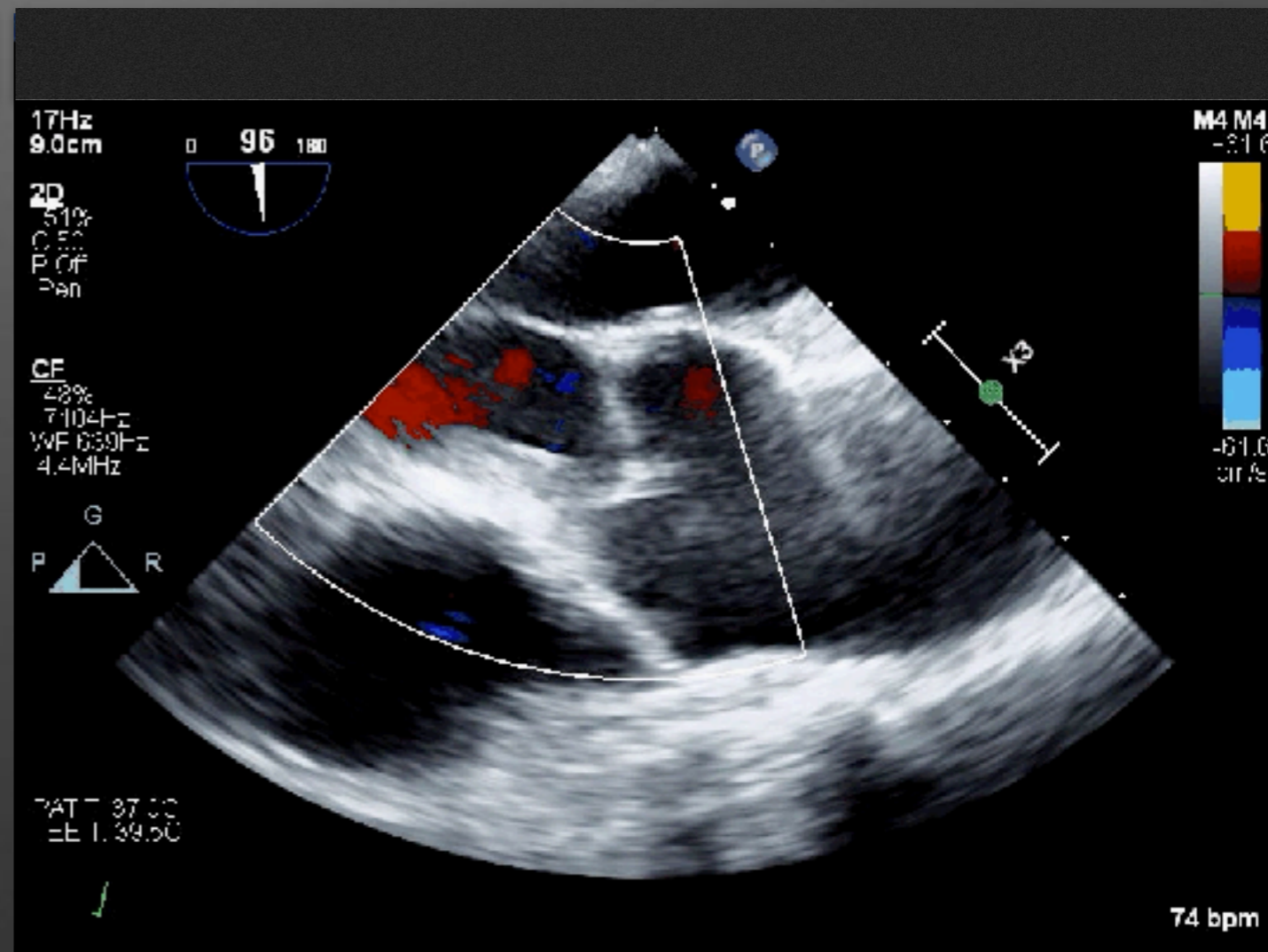
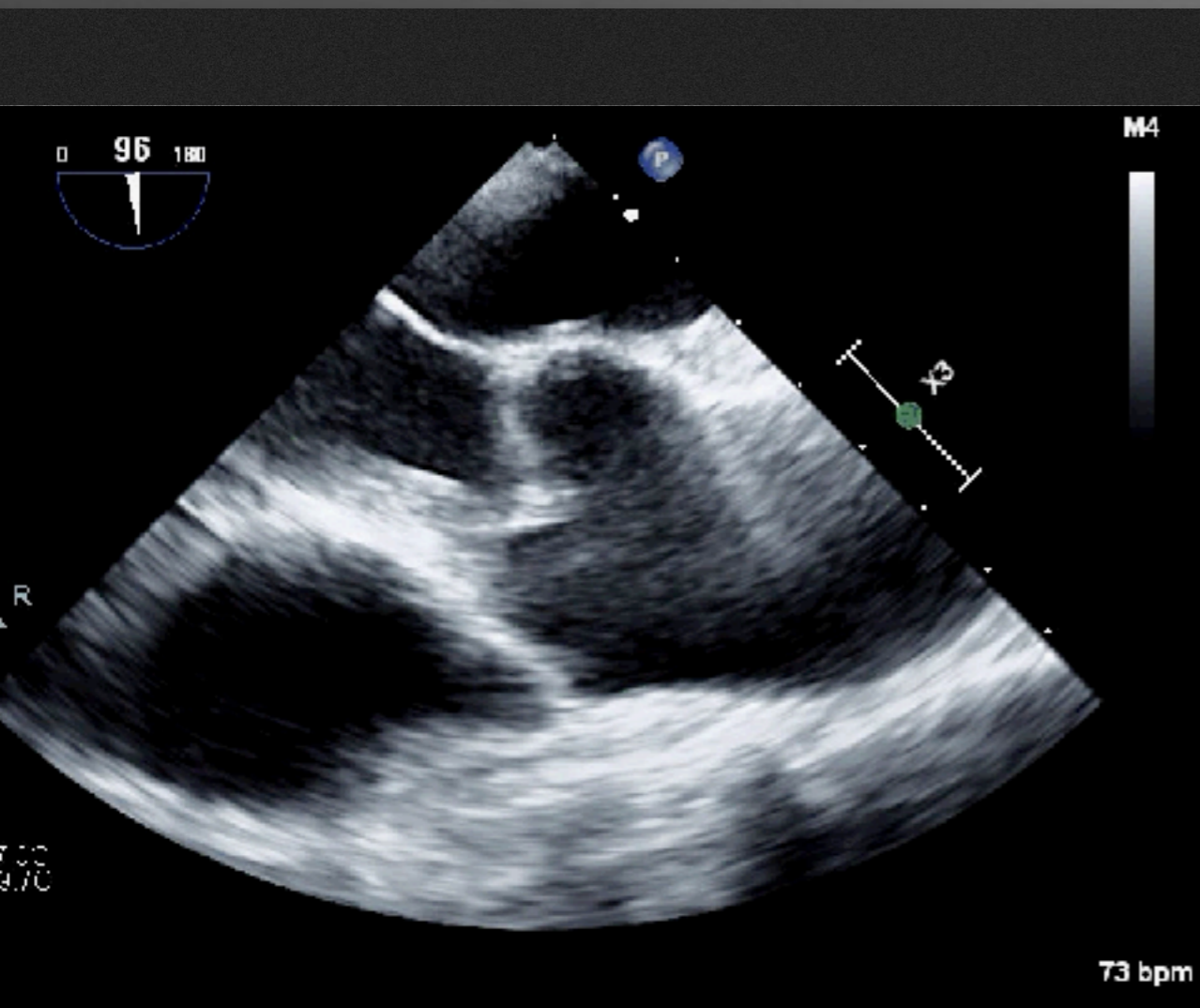


# Melody Valve



# Dilated Aorta ToF

- 50 year old female ToF right thoracotomy in infancy and sternotomy at age 5.

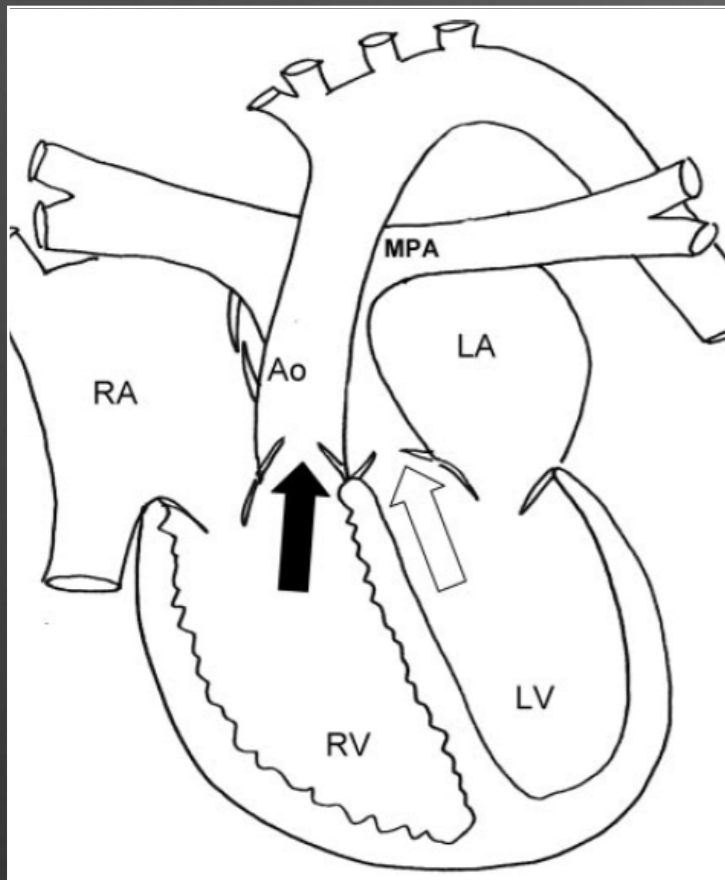


# Known complication of ToF

- Mean Aortic root  $43 \pm 6$  (echo),  $44 \pm 7$  (MPR) , 51%
- Mean ascending aorta  $42 \pm 8$  (echo) ,  $44 \pm 5$  (MPR), 69%
- $>50$  mm present in 9% of ToF
- 3% with  $>$  moderate AI
- $<10\%$  showed progression over an average of 9 years
- Dissection is very very rare.



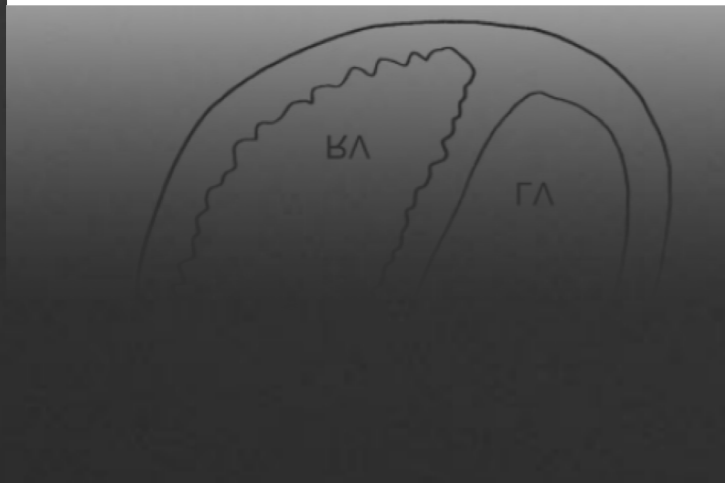
# D-TGA



## Multimodality Imaging Guidelines of Patients with Transposition of the Great Arteries: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society of Cardiovascular Computed Tomography

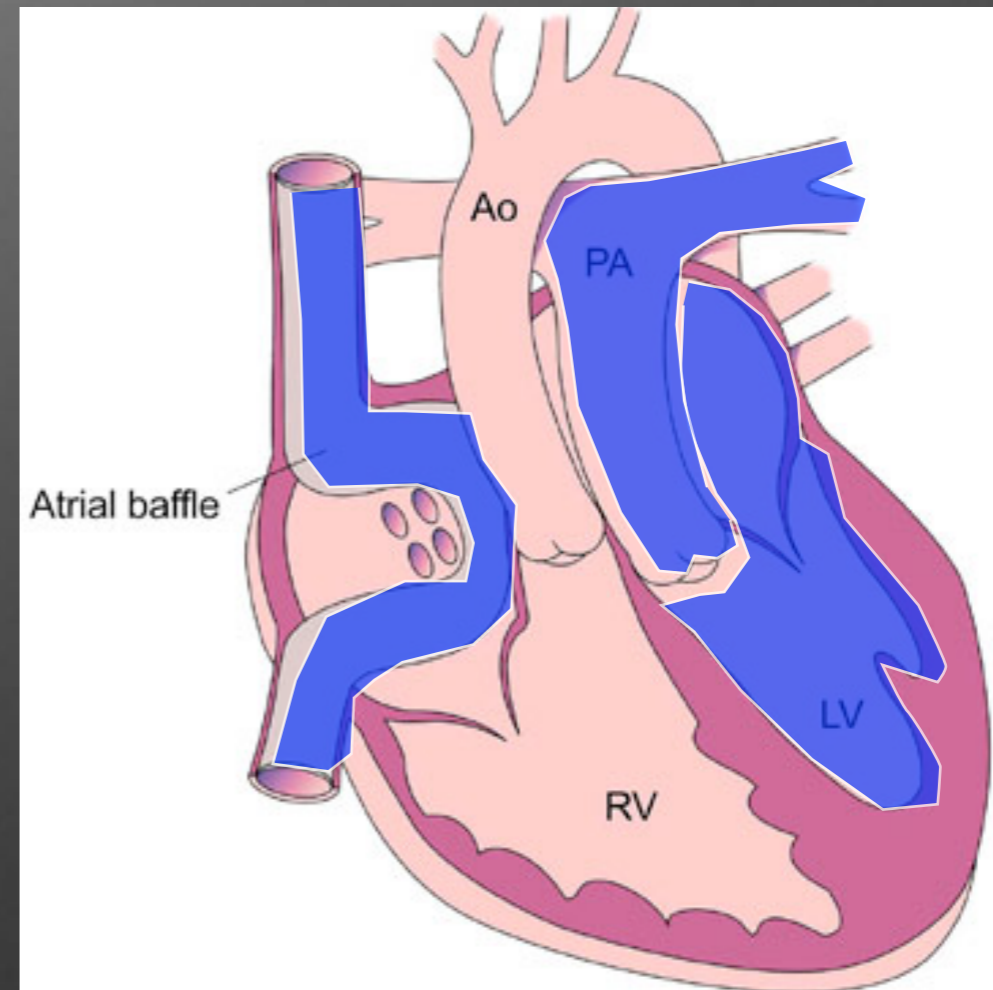
Meryl S. Cohen, MD, FASE (Chair), Benjamin W. Eidem, MD, FASE (Co-Chair), Frank Cetta, MD, FASE, Mark A. Fogel, MD, Peter C. Frommelt, MD, FASE, Javier Ganame, MD, PhD, FASE, B. Kelly Han, MD, Thomas R. Kimball, MD, FASE, Rebecca K. Johnson, RDCS, FASE, Luc Mertens, MD, PhD, FASE, Stephen M. Paridon, MD, Andrew J. Powell, MD, FASE, and Leo Lopez, MD, FASE, *Philadelphia, Pennsylvania; Rochester and Minneapolis, Minnesota; Milwaukee, Wisconsin; Hamilton and Toronto, Ontario, Canada; Cincinnati, Ohio; Boston, Massachusetts; and Miami, Florida*

(J Am Soc Echocardiogr 2016;29:571-621.)



# TGA - Atrial Switch (prior to 1980's)

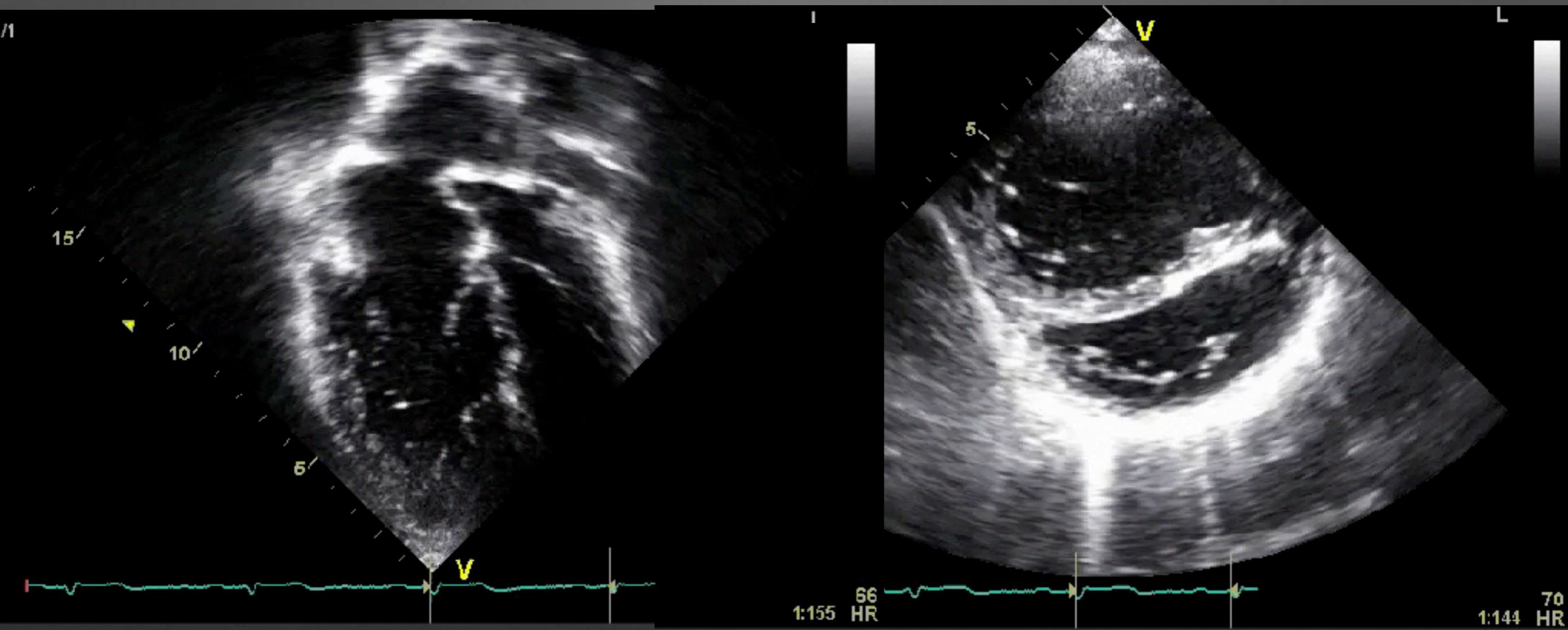
- Systemic RV failure
- Tricuspid Regurgitation
- Baffle leaks/obstructions
- Pulmonary hypertension



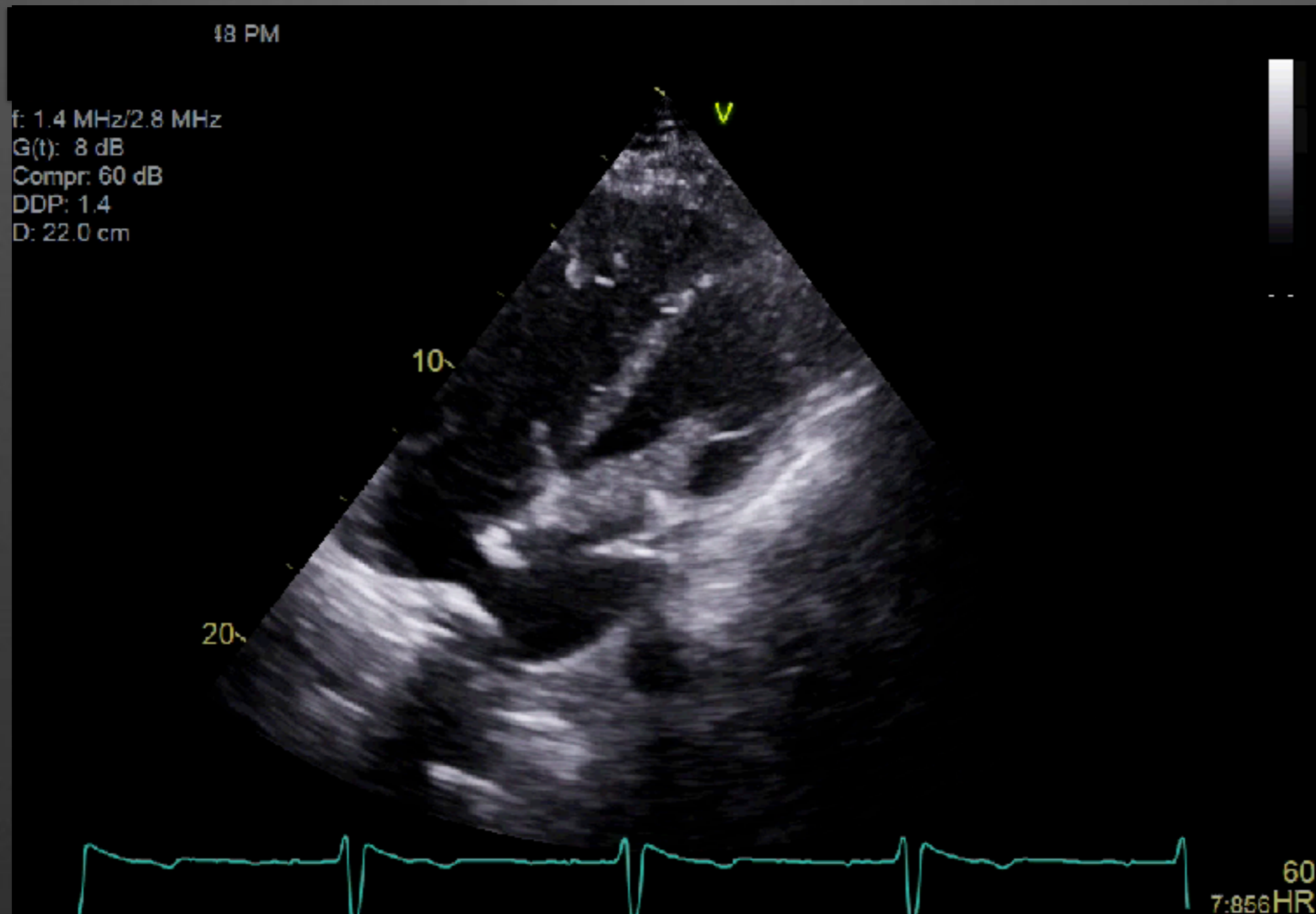
# Case 3 D-TGA- Atrial switch

- 32 y.o. male
- Who works construction presents with sudden onset of left sided weakness to his local ER and is sent to our ACHD center.
  - Vitals: BP 110/70; HR of 150 bpm; Pox 92%
  - Cardiovascular exam: RRR with systolic murmur; JVP of 10cm; HJR; +1 LE edema



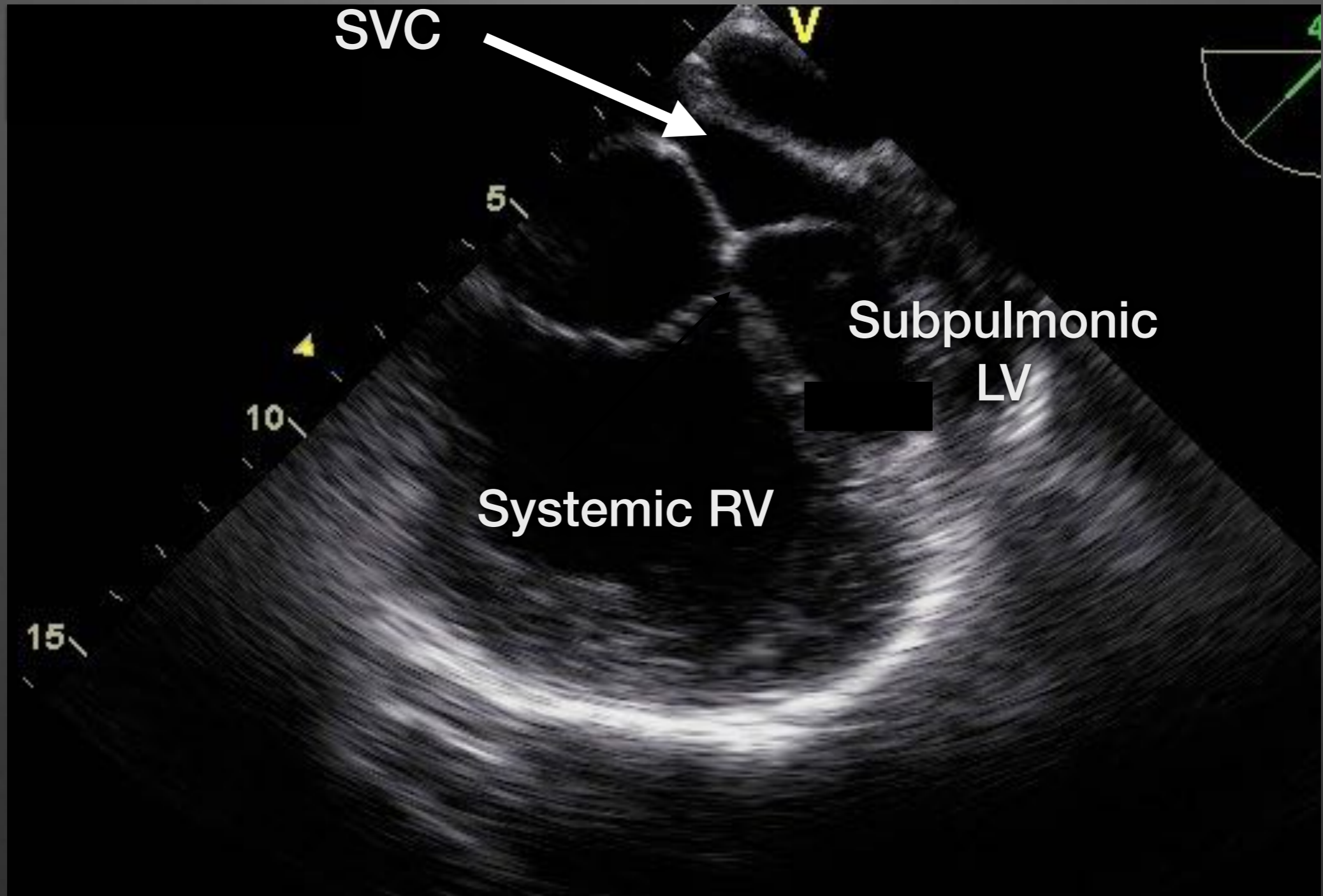


# Agitated Saline Injection

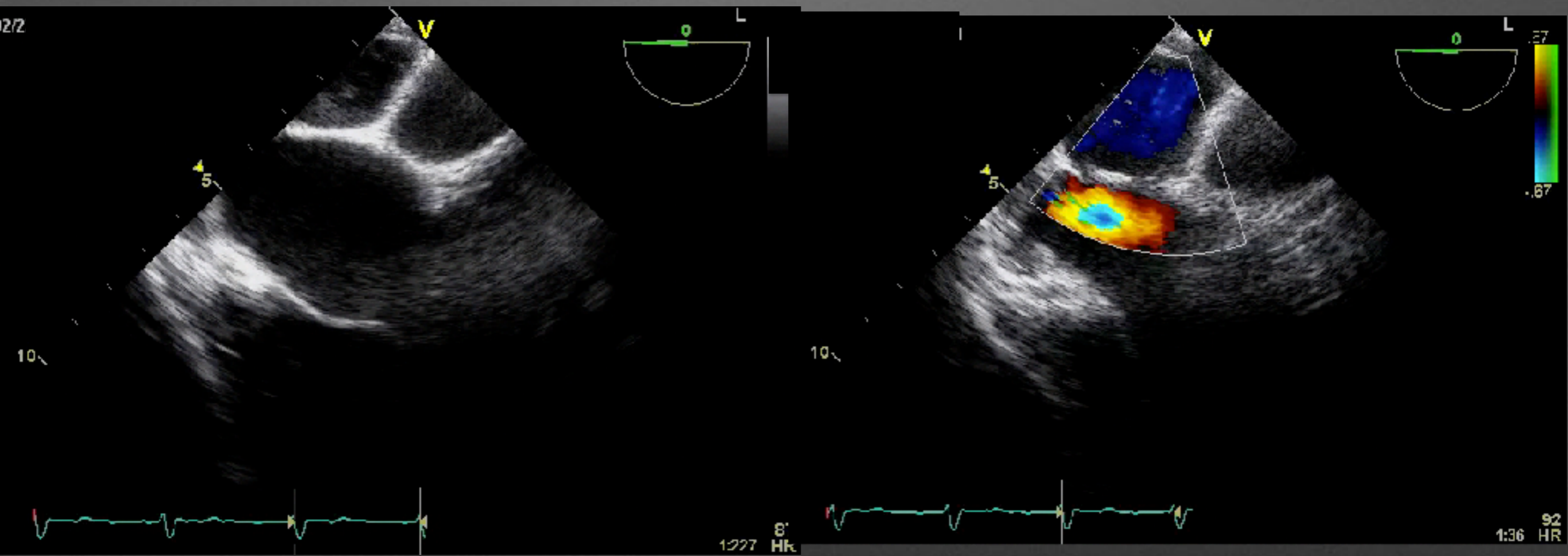




# TEE



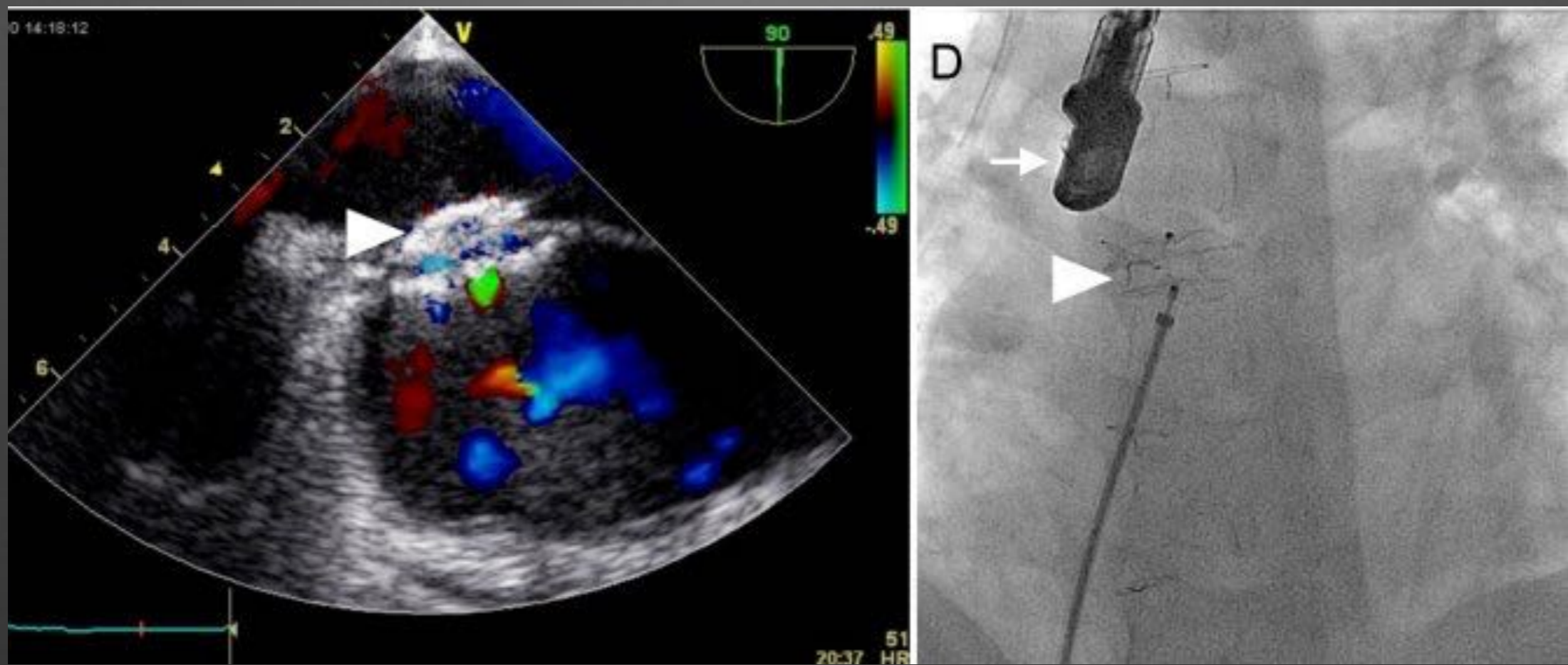
# TEE



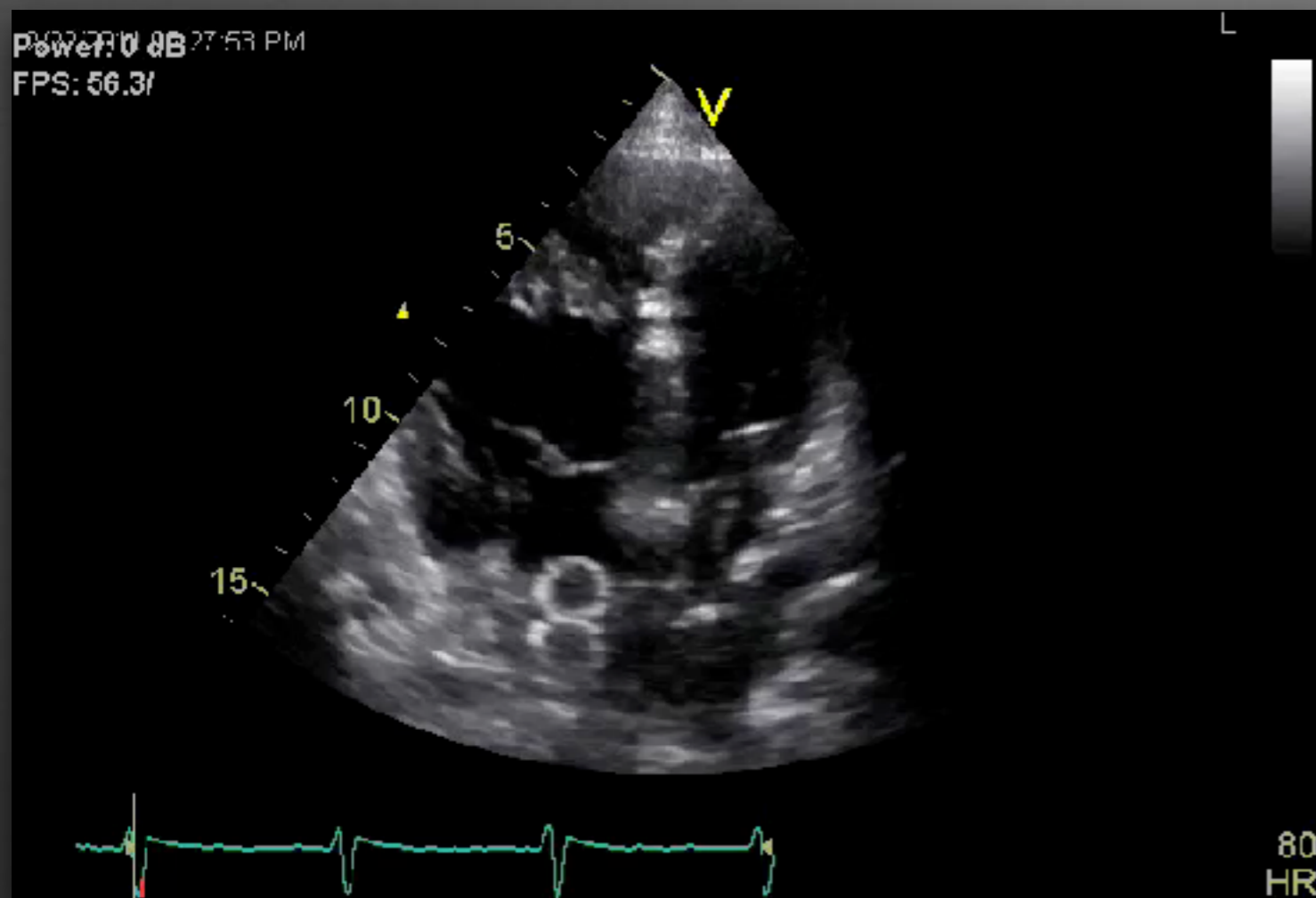
# Baffle Leak

- Saturations poor sensitivity for detection of baffle leak
- Incidence of baffle leak of 50-75%
- Especially important if desaturation with exercise or cardiac lead placement
- Agitated Saline injection superior to MRI for evaluation of baffle leak

# Percutaneous Intervention



# Diagnosis and Plan

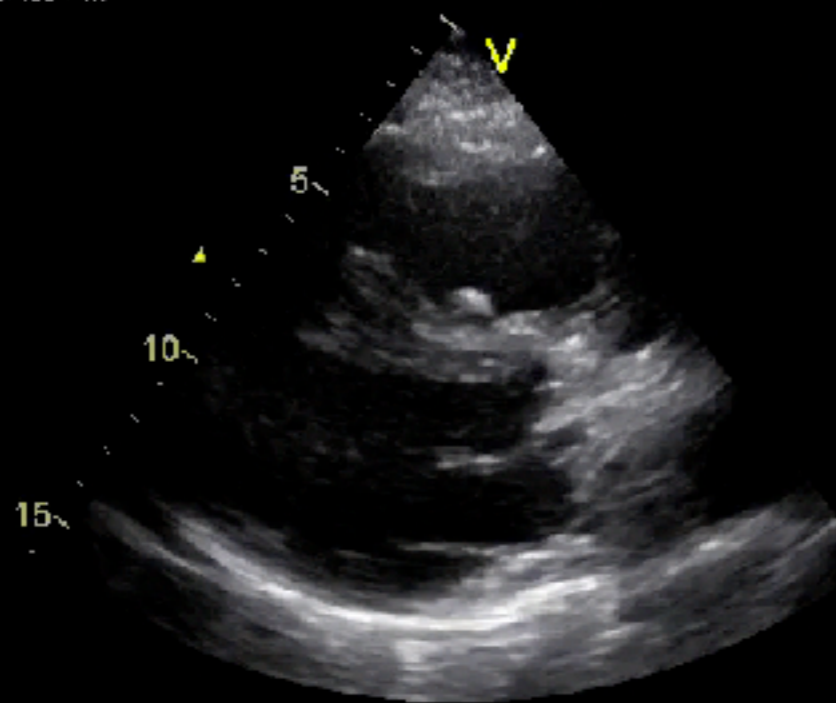


- D-TGA with atrial switch with baffle leak
- Percutaneous Amplatzer Closure

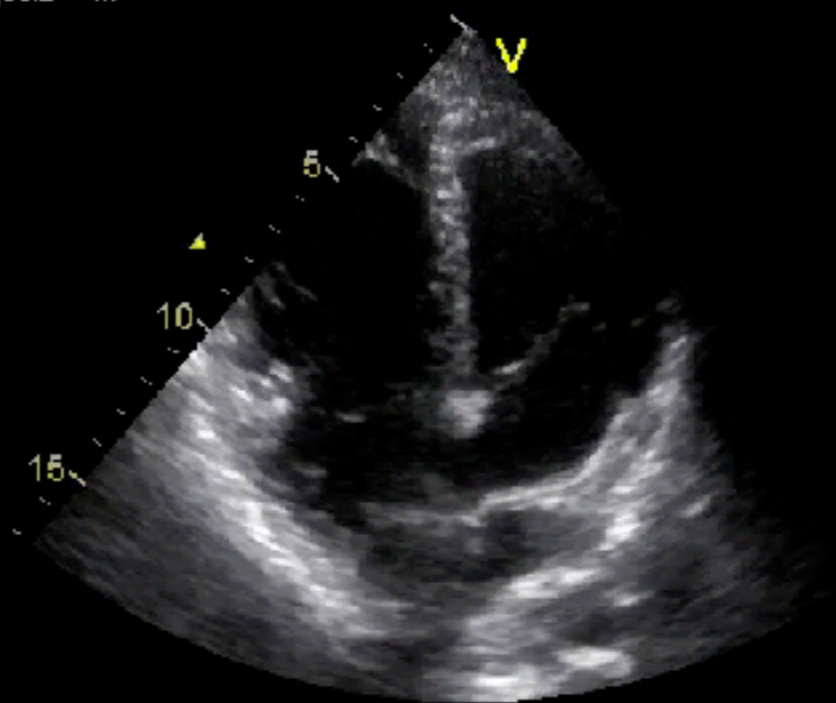
# Case 4

- 32 y.o. D-TGA Atrial switch complains of progressive dyspnea on exertion
- She has two children 3 and 6 years old
- BP 100/70 P 80 Pox 80%
- Cardiovascular: RRR 3/6 systolic on RSB and 2/6 at the mid clavicular line. JVP is 10cm

Power/D 08:07:05 AM  
FPS: 56.3

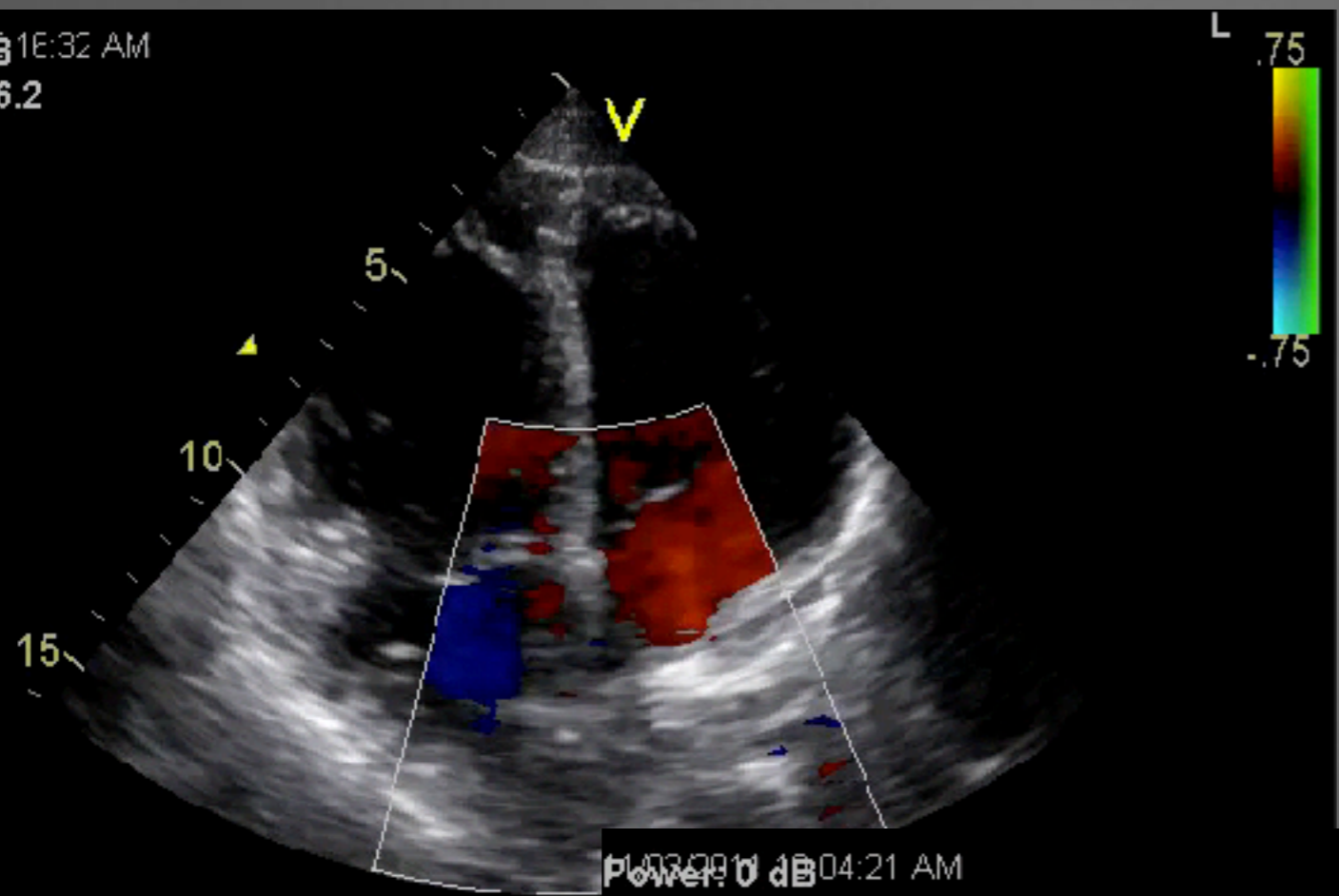


Power/D 08:07:21 AM  
FPS: 52.9

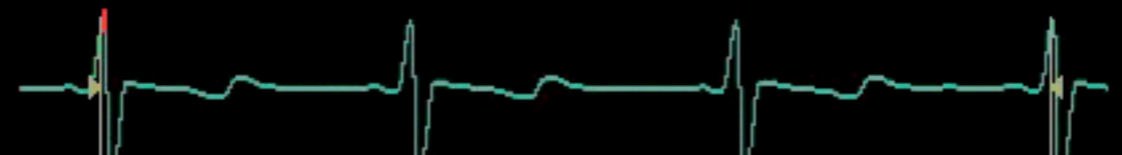
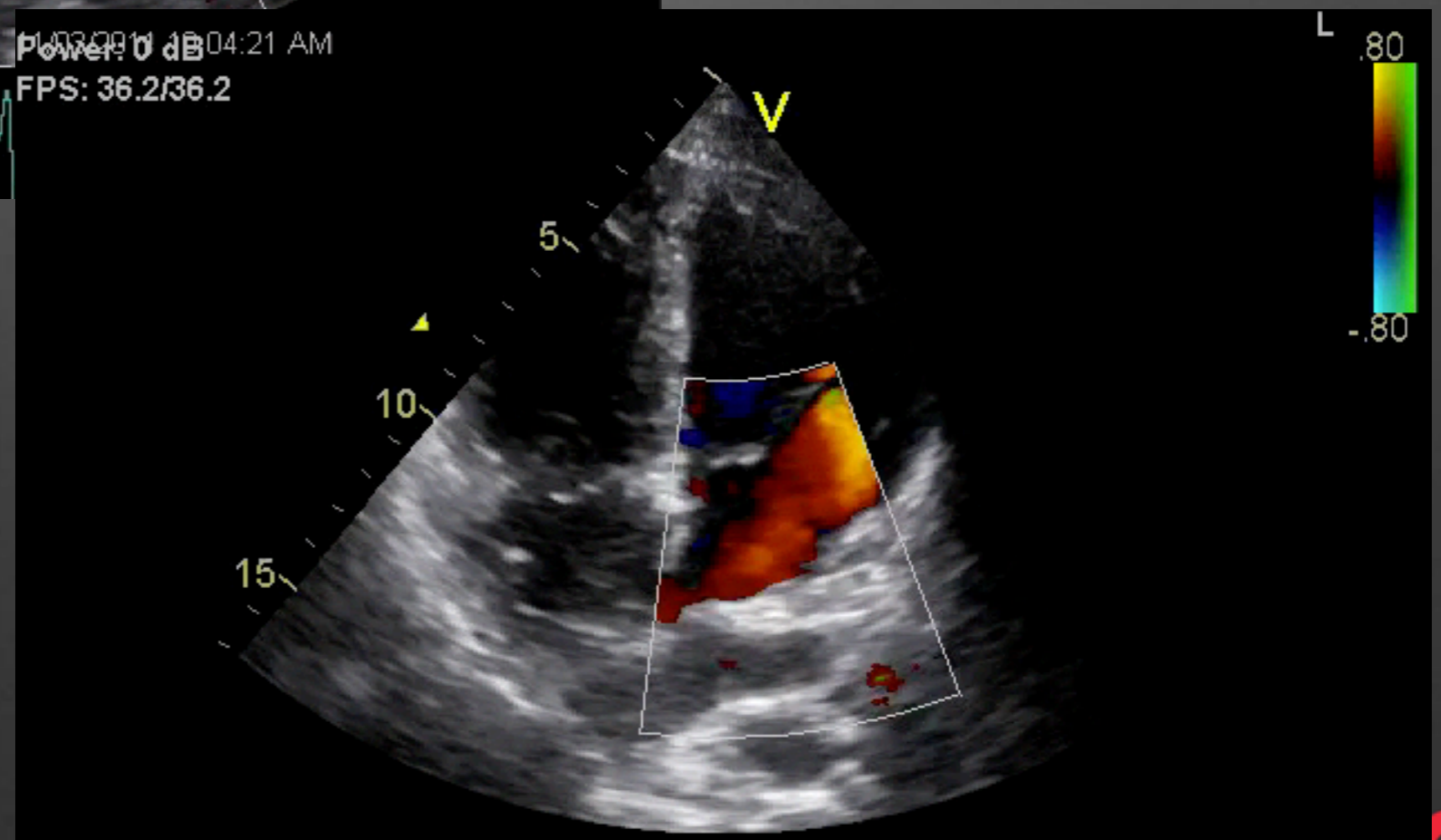


64  
HR

Power: 0 dB  
16:32 AM  
FPS: 26.2/26.2



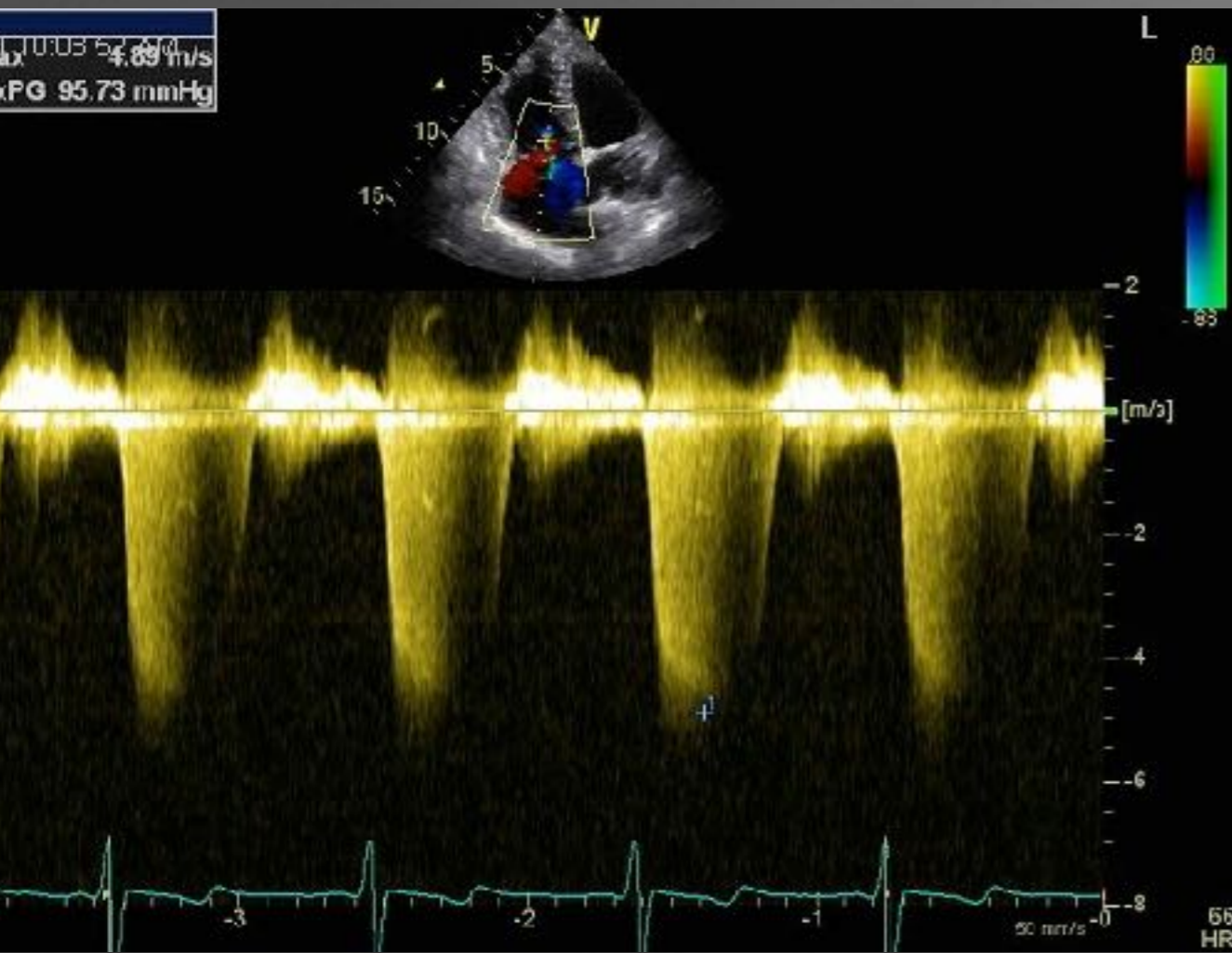
Power: 0 dB  
04:21 AM  
FPS: 36.2/36.2



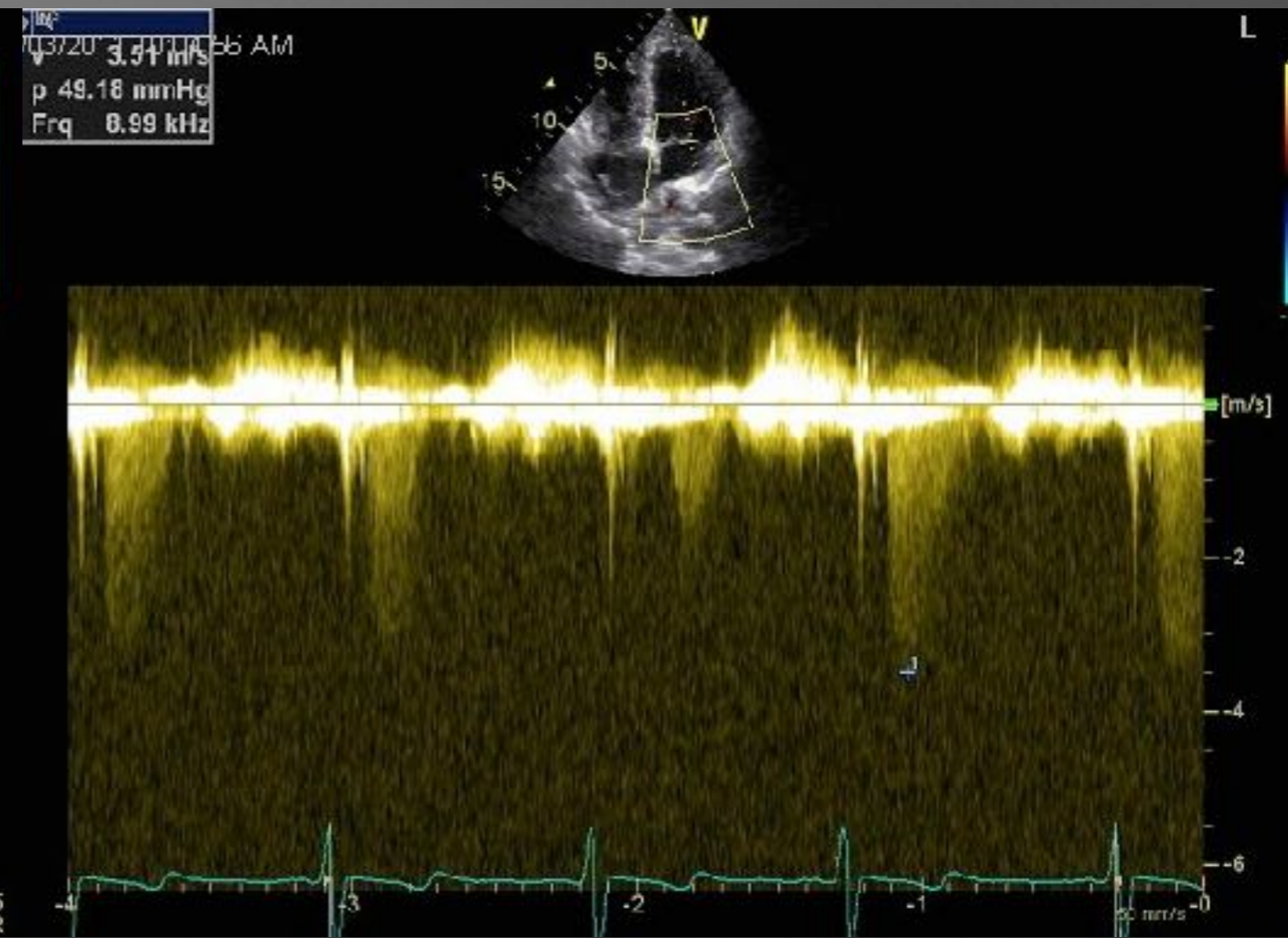
67  
HR







**Systemic TR - RV pressures of 95mmHg +**



**Subpulmonic MR - LV pressures of 50mmHg +**



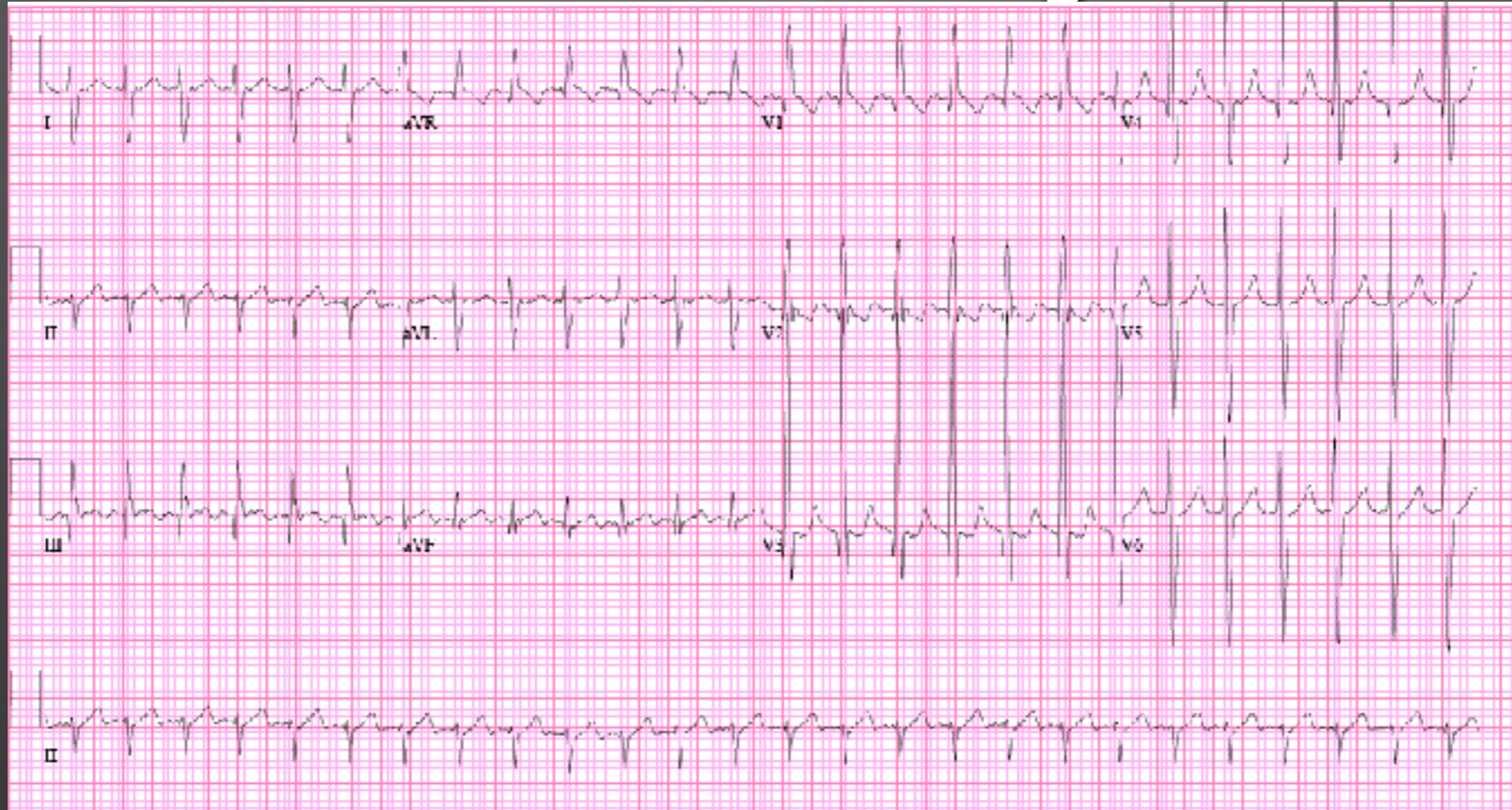
# Diagnosis and Plan

- D-TGA with atrial switch - pulmonary hypertension and large baffle leak
- Surgical correction of baffle leak

# Case 5

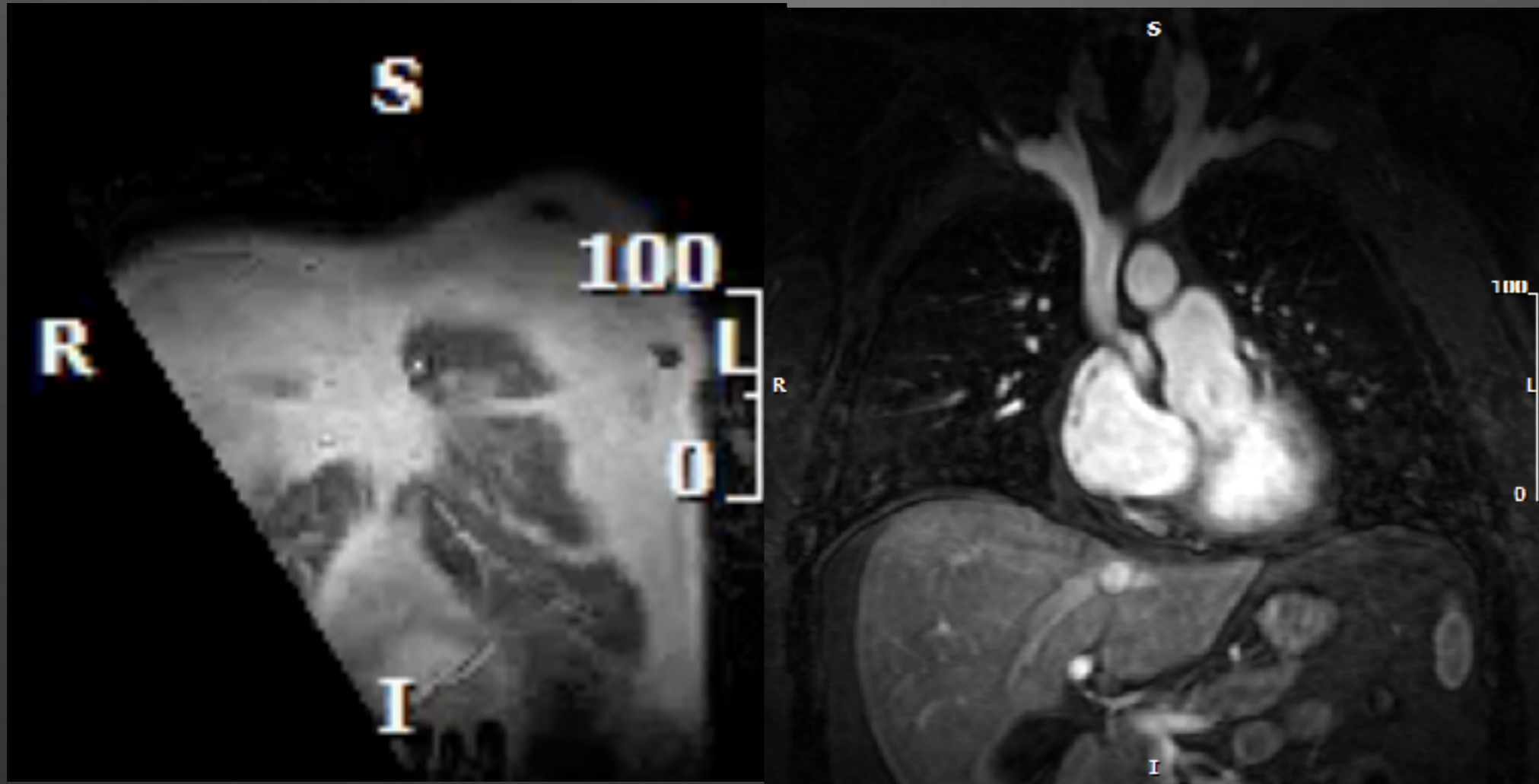
- 35 y.o. male with D-TGA with atrial switch with symptoms of palpitations associated with decreased exercise tolerance

# Intra-atrial Tachycardia

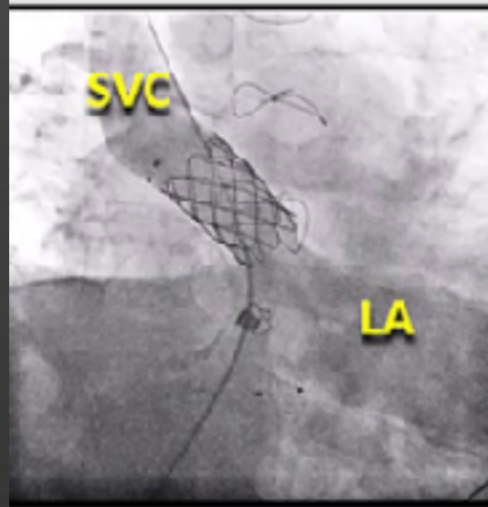
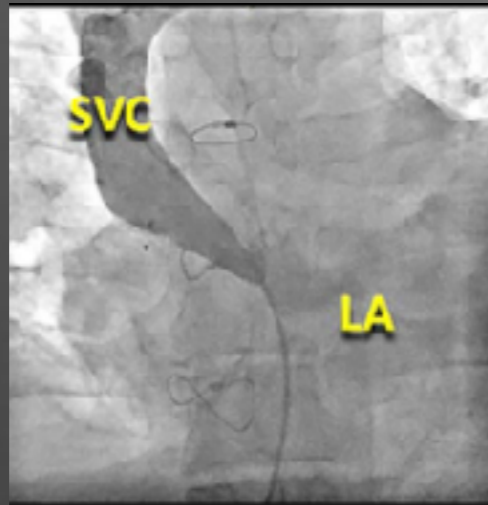


Has a TEE and undergoes DCCV  
Develops 4-6 second sinus pauses

# MRA

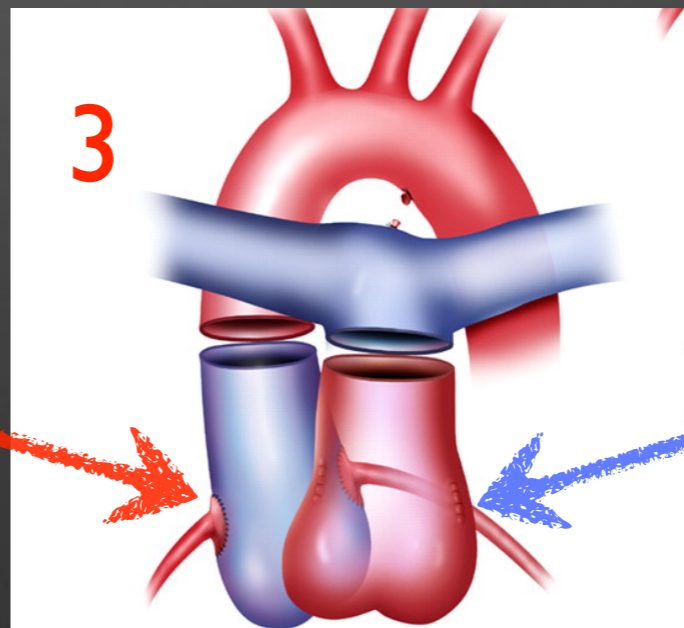
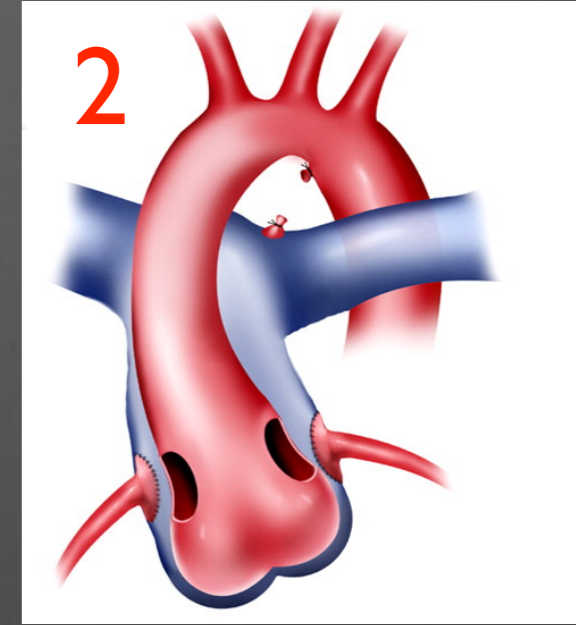
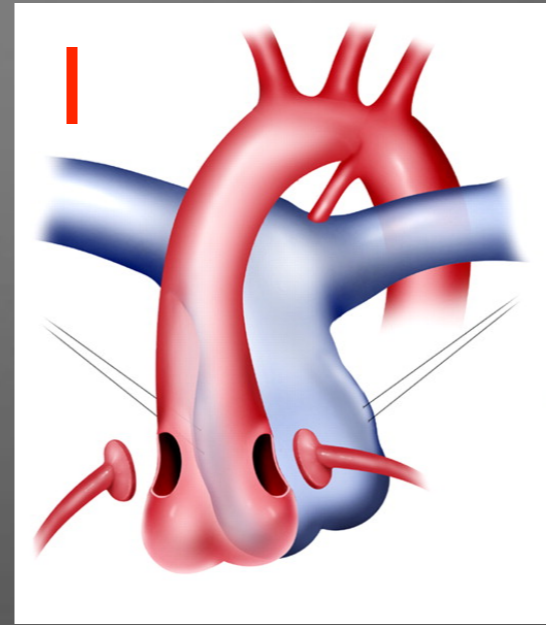
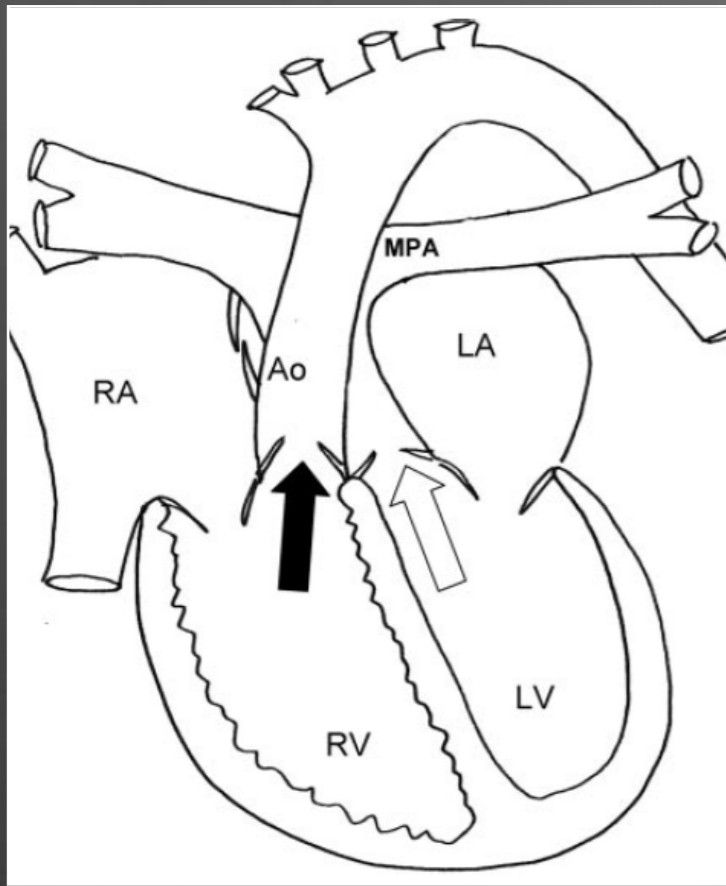


# Percutaneous Stenting



- 1/3 D-TGA atrial switch develop baffle obstruction
- Recommend Stenting
  - mean gradient of 6mmHg
  - <10mm diameter
  - prior to device placement

# D-TGA: Arterial Switch



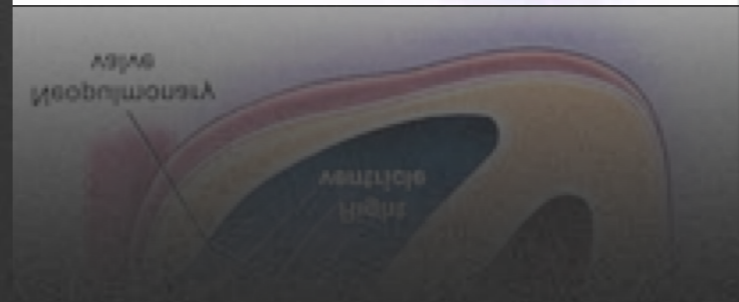
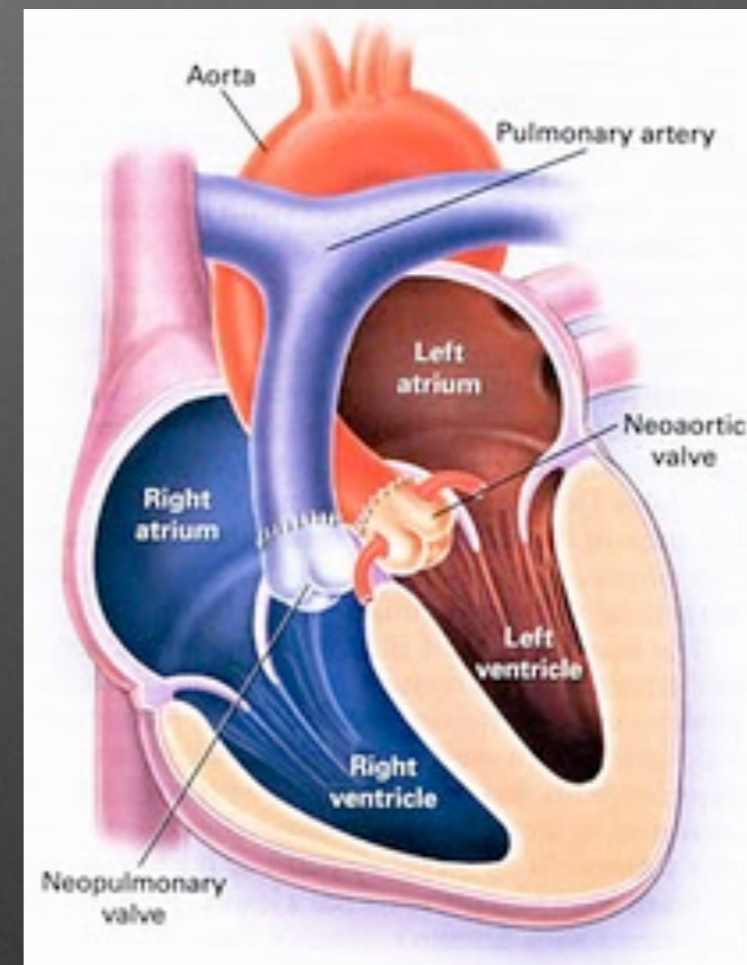
Neo aorta: proximal PA  
and pulmonic valve

Neo pulmonary: proximal  
aorta and aortic valve



# D- TGA with Arterial Switch (after 1980's)

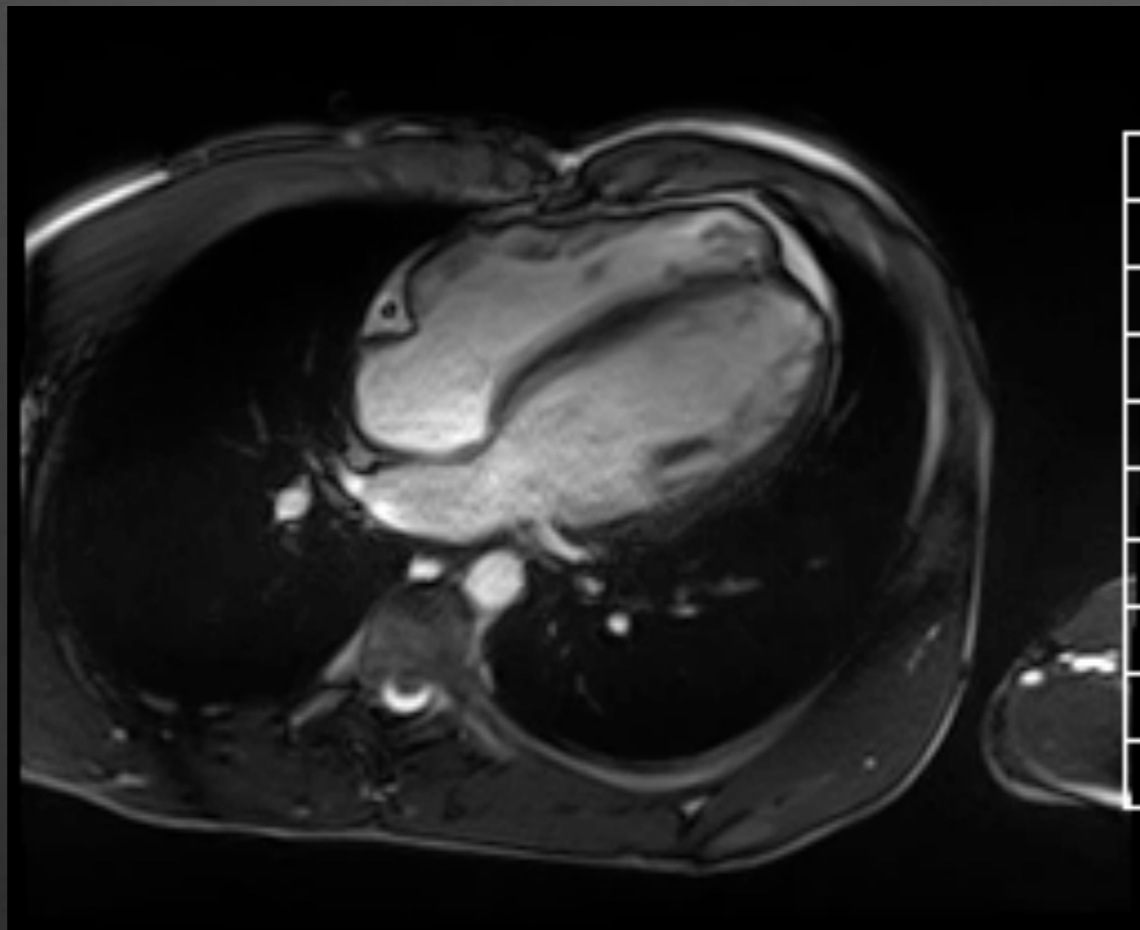
- Branch and PA stenosis
- Neo-aortic dilation
- PV and AV regurgitation
- Ostial coronary stenosis

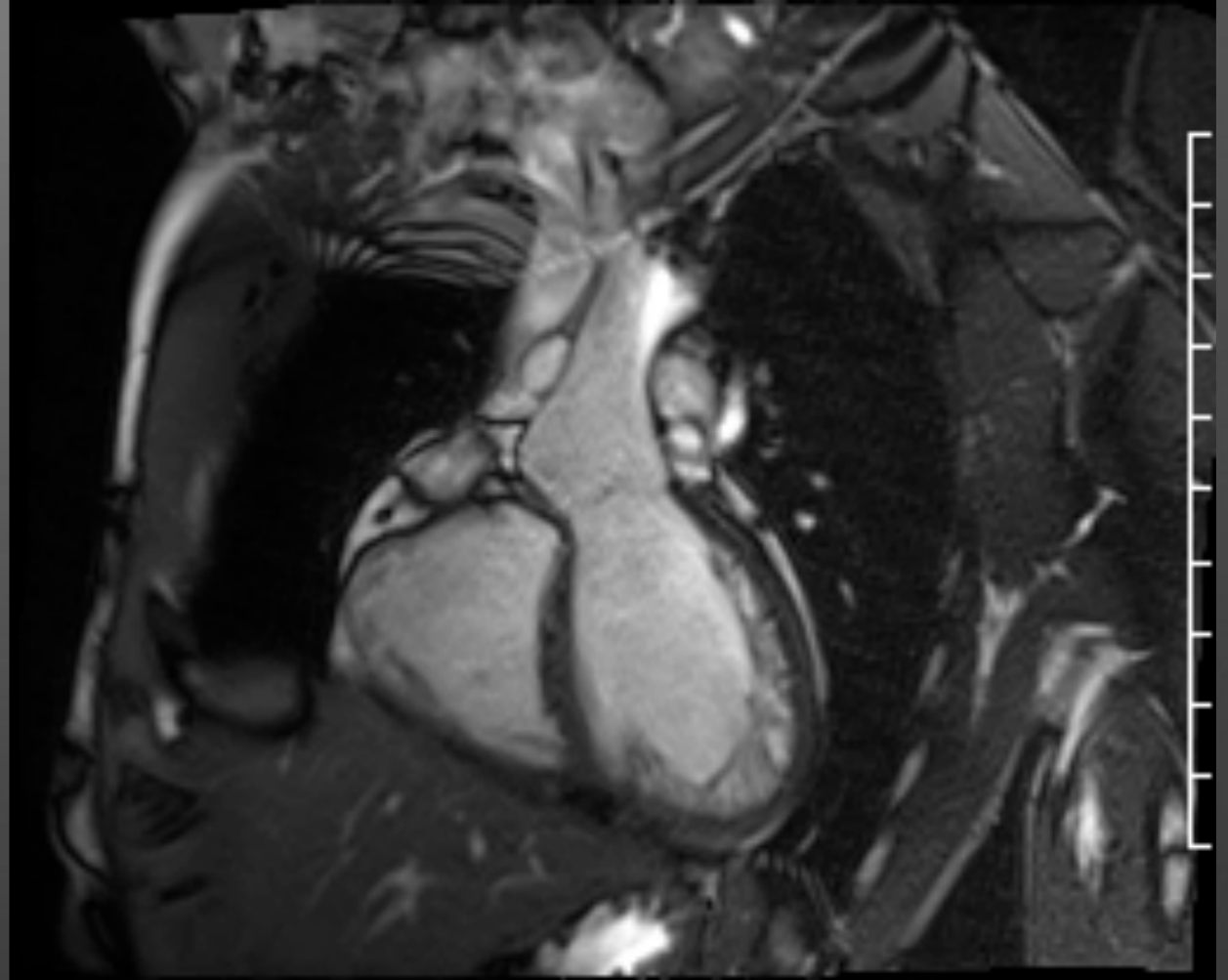
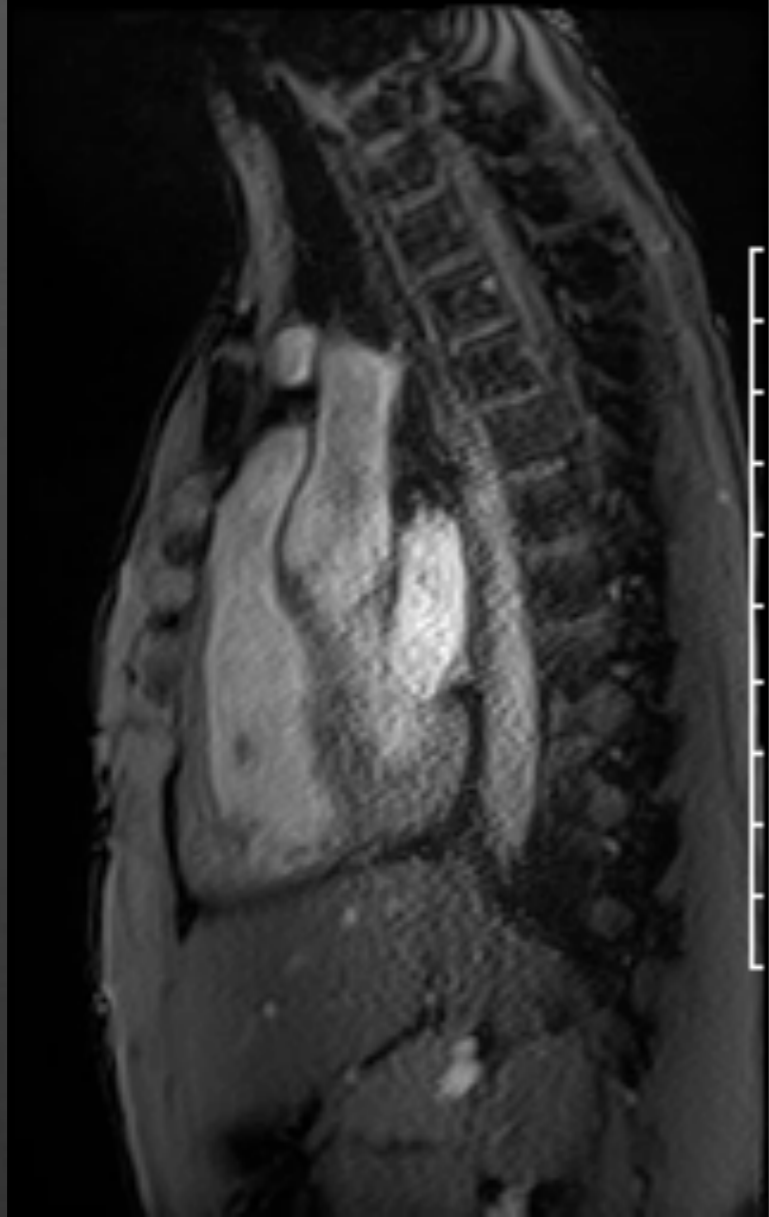




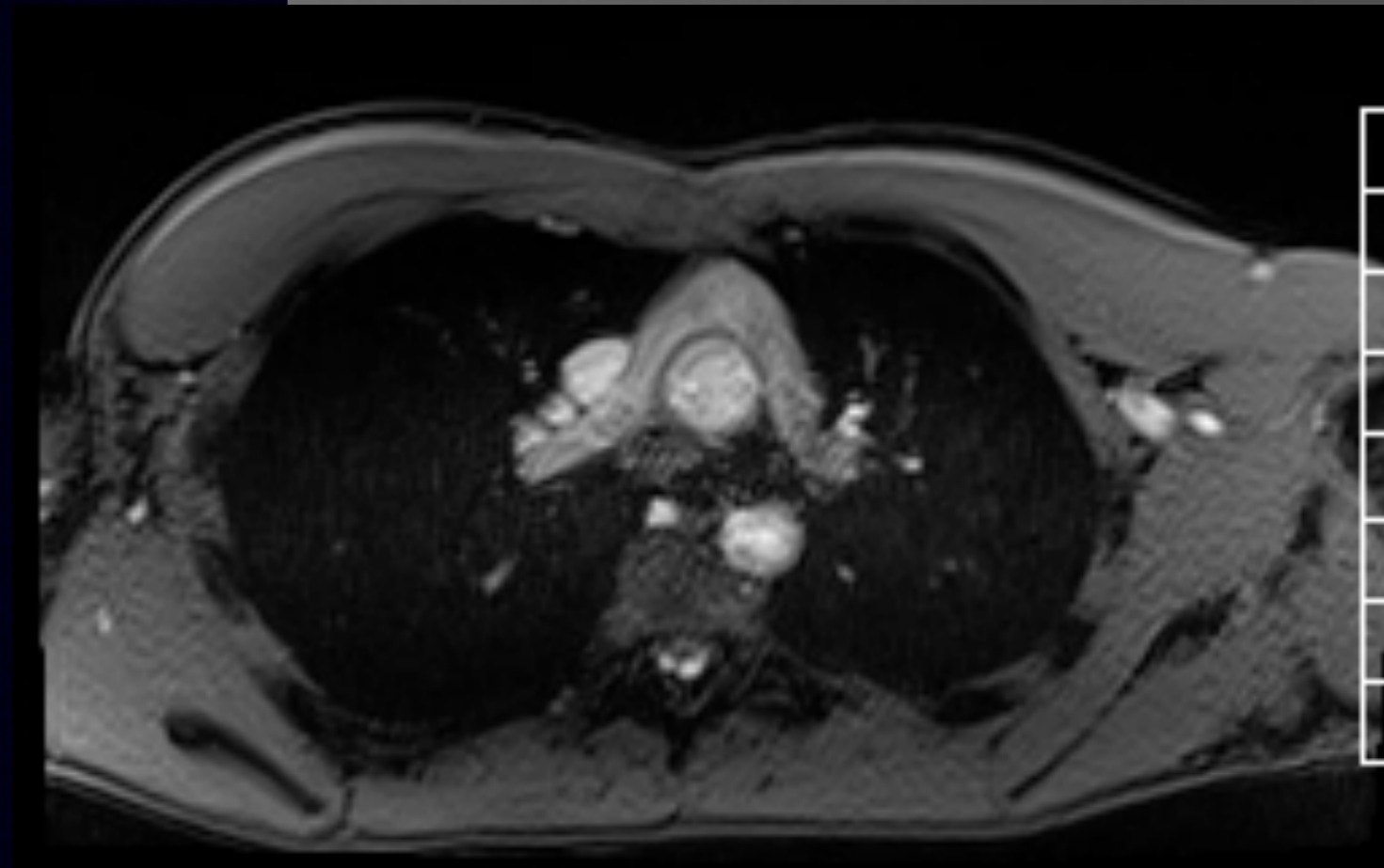
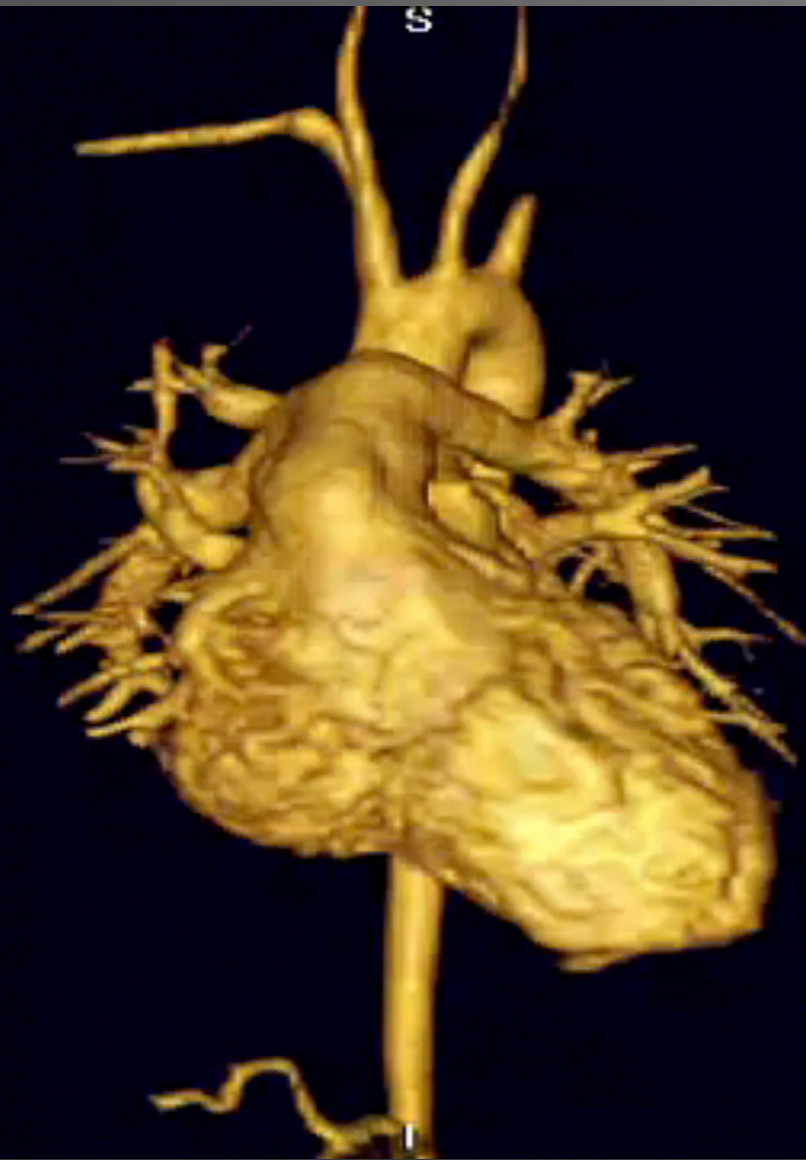
# Case 6

- 19 y.o. male who DTGA with arterial switch presents for routine visit
- Normal vitals 3/6 systolic mumur LSB



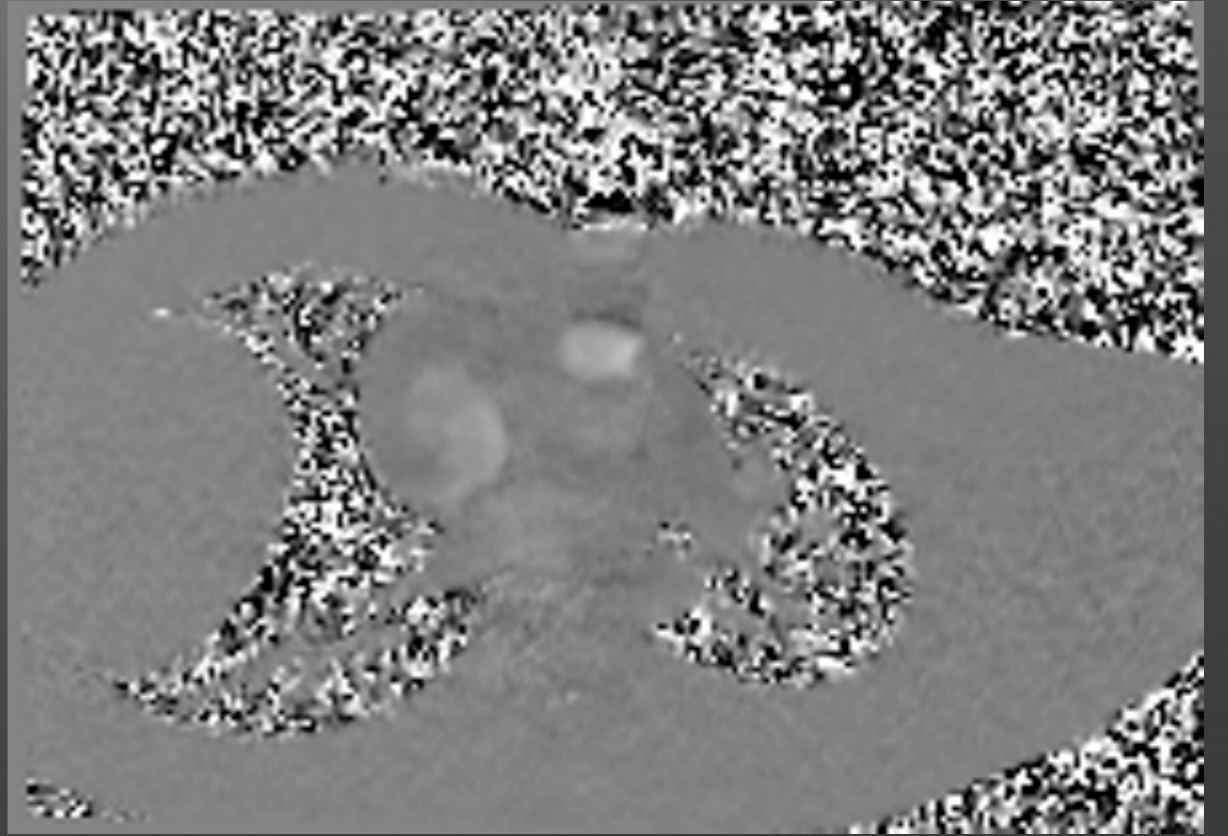
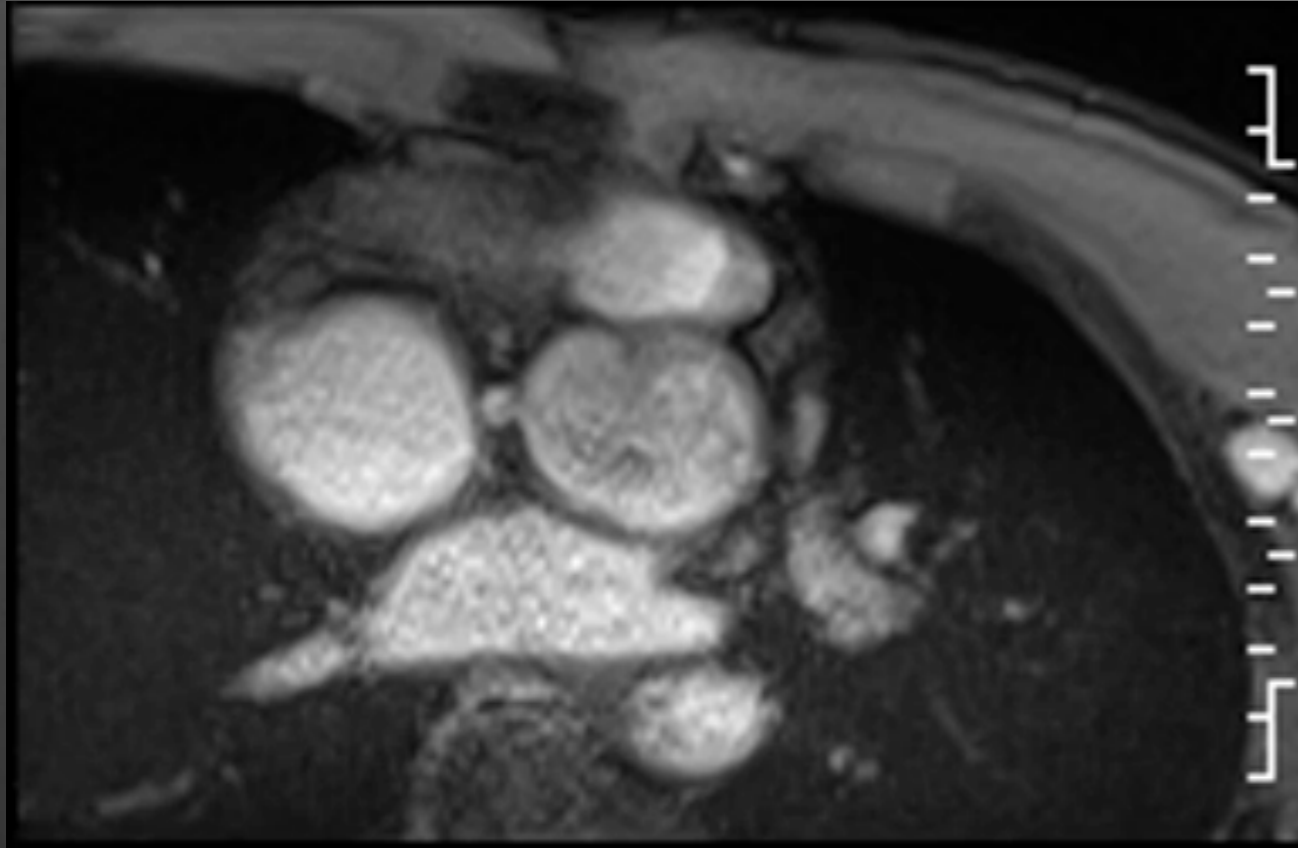


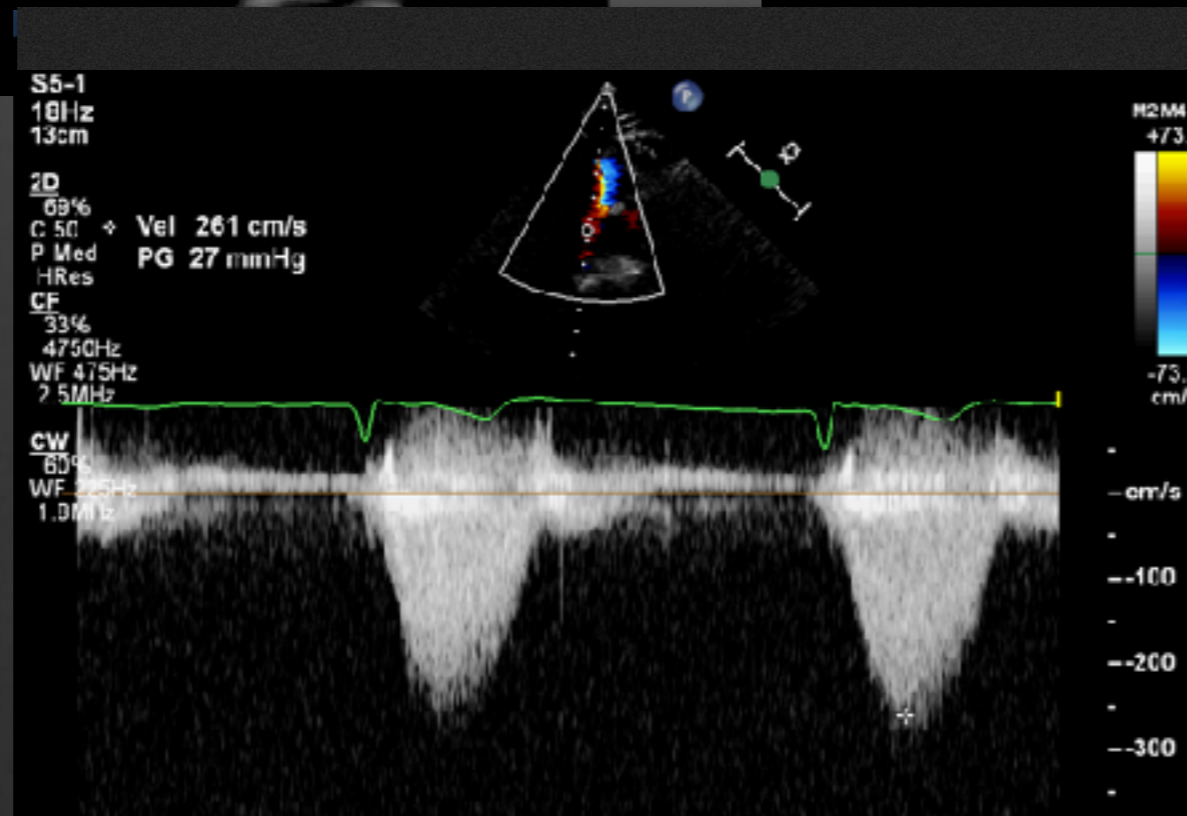
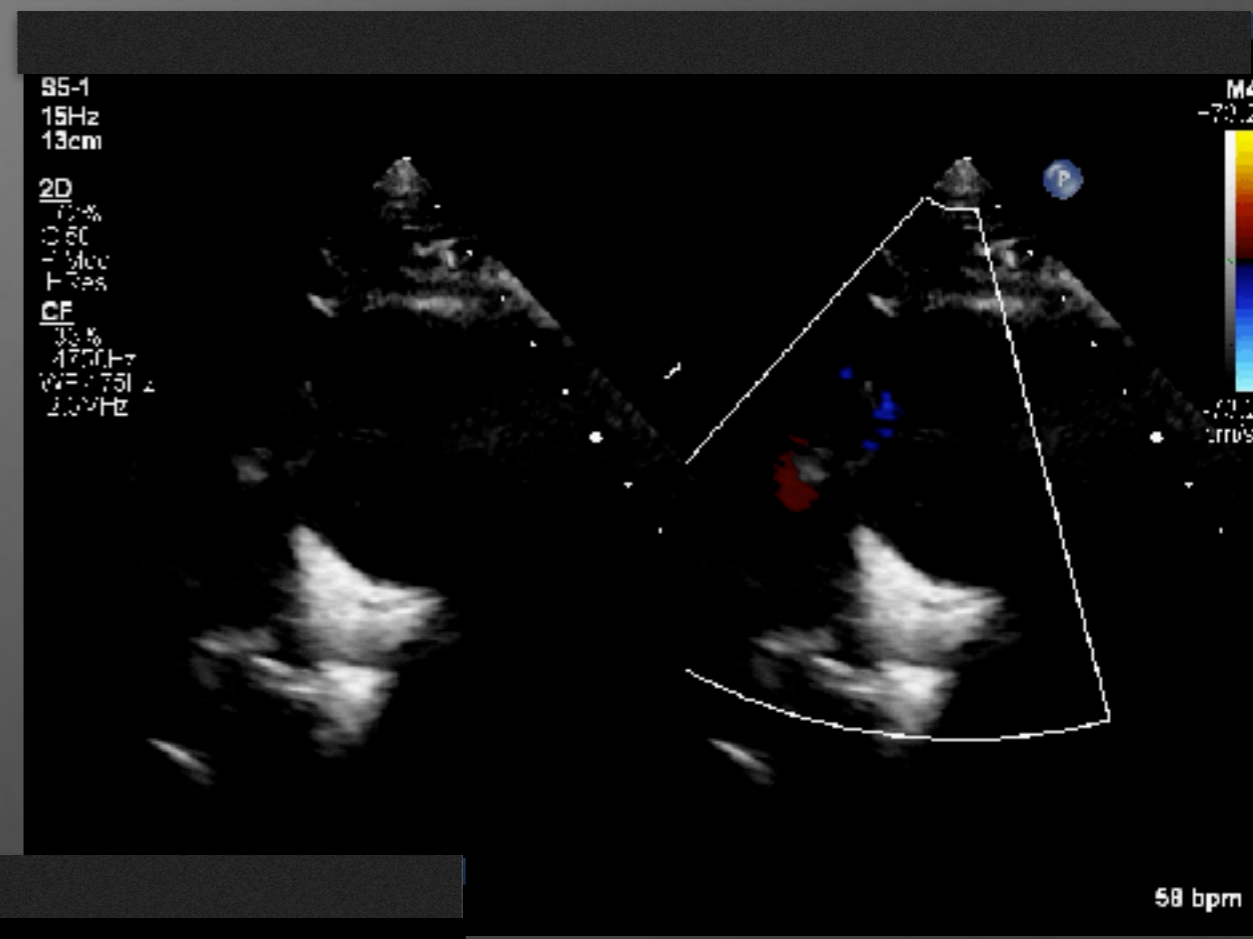
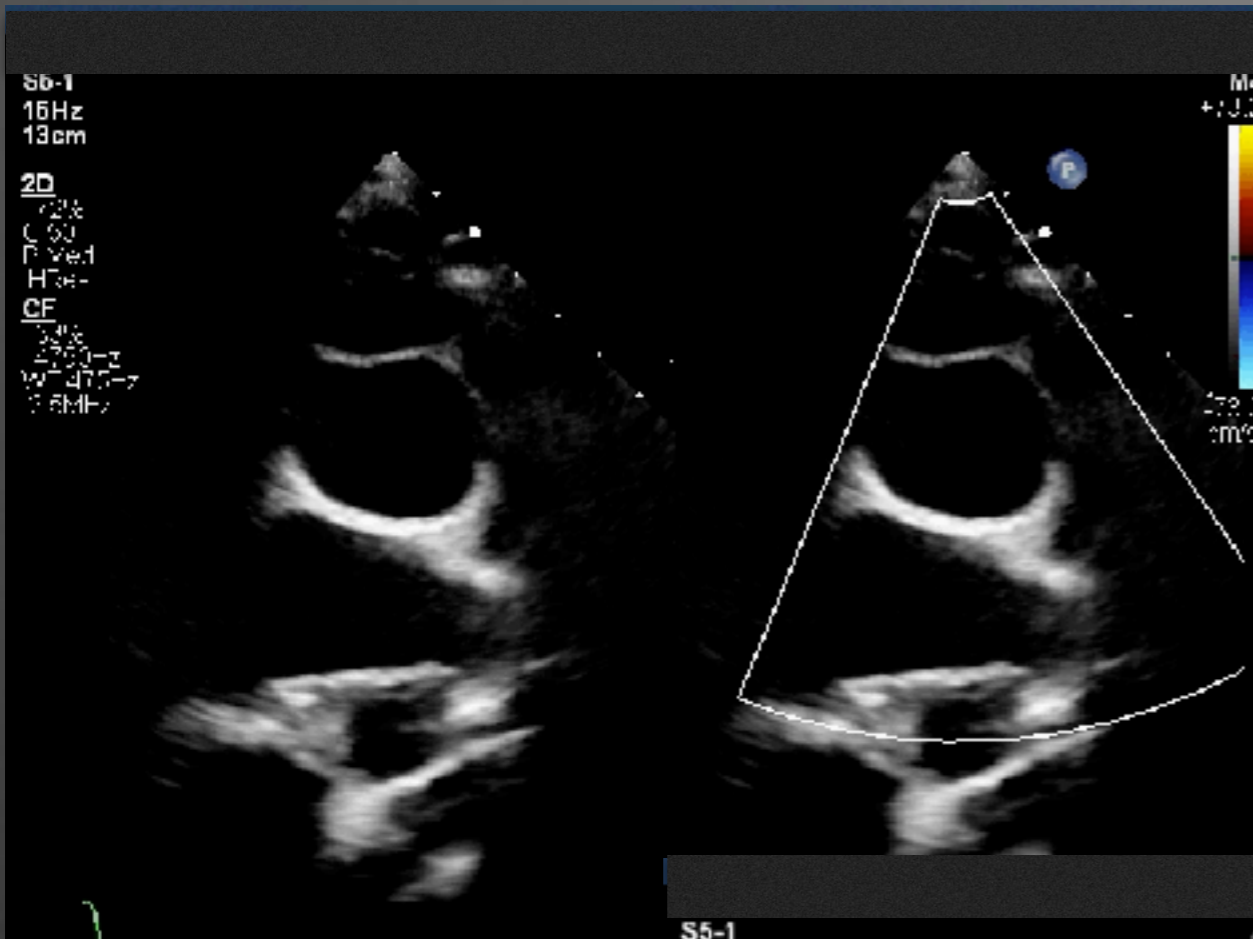
# Arterial switch Lecompte Procedure



W/L: 144/77  
Segmented





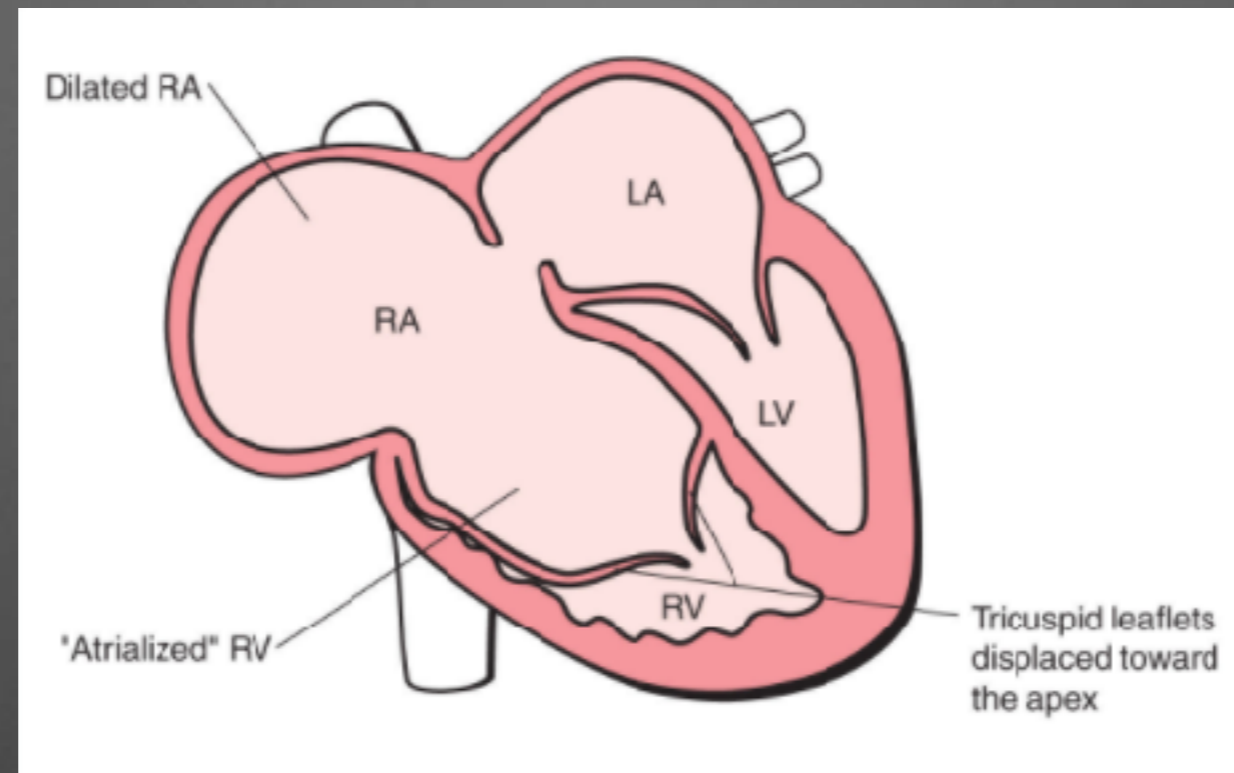


# Valvular Pathology: ASO

- neo-aortic regurgitation
  - 3% incidence 3+ AI
  - Incidence higher in TGA with VSD
- proximal neo-aorta dilation
- Toronto group 20 year follow up no reintervention



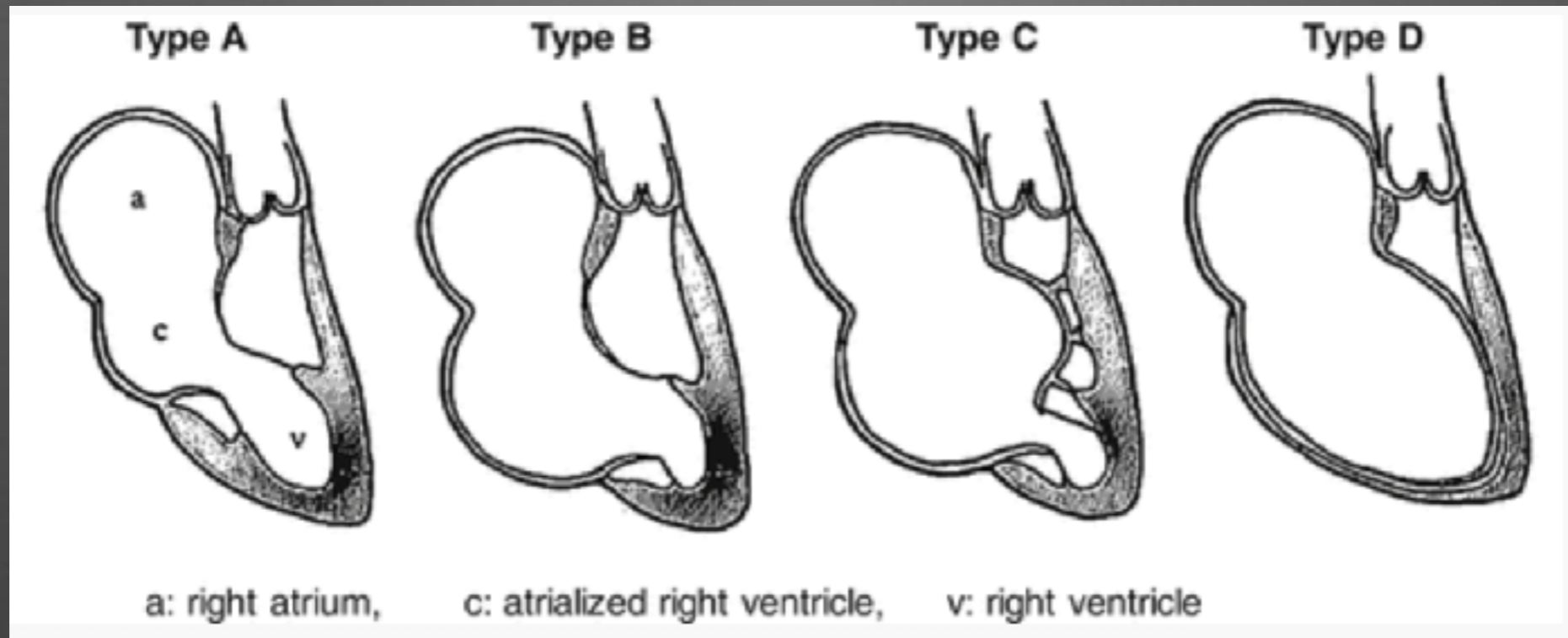
# Ebstein anomaly



- Displacement of septal leaflet of  $8\text{mm}/\text{m}^2$
- Average male BSA  $1.9 = 1.5\text{ cm}$
- Average female BSA  $1.6 = 1.3\text{ cm}$



# Carpentier Classification



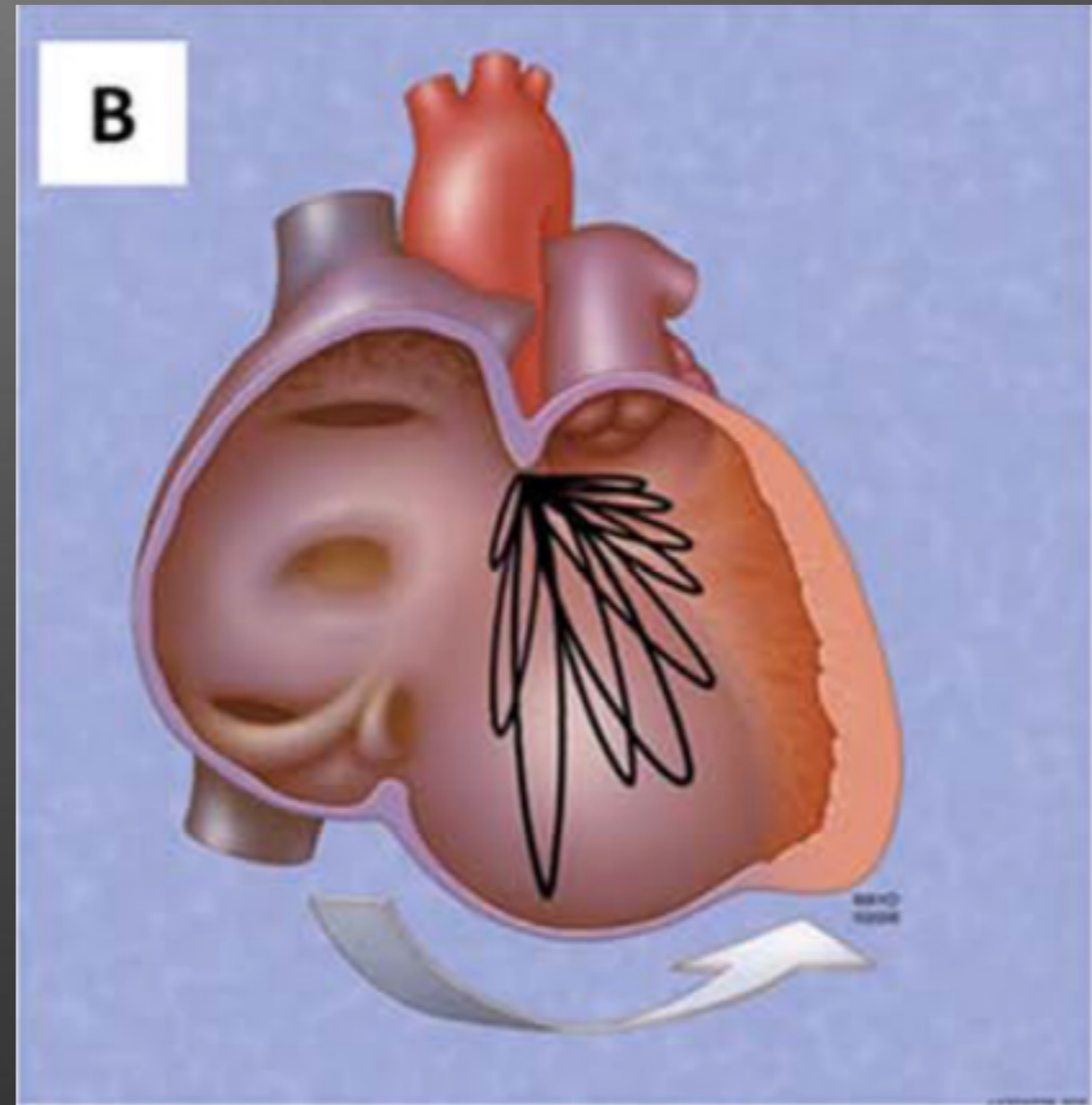
- Volume of RV
- Motion of the anterior leaflet
- Obstruction of the RVOT





# TV orifice

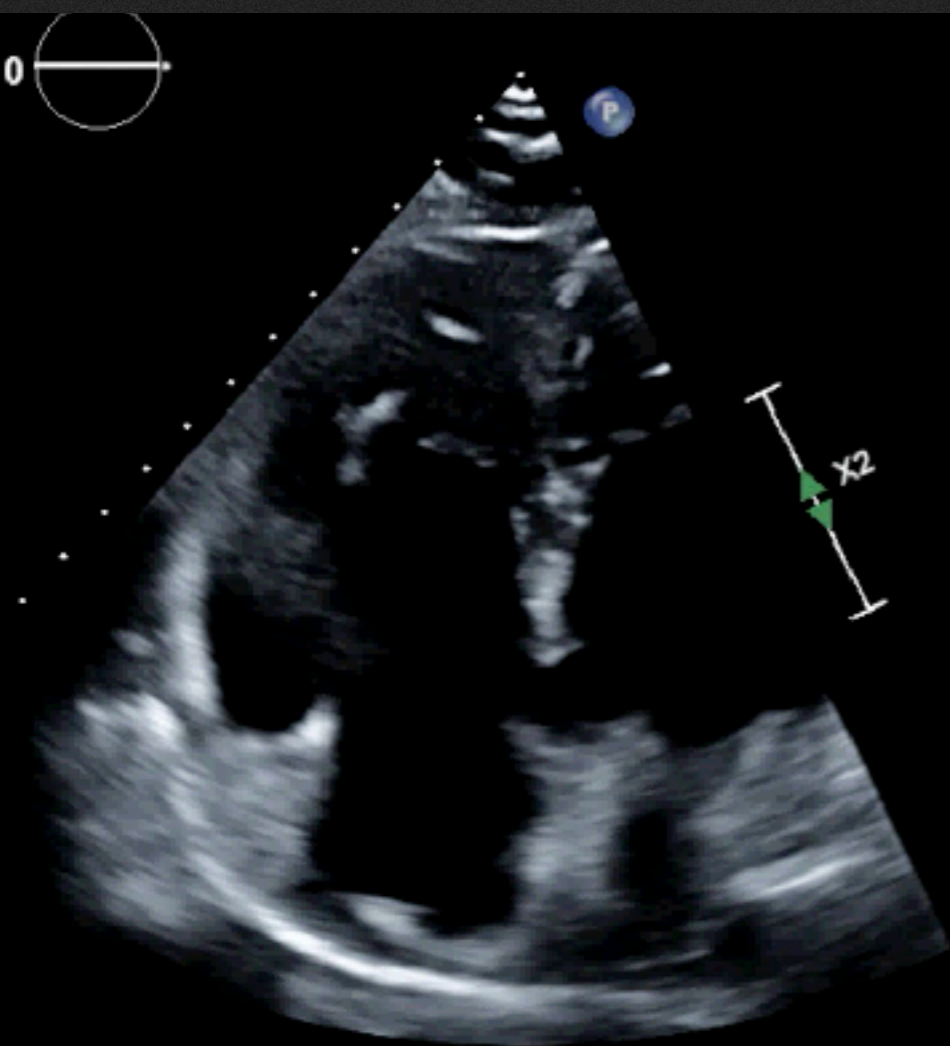
- Regurgitation
- Stenosis



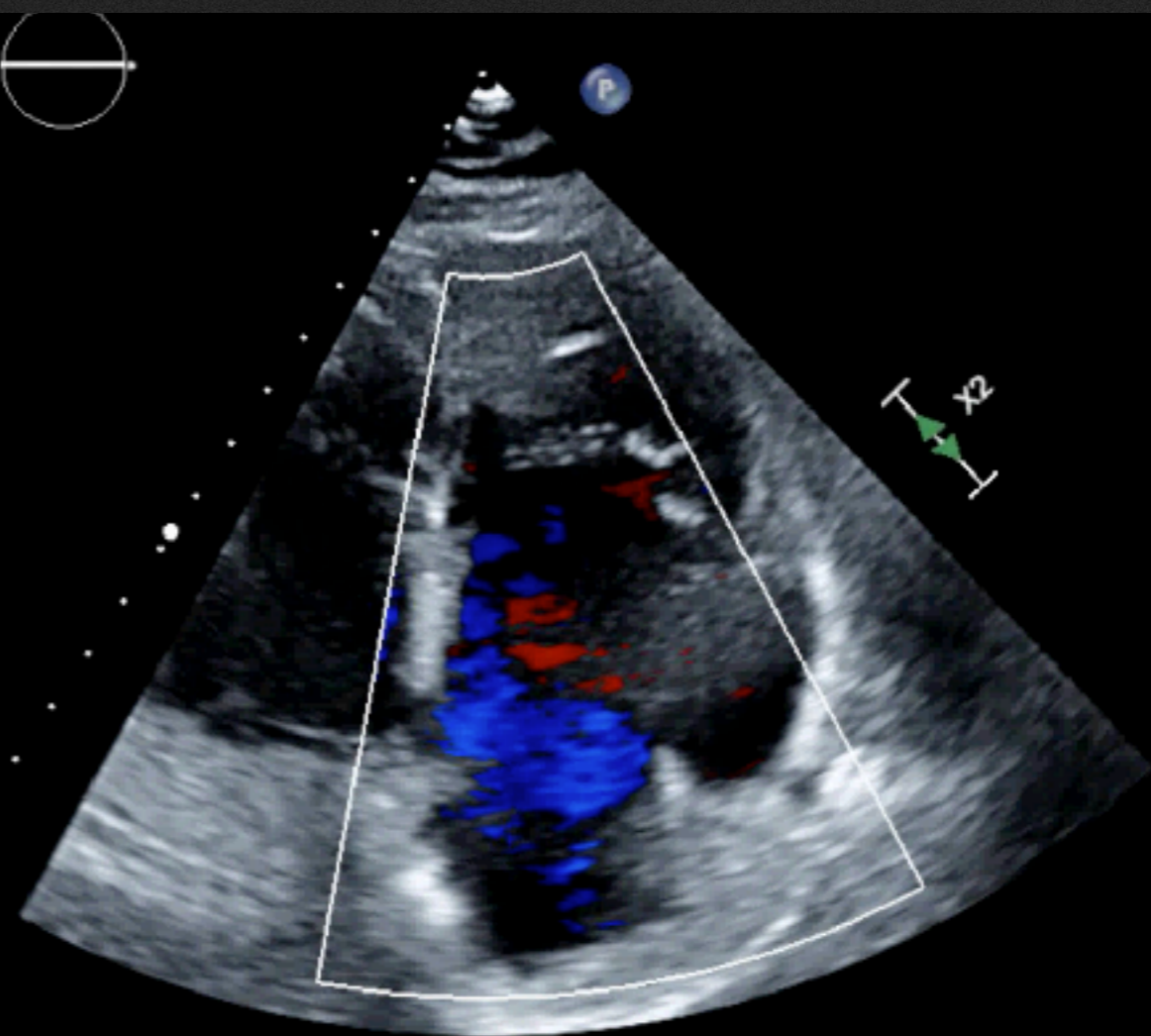
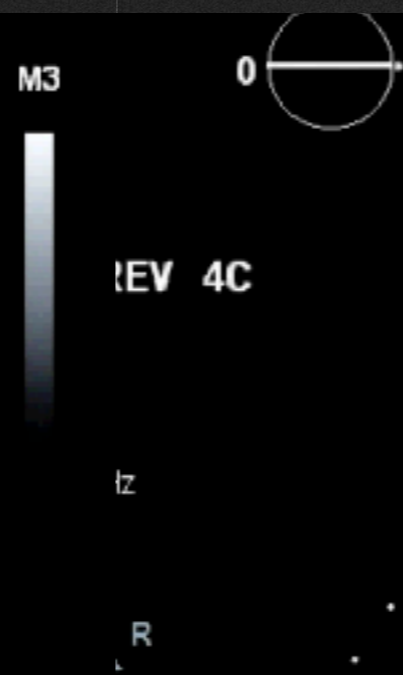
# Case 7

- 29 y.o. female who dx age 8 with Ebstein anomaly based on a murmur
- Presents for assessment of risk of pregnancy



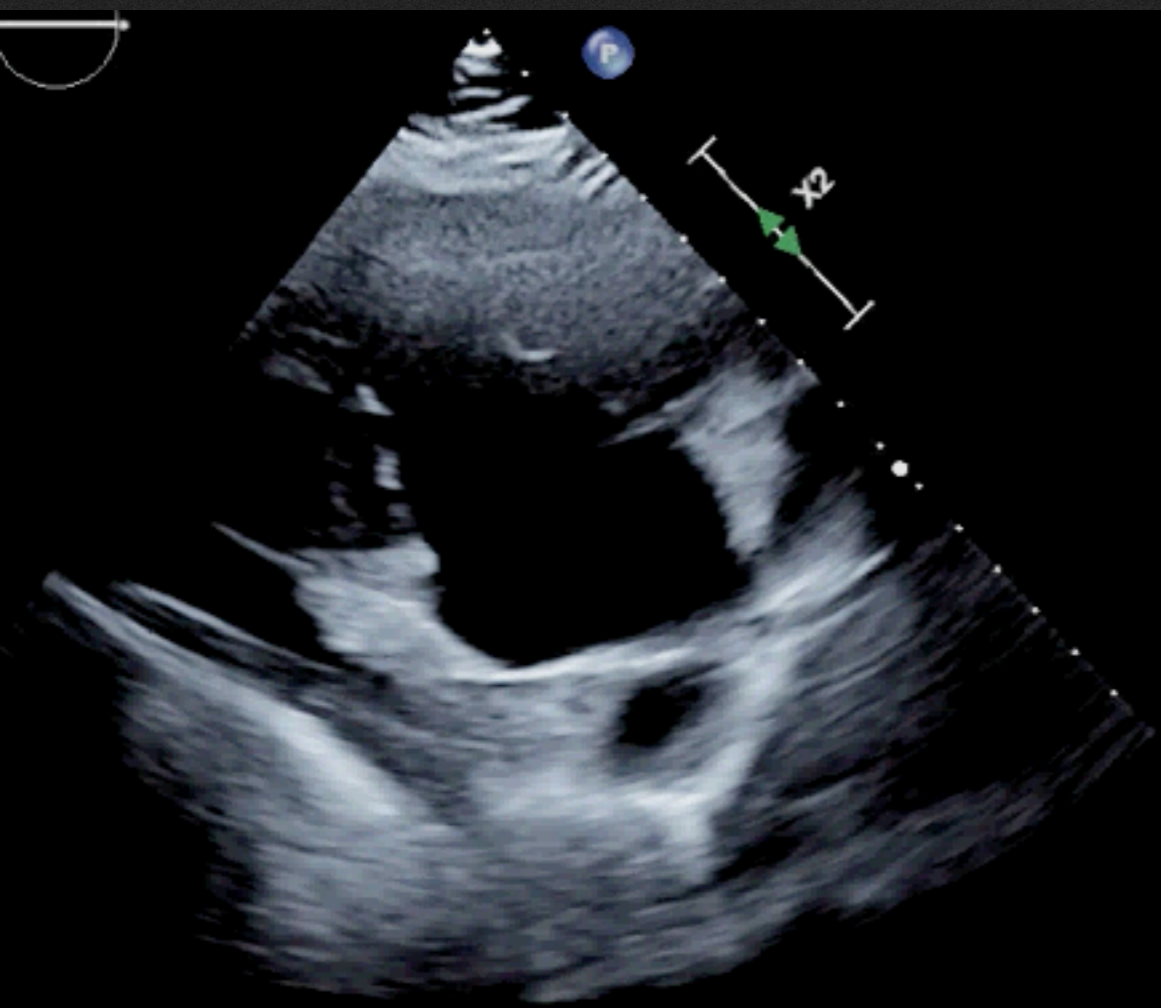


71 bpm



65 bpm

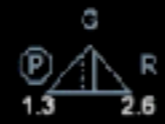




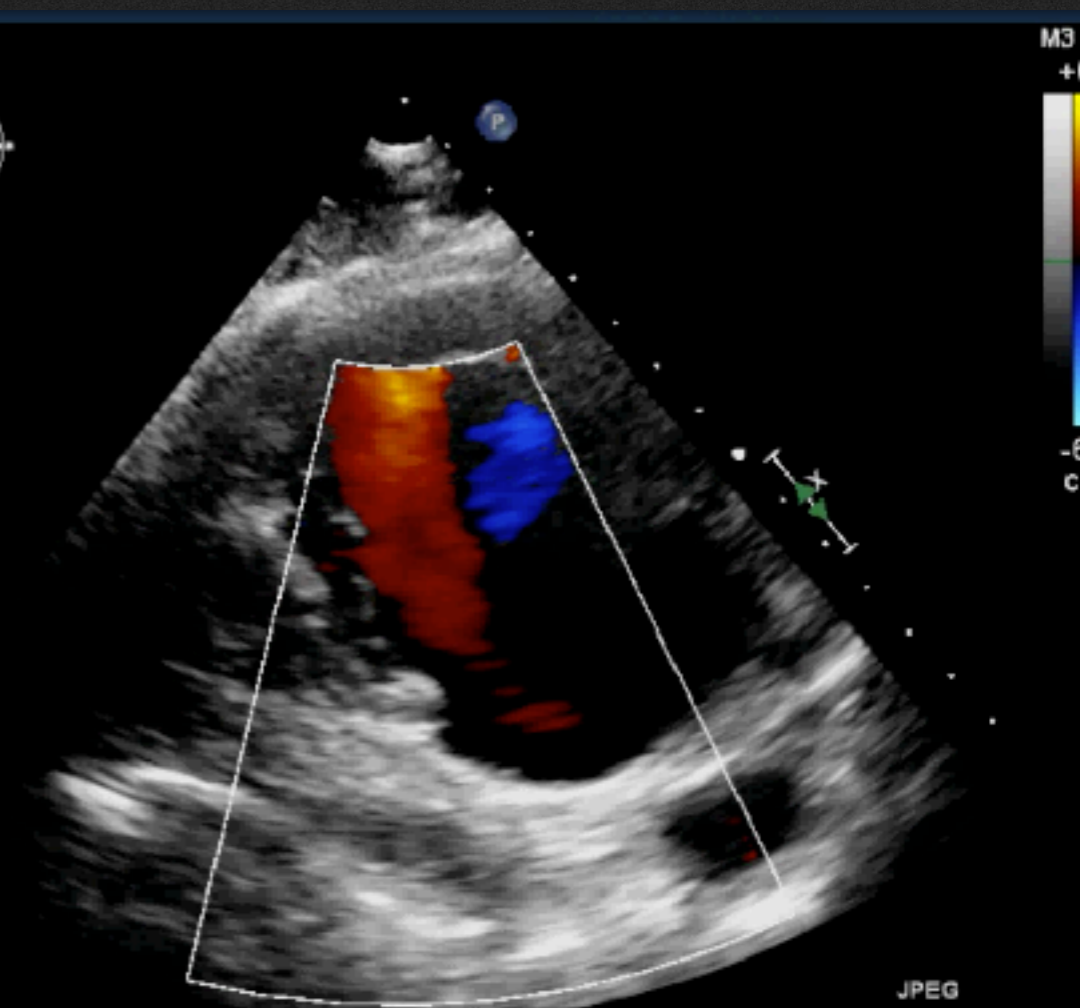
M3 FR 15Hz  
15cm

2D  
59%  
C 50  
P Low  
HPen

CF  
63%  
2.5MHz  
WF High  
Med



74 bl



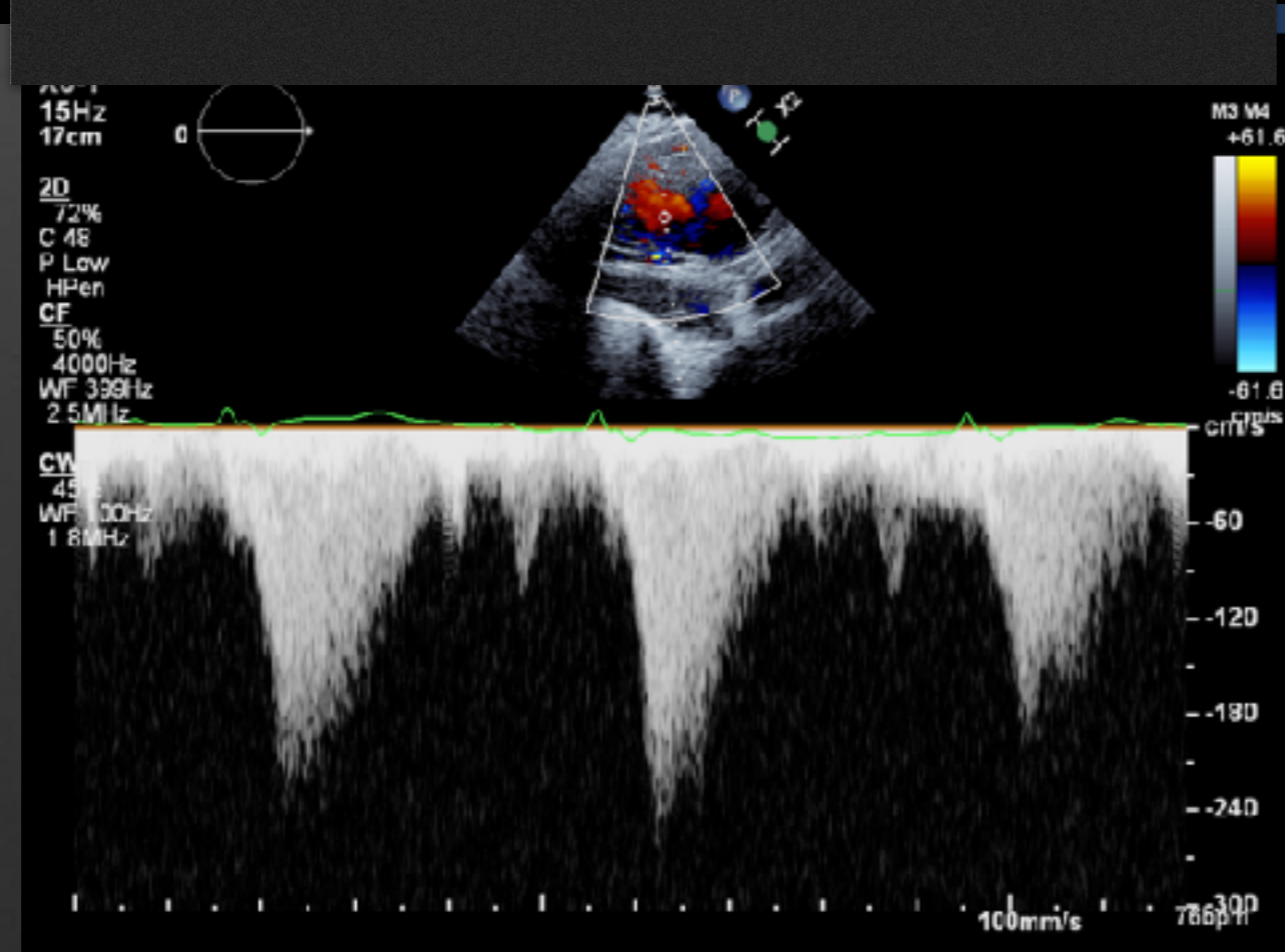
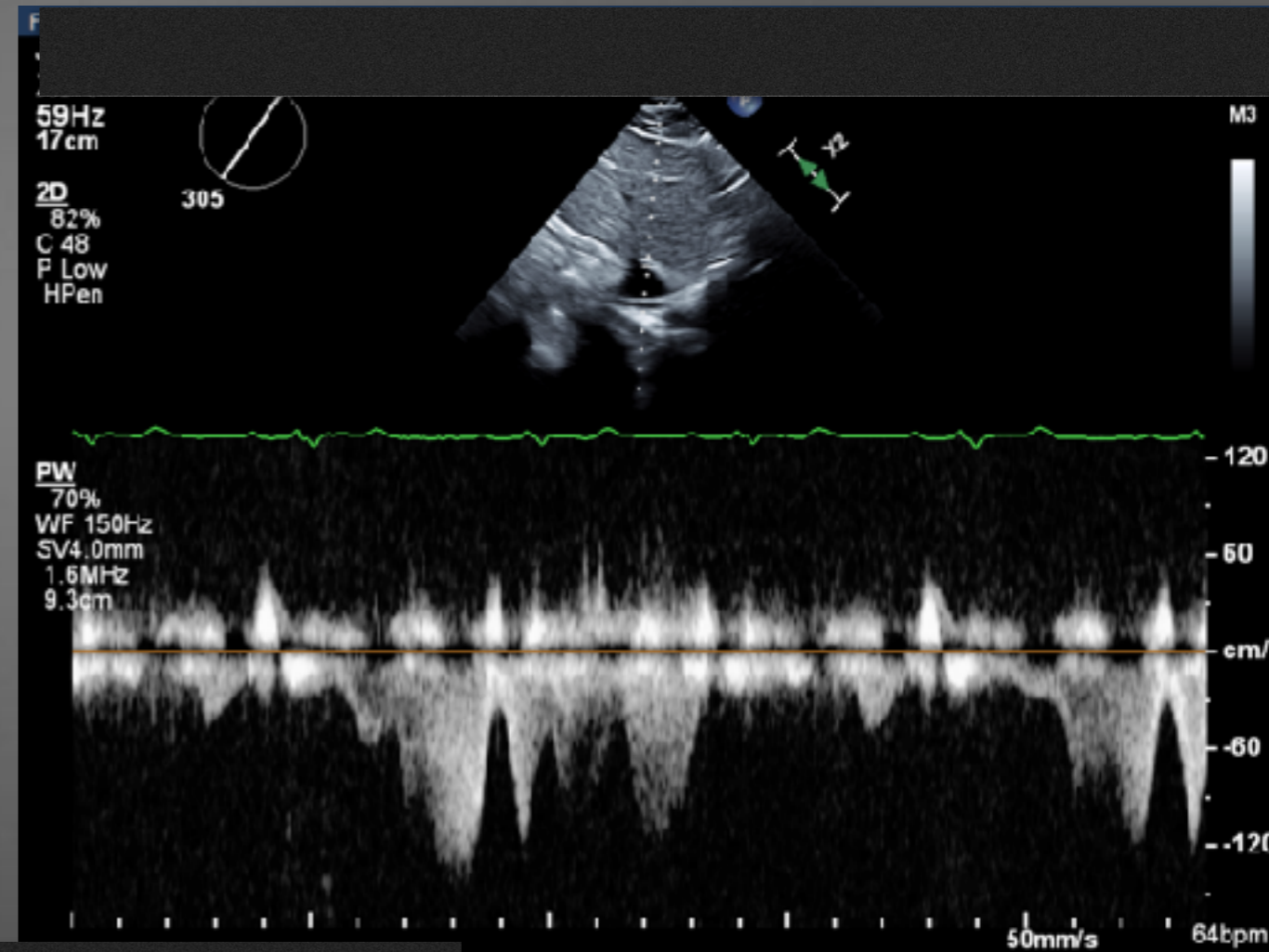
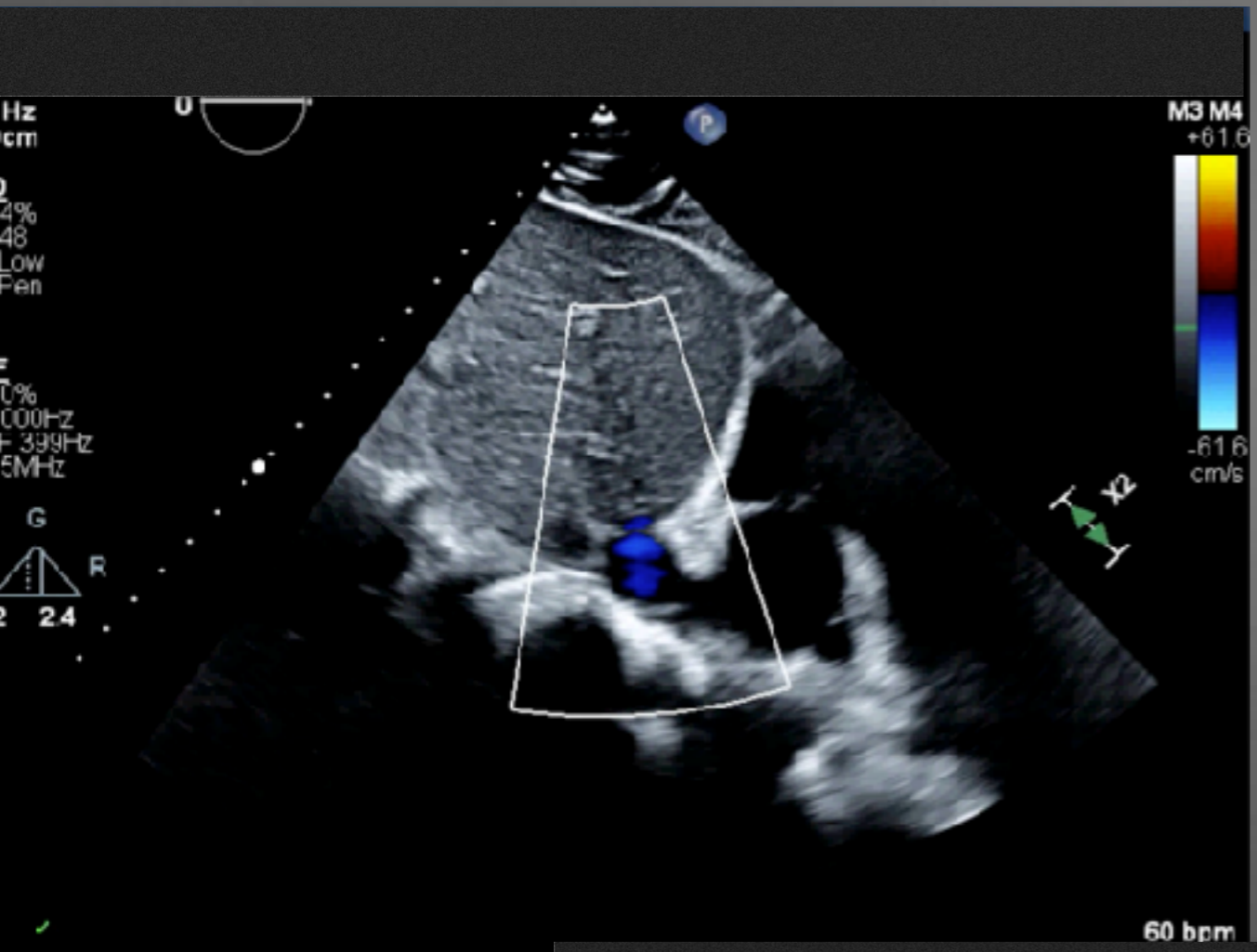
M3 +

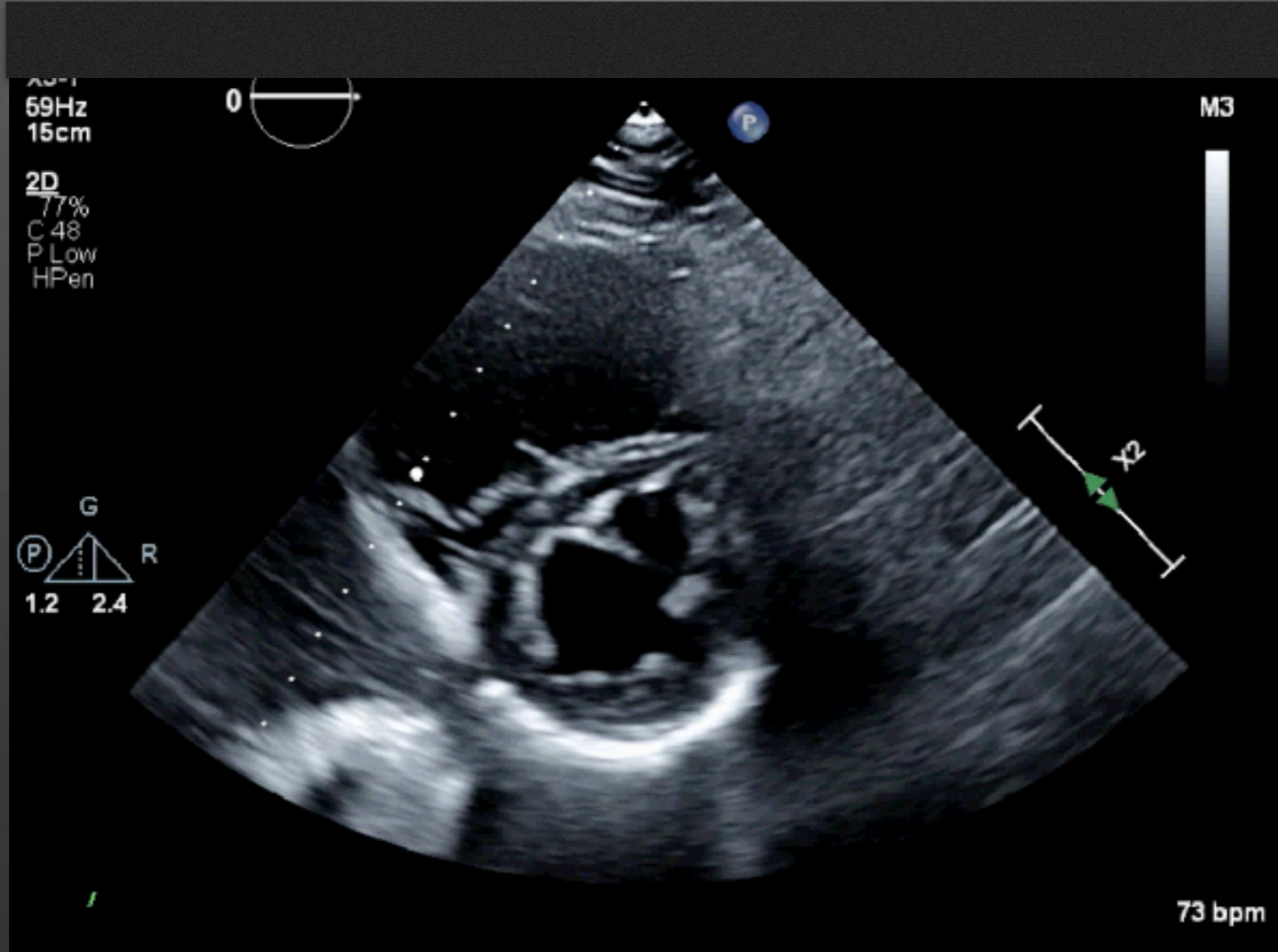
-6  
C

JPEG

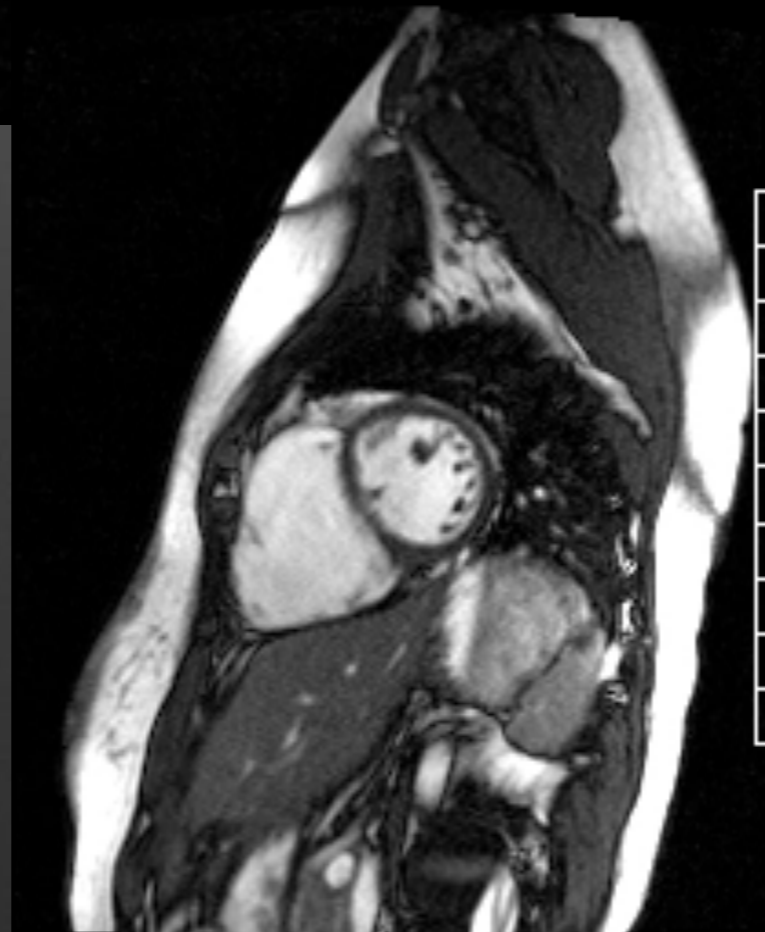
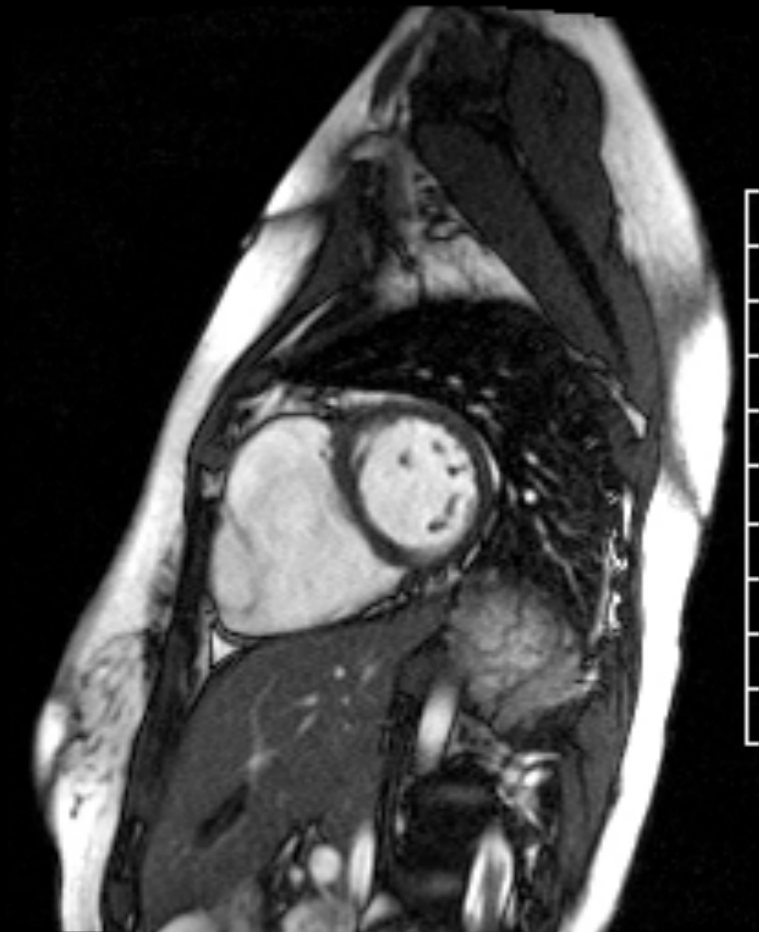
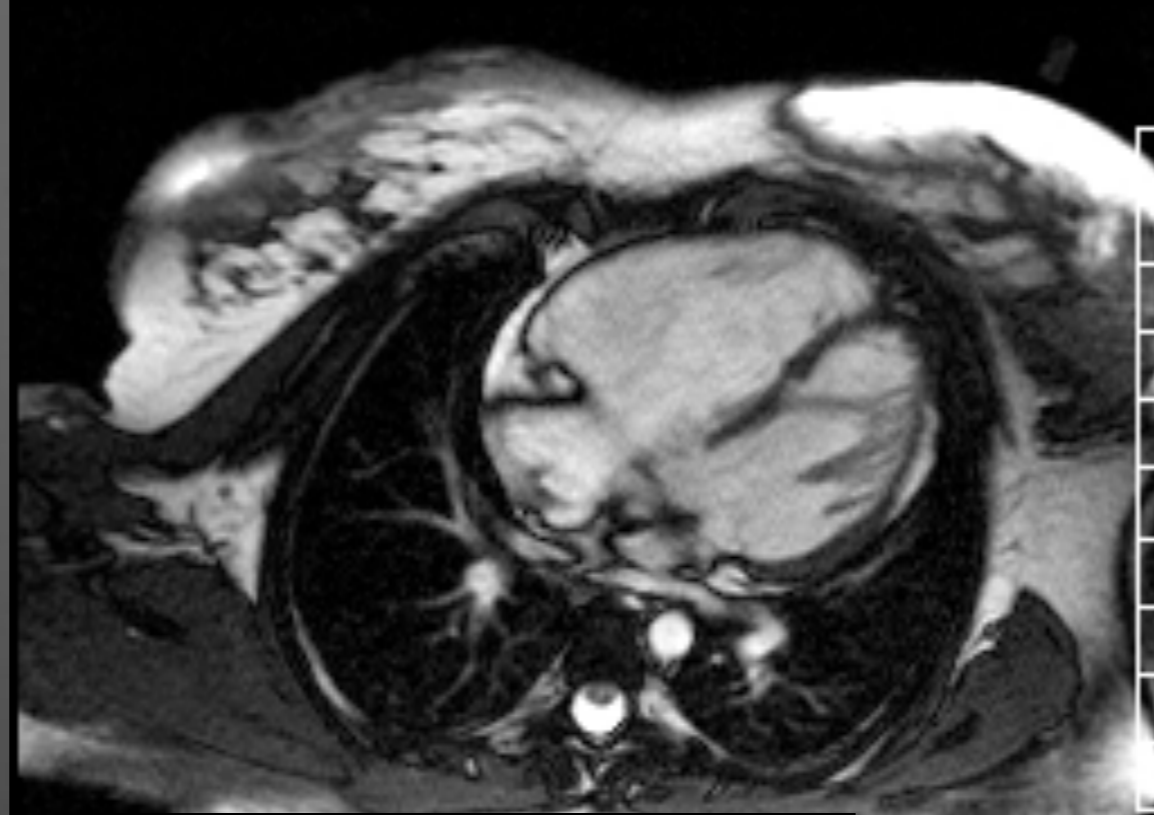
70 bl





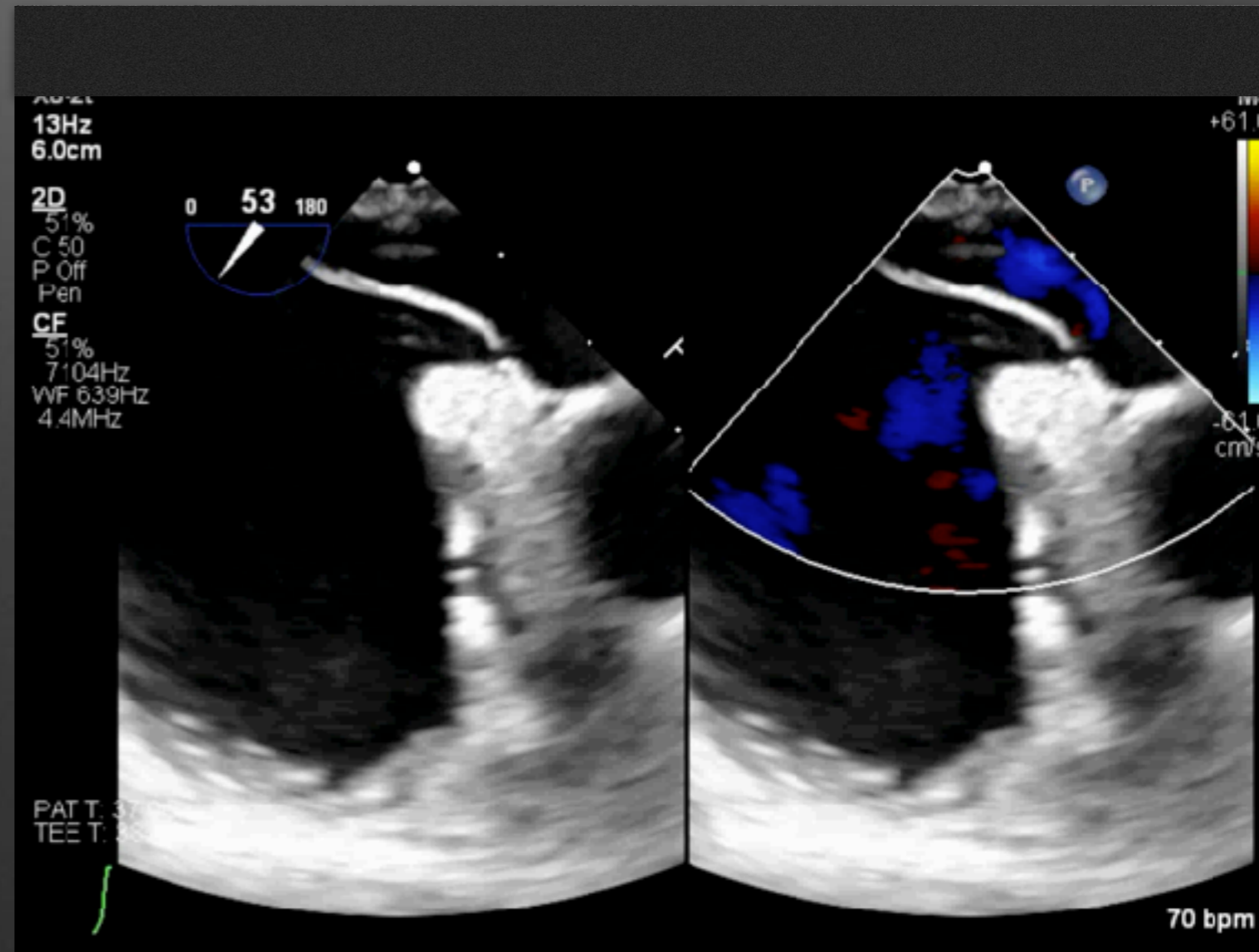
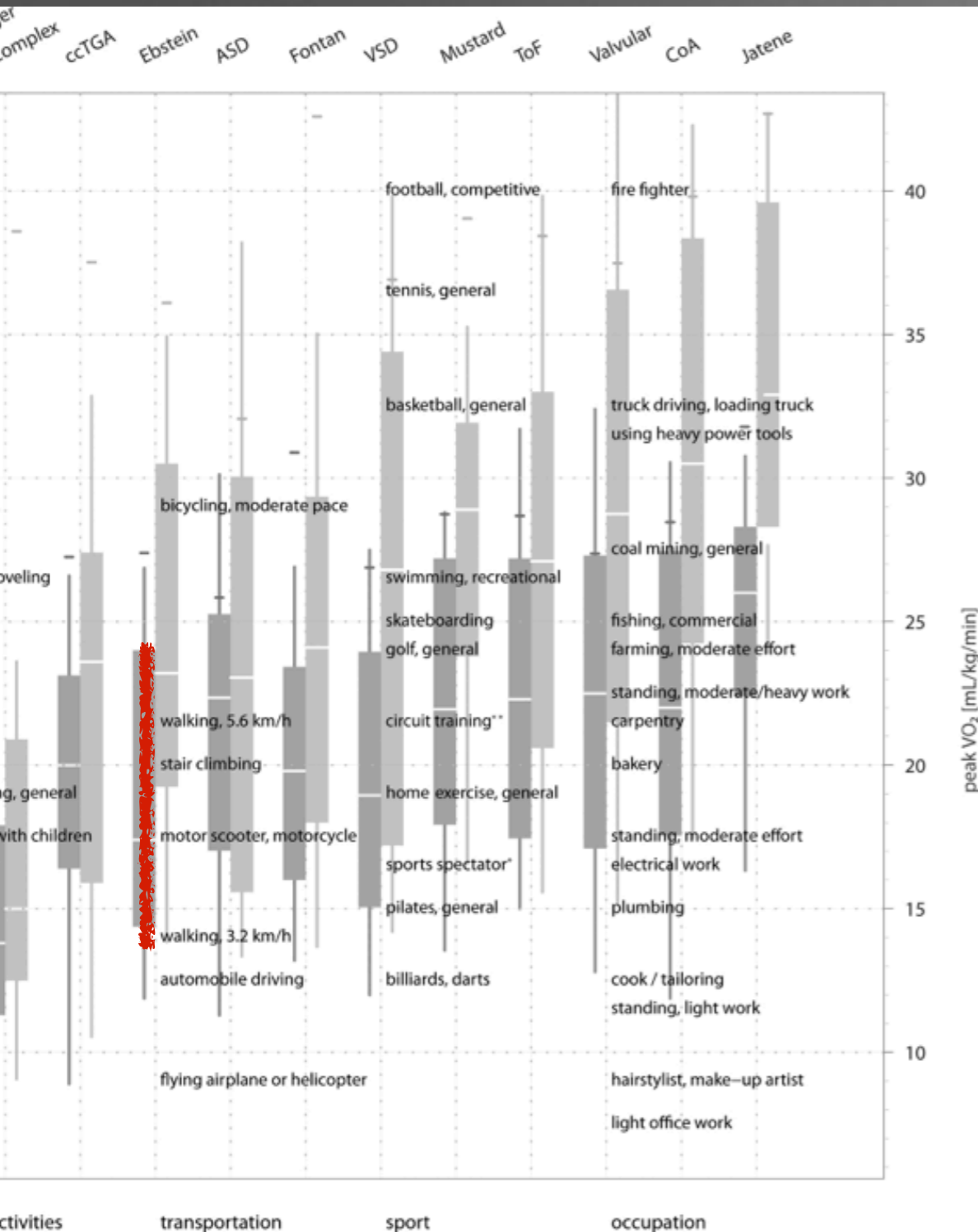


# CMR



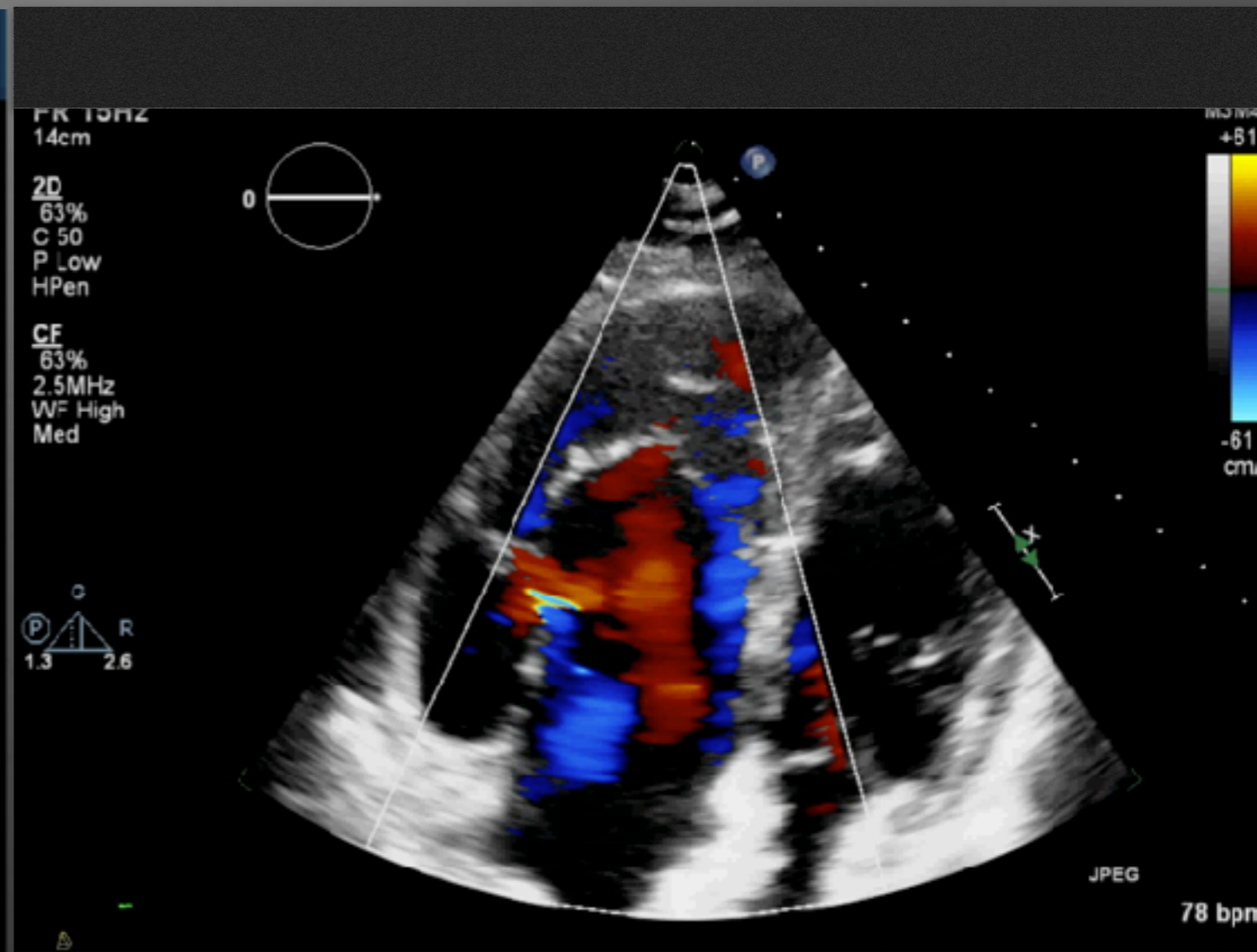
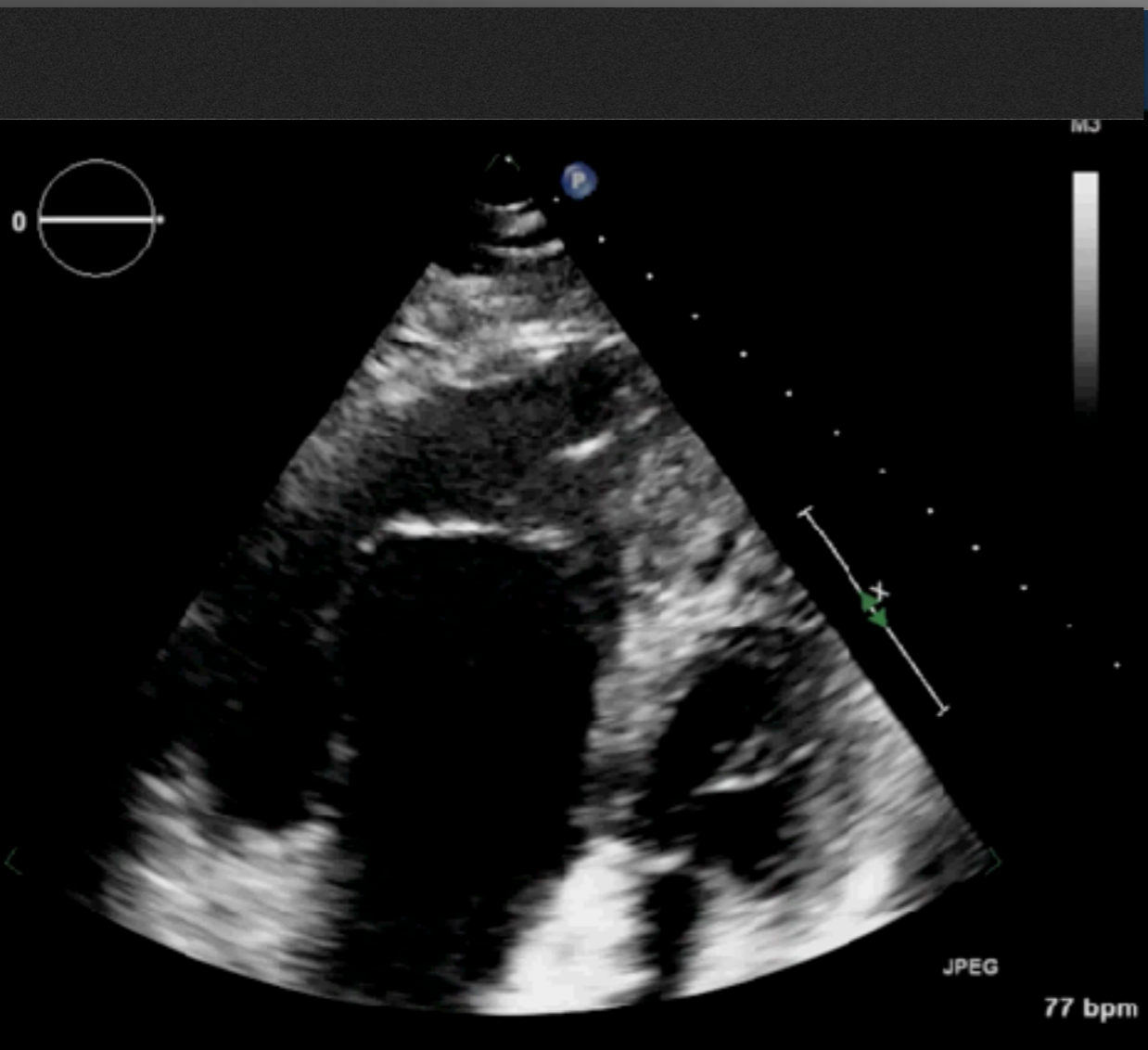
# CPET

- Peak Vo2 of 24
- Saturations decreased





# Pregnancy Echo



# Adult Congenital Heart Disease

Heterogeneity Anatomy and Complications

CV Testing

Percutaneous/Surgical