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**Children's Health**  
Medical University of South Carolina



# **Atrioventricular Septal Defects: Imaging Techniques after Surgery**

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**Associate Professor**

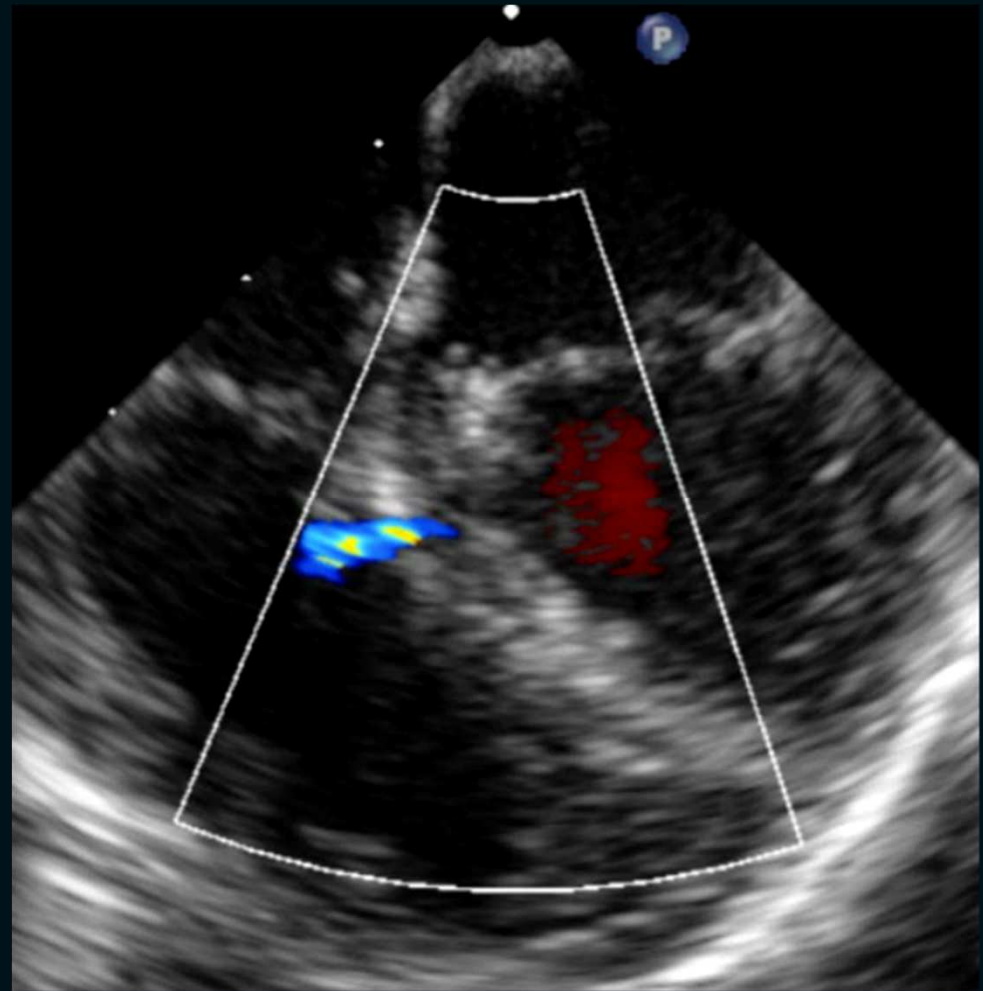
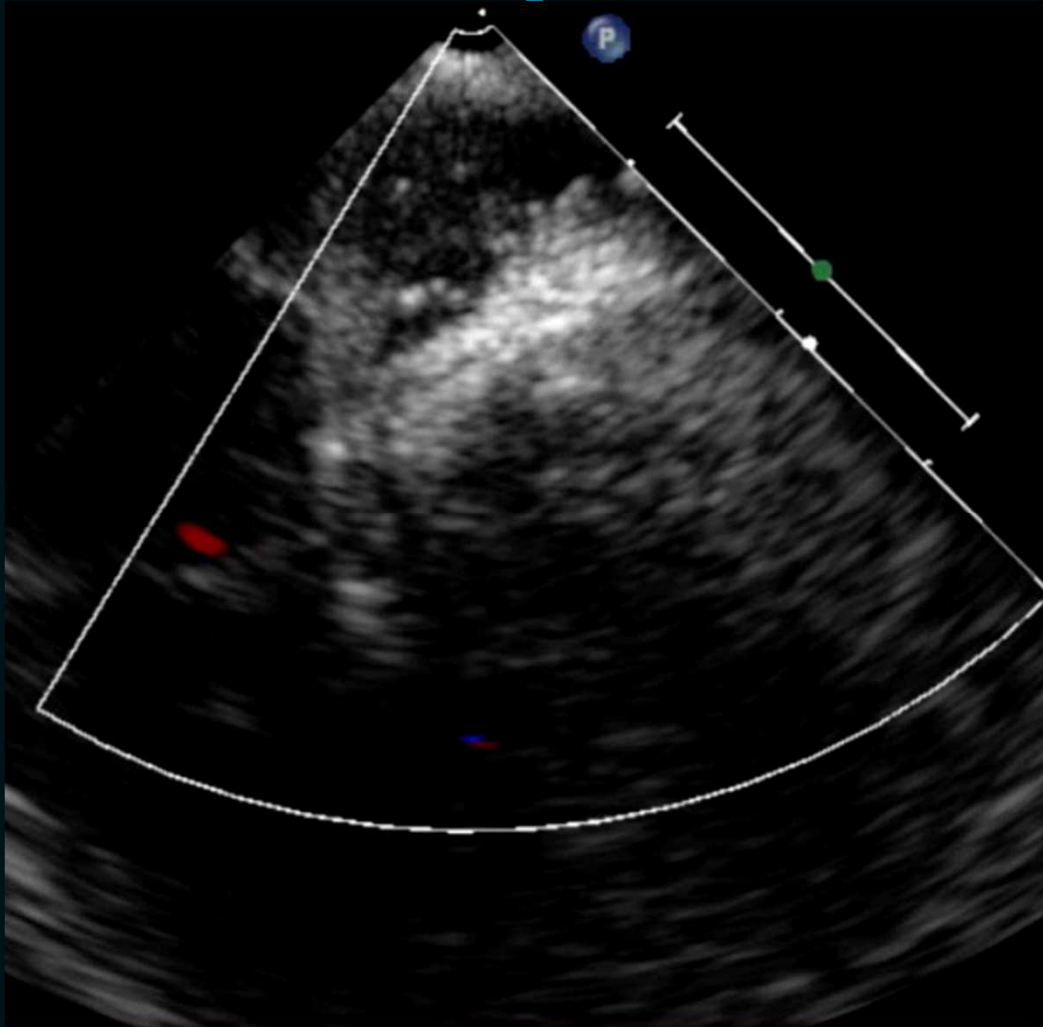
**Pediatrics and Radiology**

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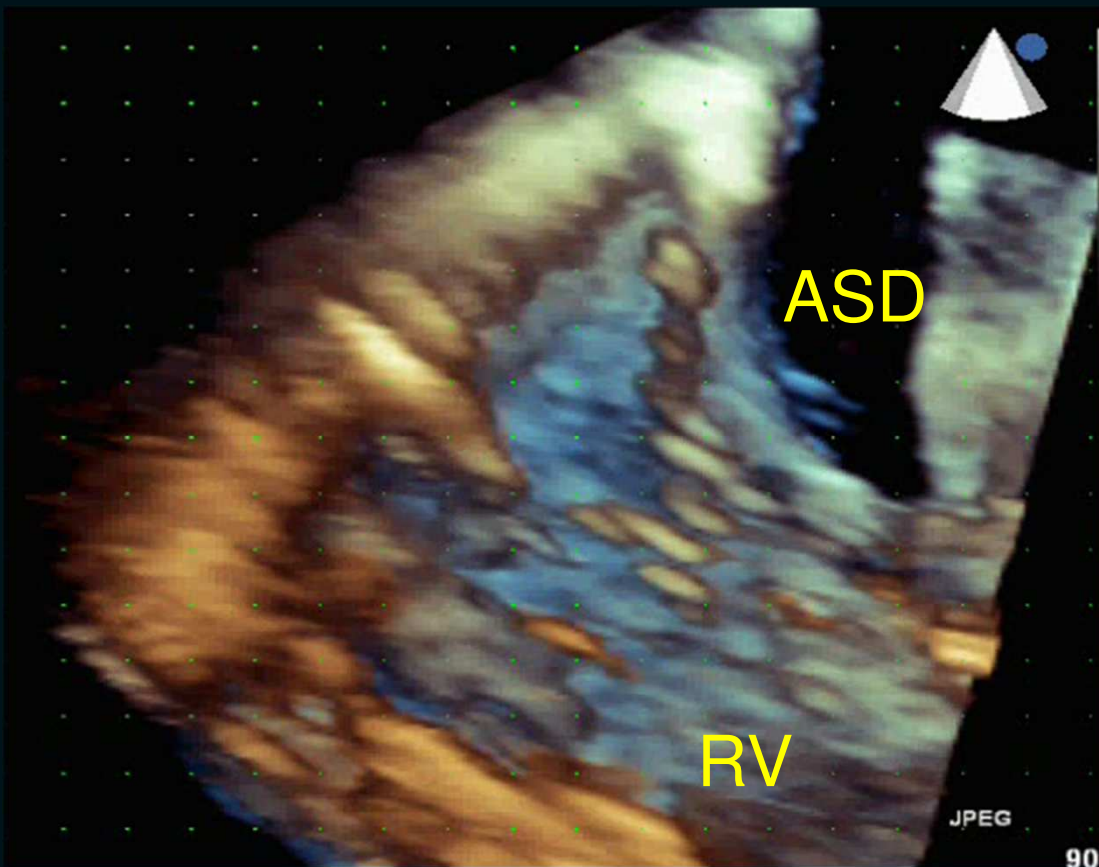
Changing What's Possible

[MUSCkids.org](http://MUSCkids.org)

# Intraop TEE-Residual Defects

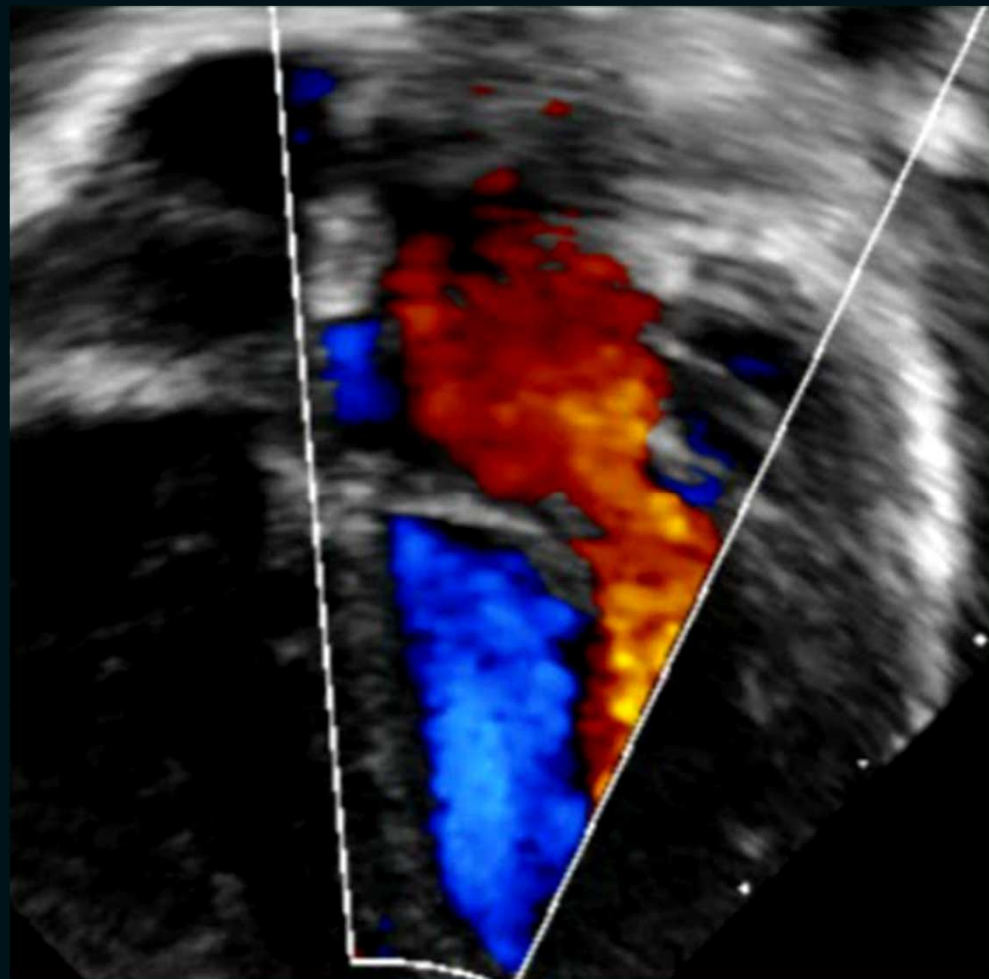
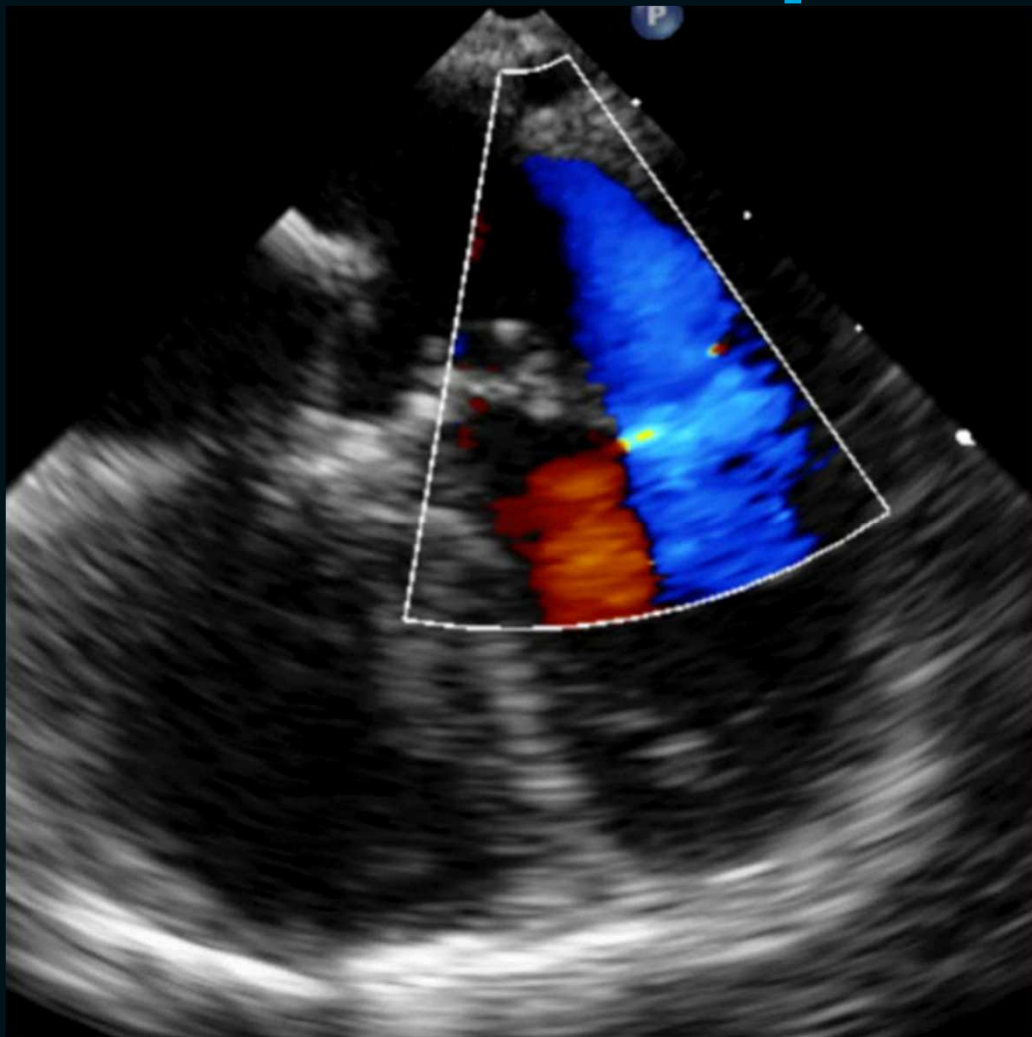


# TEE—larger residual ASD



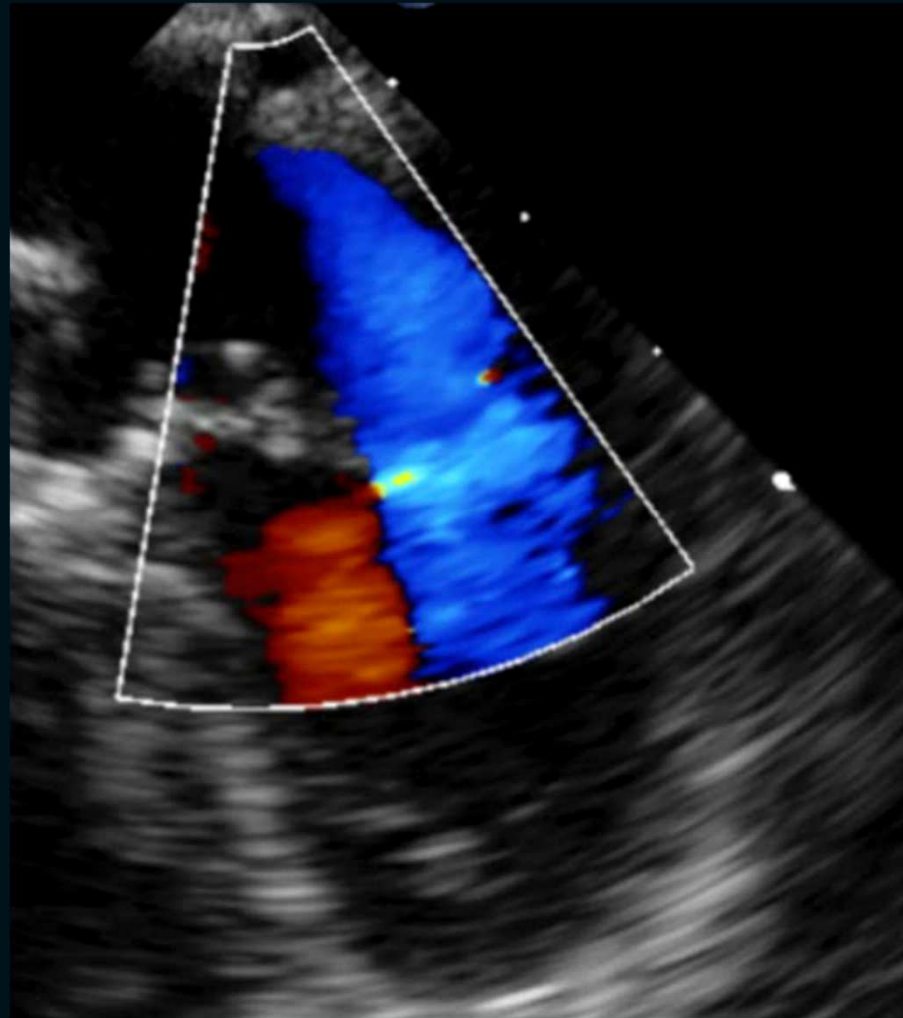
Courtesy of Girish Shirali

# Intraop TEE--LAVVR

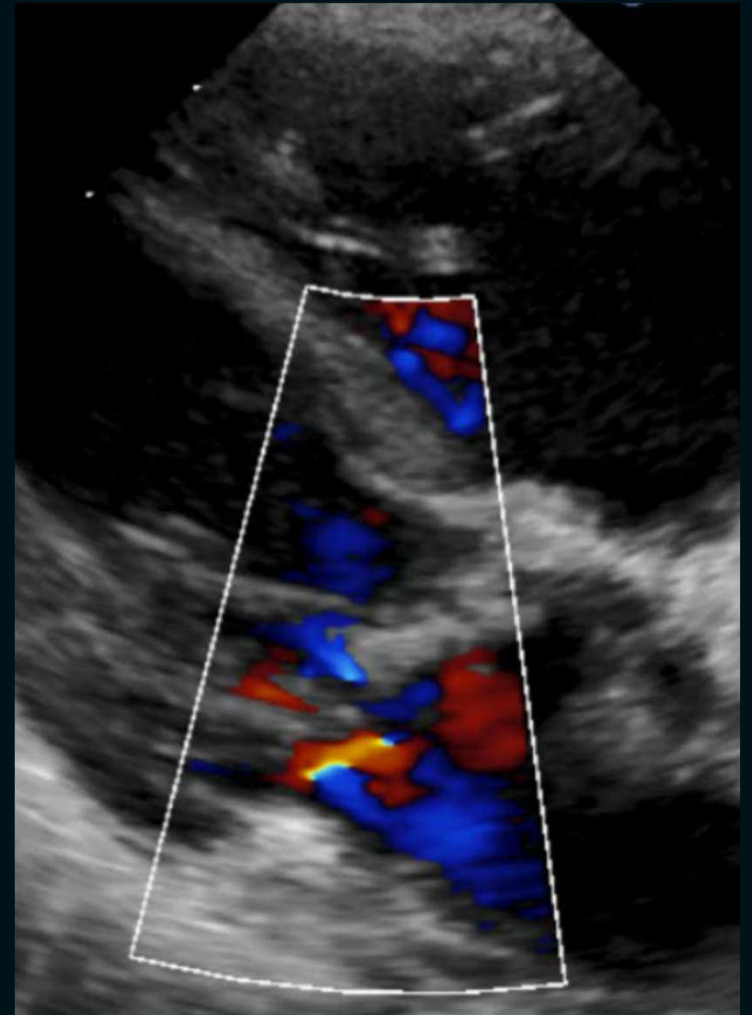
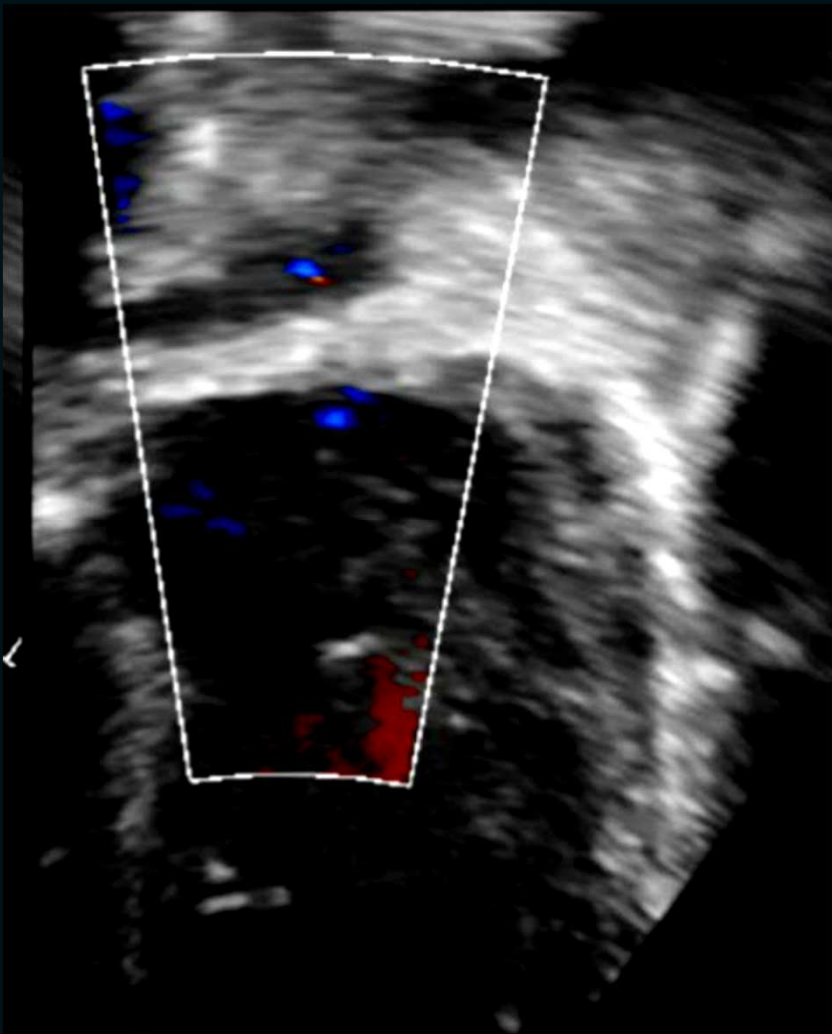




# Intraop TEE—Trivial LAVVR



# Discharge TTE—Same Patient



## Kim et al. Ann Thorac Surg, 2005. 80(1)



- ▶ *“Predictive value of intraoperative transesophageal echocardiography in complete atrioventricular septal defect”*
- ▶ Single center, 35 patients, 1997-2004
- ▶ Compared intraop TEE to subsequent TTE
- ▶ 29% (10/35) LAVVR progressed from grade I to II



# Do Residual Lesions Persist?



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# Atz et al. JTCVS, 2011. 141(6)

- ▶ PHN ACE MR study (Complete AVSD)
- ▶ 1 Month Echo
  - ▶ 16% had a residual ASD (1% >3mm)
  - ▶ 48% with a residual ventricular shunt (8% >3mm)
- ▶ 6 Month Echo
  - ▶ No patients with ASD >3mm
  - ▶ 1 patient (out of 117) had VSD >3mm

# Atz et al. JTCVS, 2011. 141(6)



- ▶ LV dysfunction
  - ▶ 35% at 1 month
  - ▶ 10% at 6 months
- ▶ No change in #  $\geq$  moderate LAVVR (24% vs 22%)
- ▶ No patients had significant LAVV stenosis or LVOTO at either time point



# Left AV Valve Regurgitation

- ▶ 15-40% have moderate-severe LAVVR
- ▶ Generally stable after the 1<sup>st</sup> month
- ▶ 5-12% reoperation for LAVVR (contemporary series)
- ▶ Reoperation generally required within a few years
  - ▶ 3.5mo-2yr after 1<sup>st</sup> surgery

# How do we quantify Left AV Valve Regurgitation?



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## Prakash et al. *Pediatr Cardiol*, 2012. 33(2)



- ▶ *“Challenges in echocardiographic assessment of mitral regurgitation in children after repair of atrioventricular septal defect”*
- ▶ PHN ACE MR study
- ▶ 149 patients after repair of AVSD (61% complete, 39% partial/transitional)
- ▶ 6 month post-op echo
- ▶ Compared various methods to quantify LAVVR with LV volume z-score





# Comparison of Methods for LAVVR



- ▶ Subjective grade of LAVVR vs.
- ▶ Quantitative assessment
  - ▶ Vena contracta width
  - ▶ Vena contracta cross-sectional area
  - ▶ Regurgitant volume/fraction

Prakash et al. Pediatr Cardiol, 2012. 33(2)

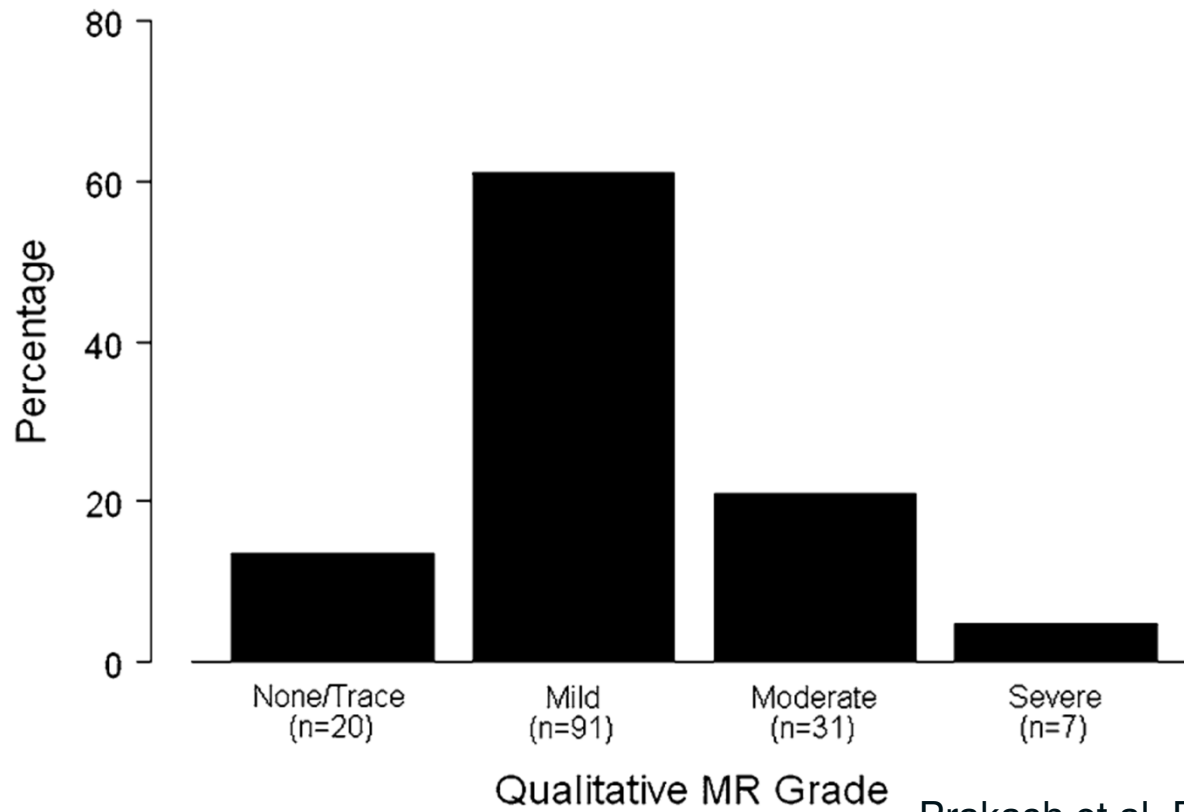


# Regurgitant Volume/Fraction

- ▶ LV stroke volume from 2D tracing - Aortic stroke volume by Doppler
- ▶ Correlated with LVEDVz (both  $R^2$  0.45), but...
- ▶ Negative value in 75% of subjects
- ▶ Poor intra-observer agreement (ICC 0.281, 0.167)

Prakash et al. Pediatr Cardiol, 2012. 33(2)

# Subjective grade of LAVVR



Prakash et al. Pediatr Cardiol, 2012. 33(2)

# Subjective grade of LAVVR



- ▶ Correlated with quantitative measurements of regurgitation
- ▶ Significantly associated with LVEDVz ( $R^2=0.53$ )
- ▶ Modest intraobserver agreement (kappa 0.56)

Prakash et al. Pediatr Cardiol, 2012. 33(2)



# Vena Contracta Width/Area

- ▶ Lateral and anterior-posterior
- ▶ Subjects with  $>1$  jet were excluded
- ▶ Significantly associated with LVEDVz ( $R^2$  0.24-0.54)
- ▶ Best interobserver agreement (ICC 0.62-0.73)
- ▶ Not significantly better than subjective grading

Prakash et al. Pediatr Cardiol, 2012. 33(2)



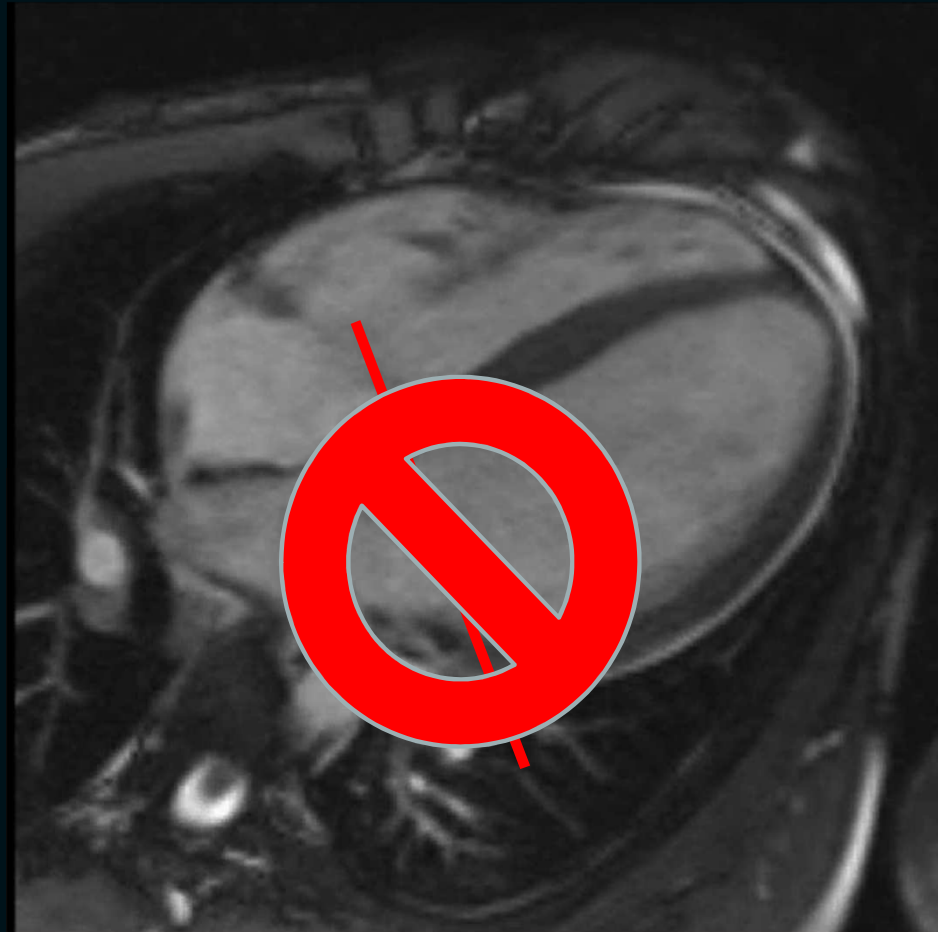
# Cardiac MRI for LAVVR

- ▶ Very limited data in literature for CHD
- ▶ Valve anatomy generally seen better by echo (TTE  $\pm$  TEE)
- ▶ Accurate measurement of LV volume and function
- ▶ Regurgitant fraction/volume
- ▶ Adult data (non CHD) suggests poorer outcomes with RF >40% or RVol >29ml/m<sup>2</sup>
  - ▶ Better predictive value than LV volume

Myerson SG, et al. Circulation. 2016;133:2287-2296



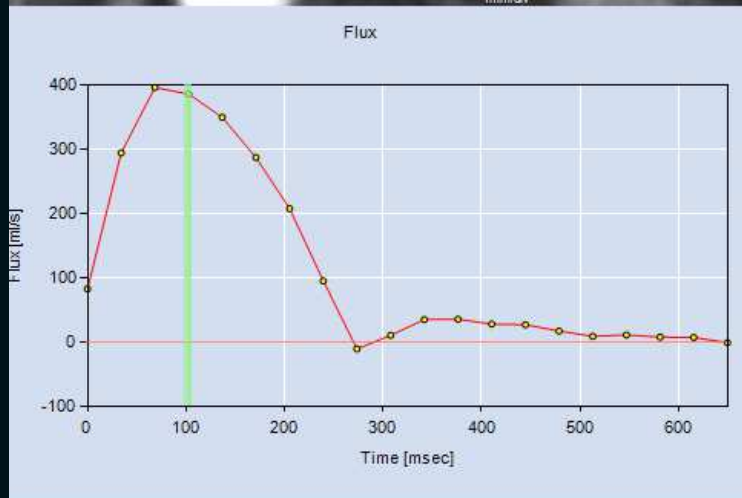
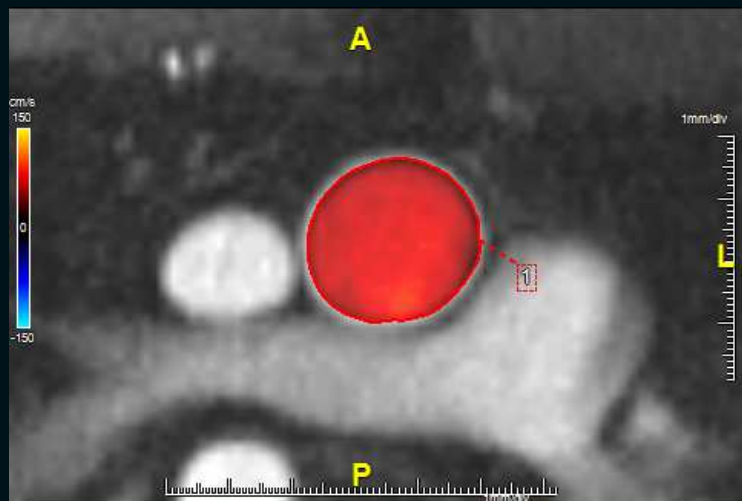
# CMR Quantification of LAVVR



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# CMR Quantification of LAVVR

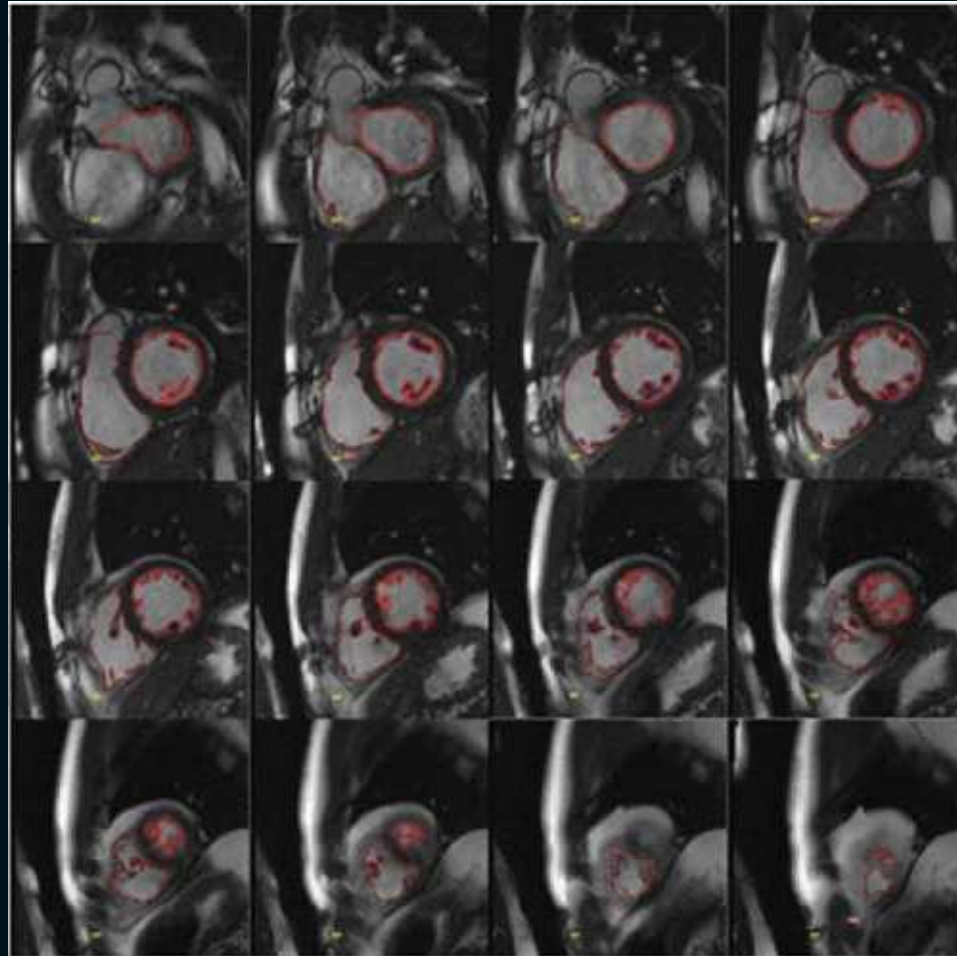


ROI No: 1  
 Stroke Volume: 76.79 ml  
 Forward Flow Volume: 76.89 ml  
 Backward Flow volume: 0.1 ml  
 Regurgitant Fraction: 0.13 %  
 Absolute Stroke Volume: 76.99 ml  
 Mean Flux: 114.34 ml/s  
 Mean velocity: 20.61 cm/s  
 Stroke Distance(time velocity integral): 13.8 cm



No	Time [msec]	Flux [ml/s]	Area [cm <sup>2</sup> ]	Velocity [cm/s]	Max Vel [cm/s]	M
01	0.00	83.25	4.79	17.37	52.00	
02	34.20	294.85	5.30	55.59	82.00	
03	68.40	396.48	5.66	70.07	90.00	
04	102.60	386.46	5.62	68.74	94.00	
05	136.80	350.15	5.89	59.42	89.00	
06	171.00	287.70	5.79	49.73	77.00	
07	205.20	208.00	5.60	37.12	62.00	

# CMR Quantification of LAVVR



## LV Volumes

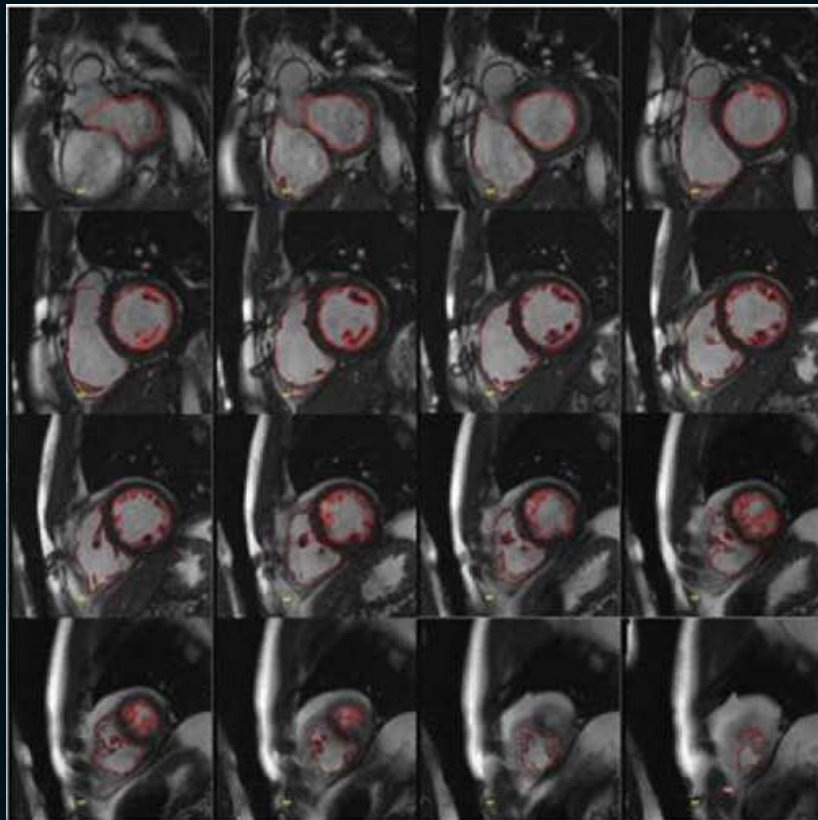
- Diastolic
- Systolic

Fratz S, et al. Am J Cardiol 2009;103:1764 –1769



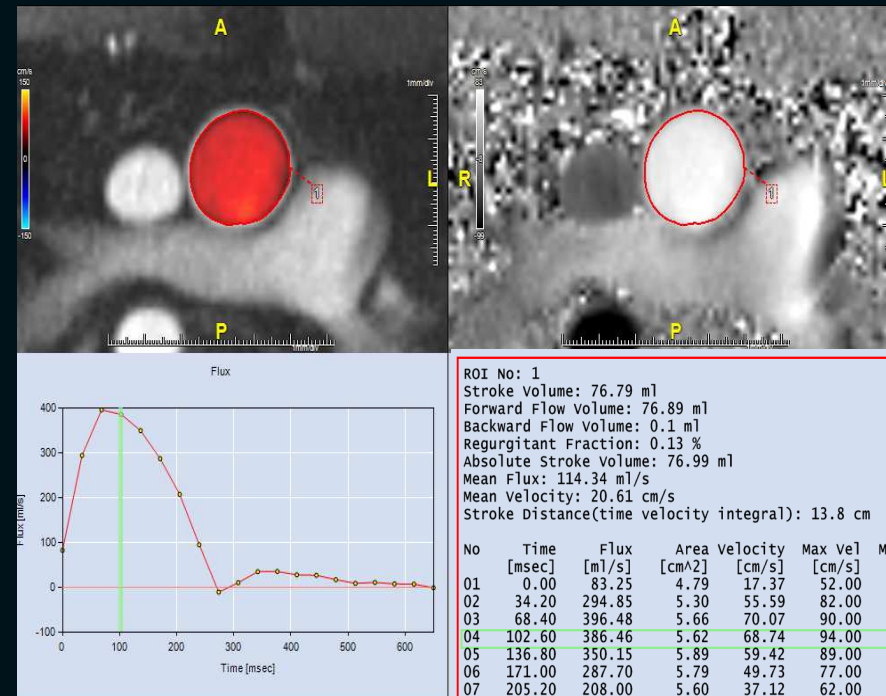
# CMR Quantification of LAVVR

## LV Stroke Volume



Fratz S, et al. Am J Cardiol 2009;103:1764 –1769

## - Ao Stroke Volume



= Mitral Regurgitation\*

\* Assuming no VSD

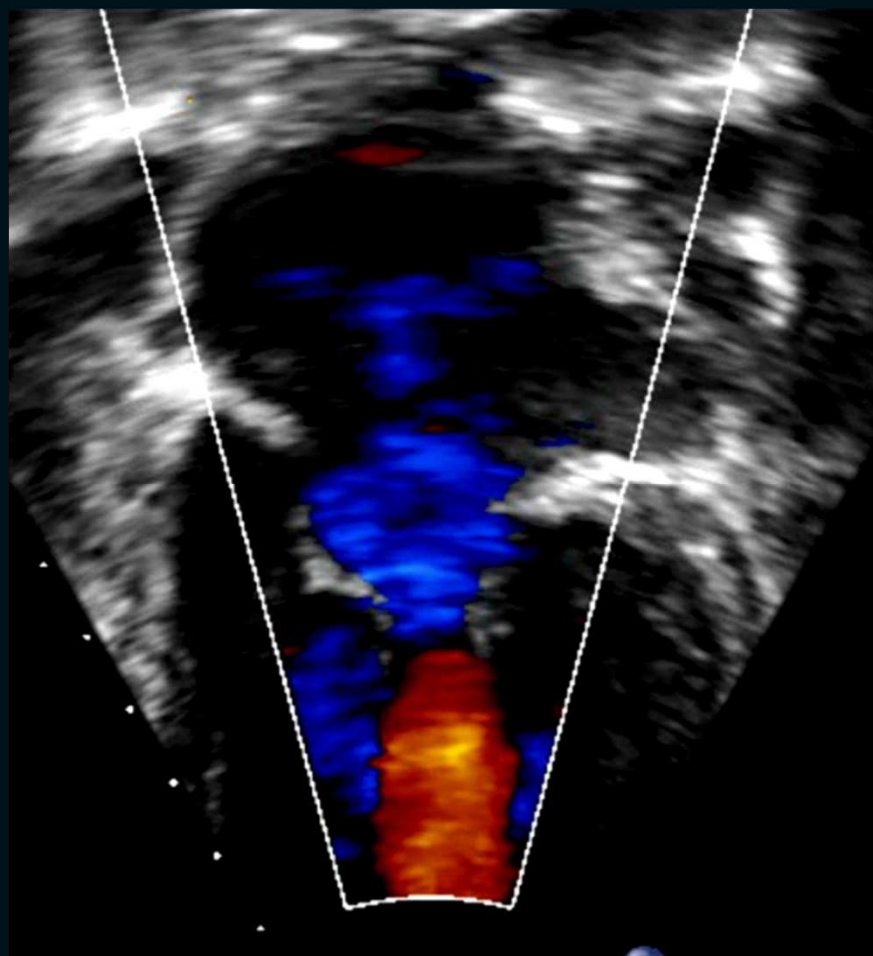
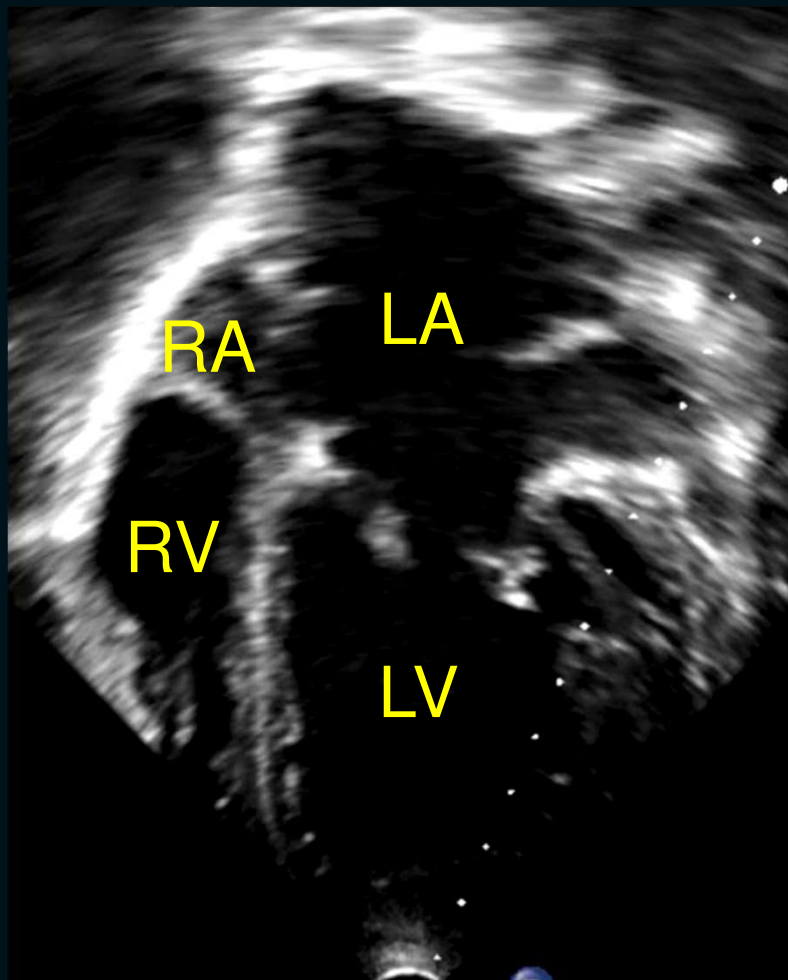
# Mechanisms of Left AV Valve Regurgitation



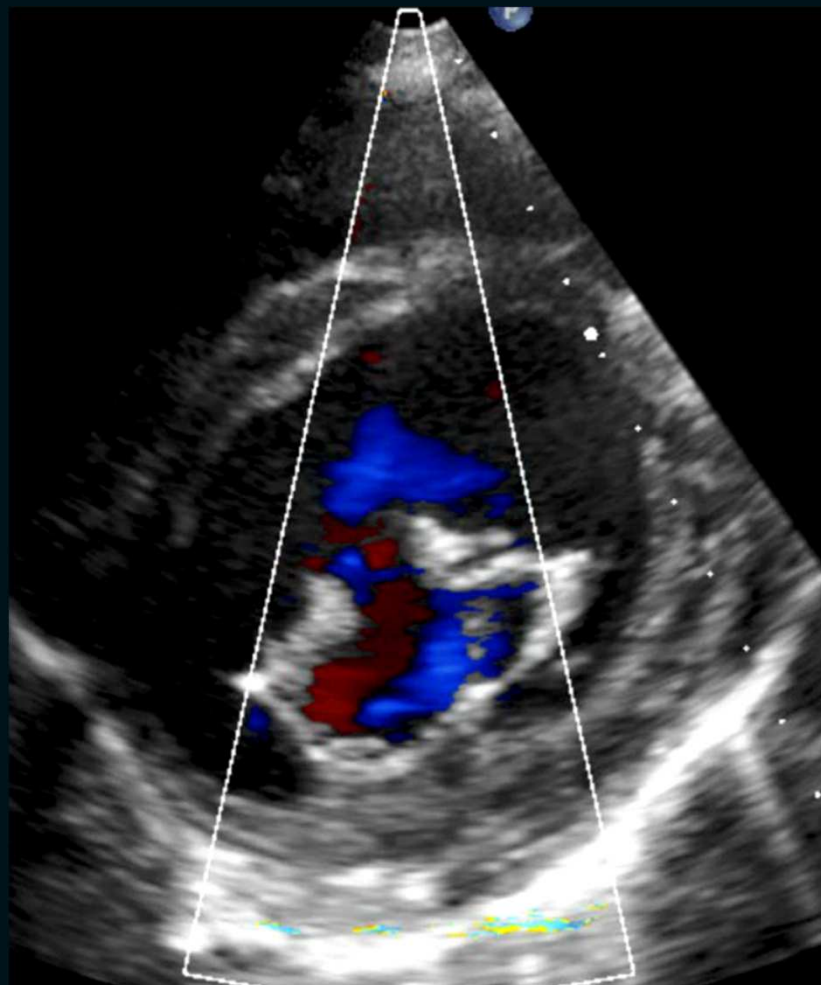
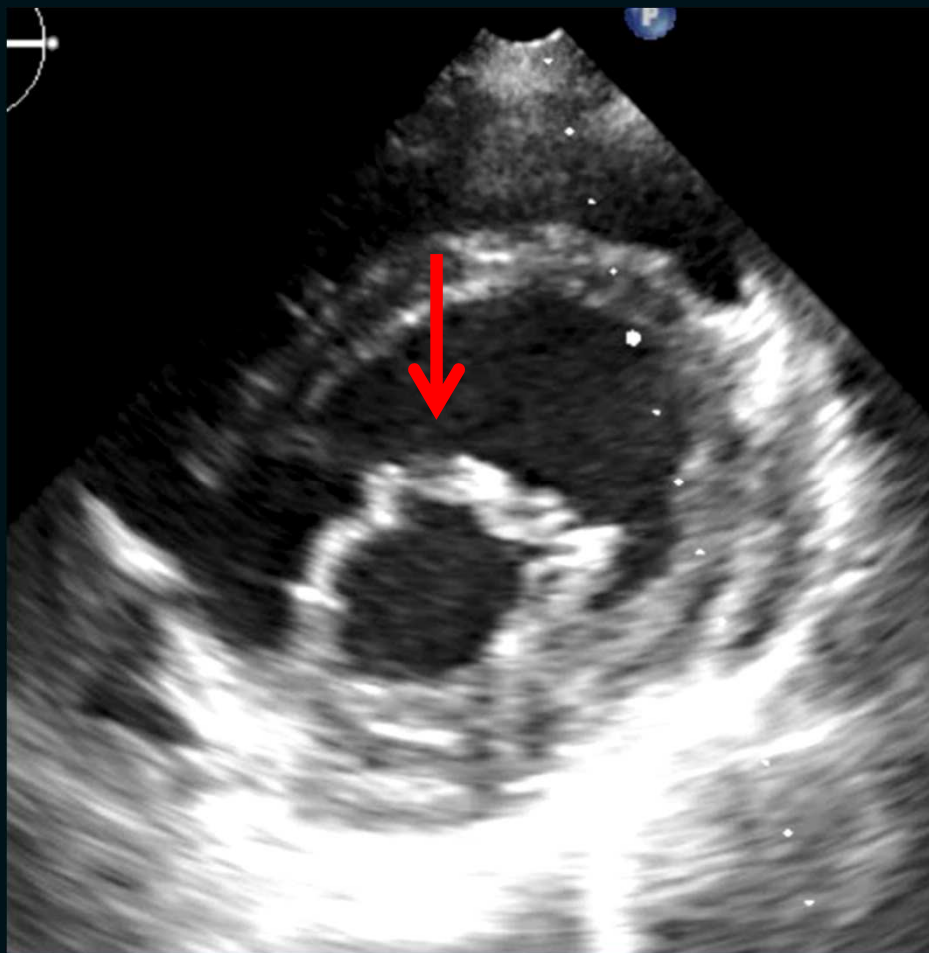
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## LAVVR—Residual “Cleft”

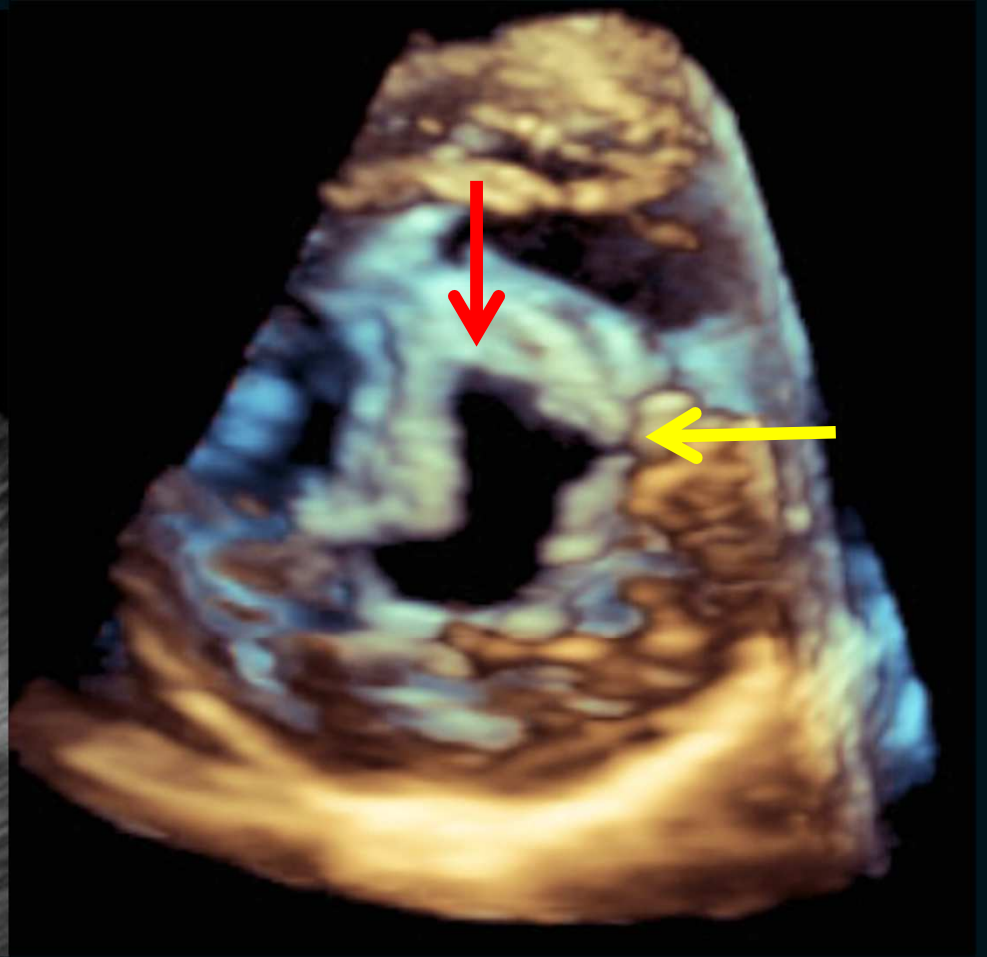
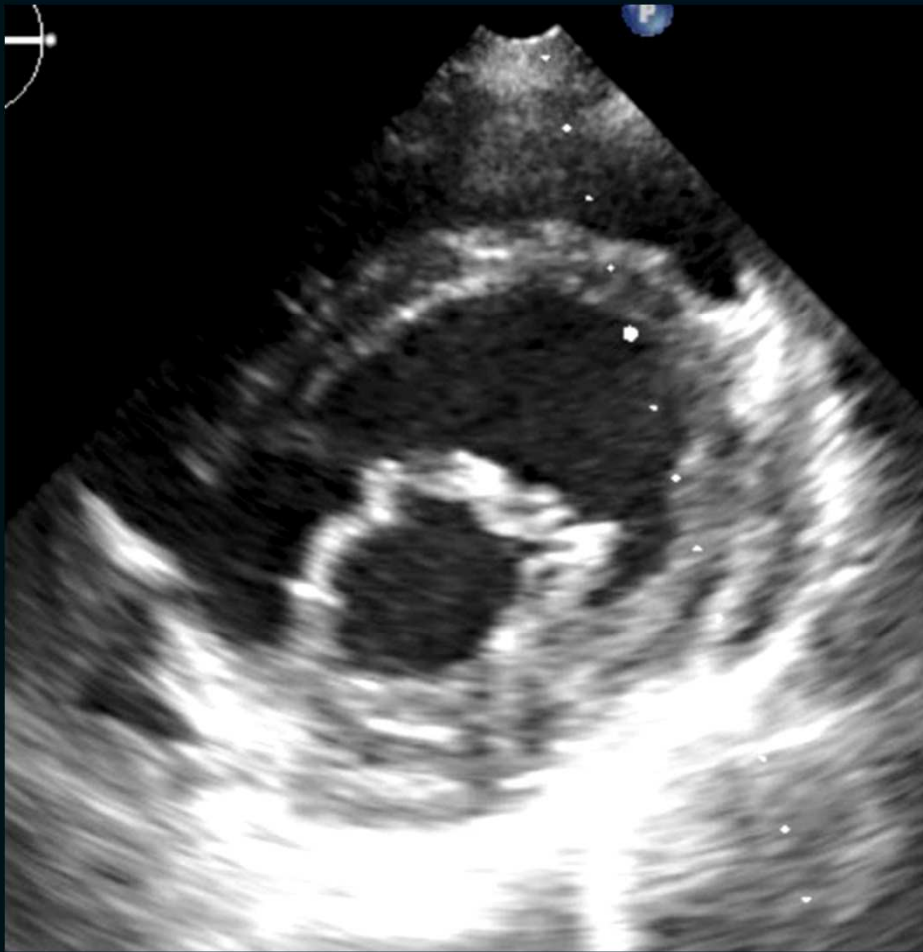


## LAVVR—Residual “Cleft”



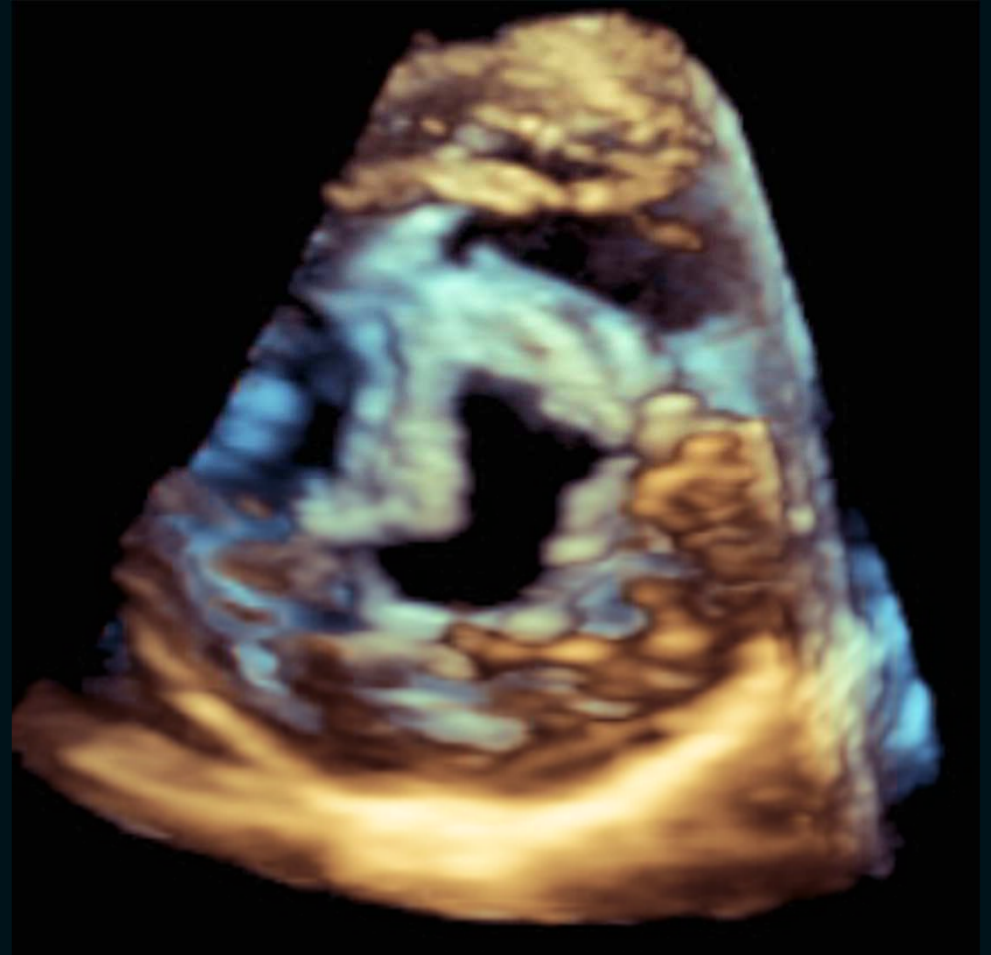


## LAVVR—Residual “Cleft”

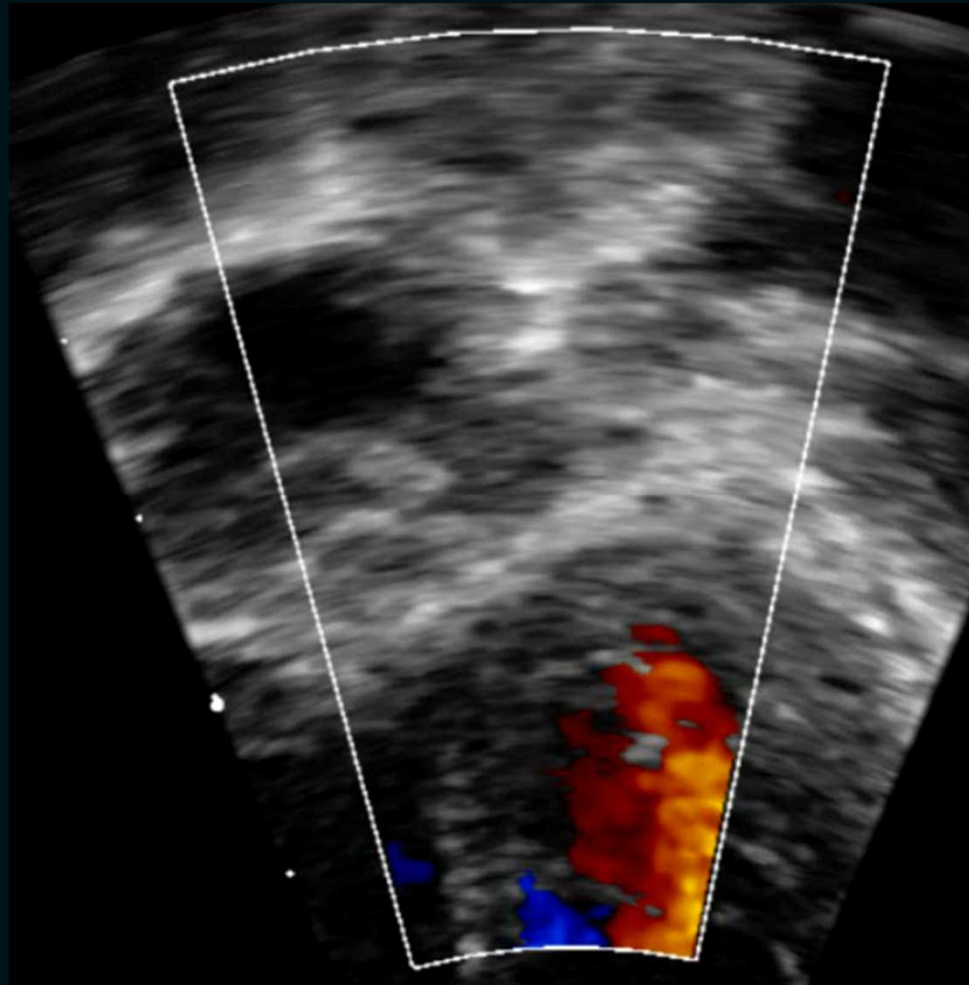




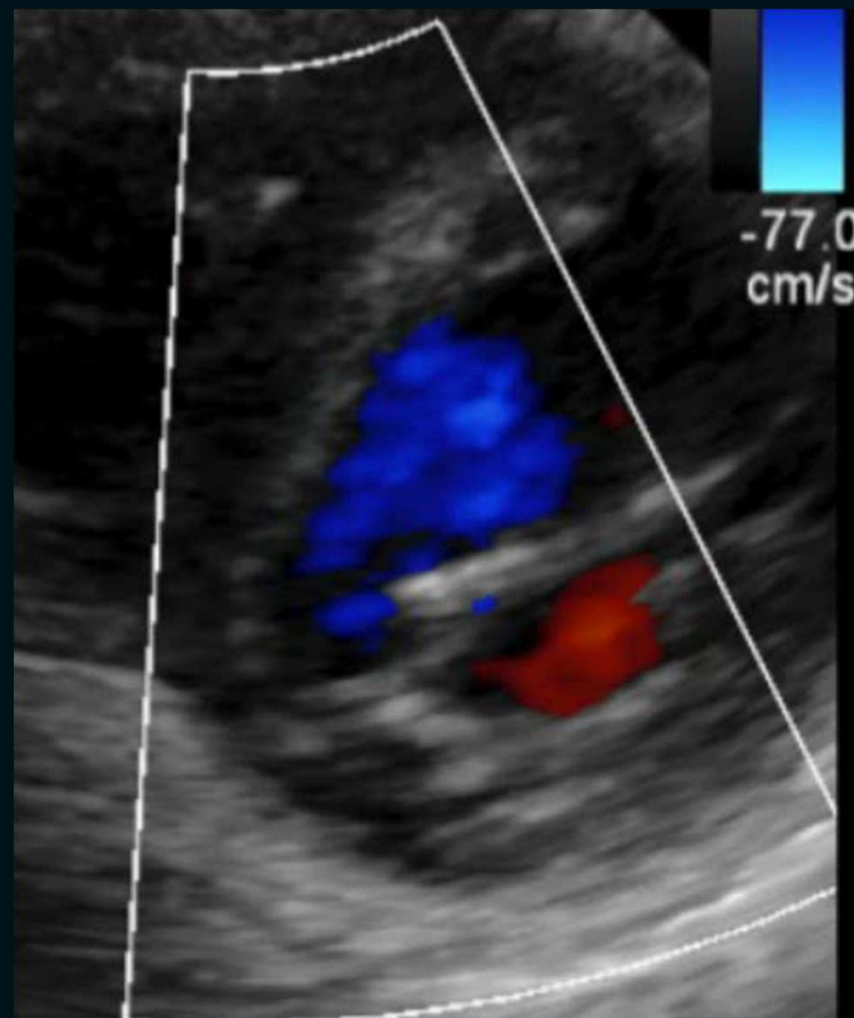
## LAVVR—Residual “Cleft”



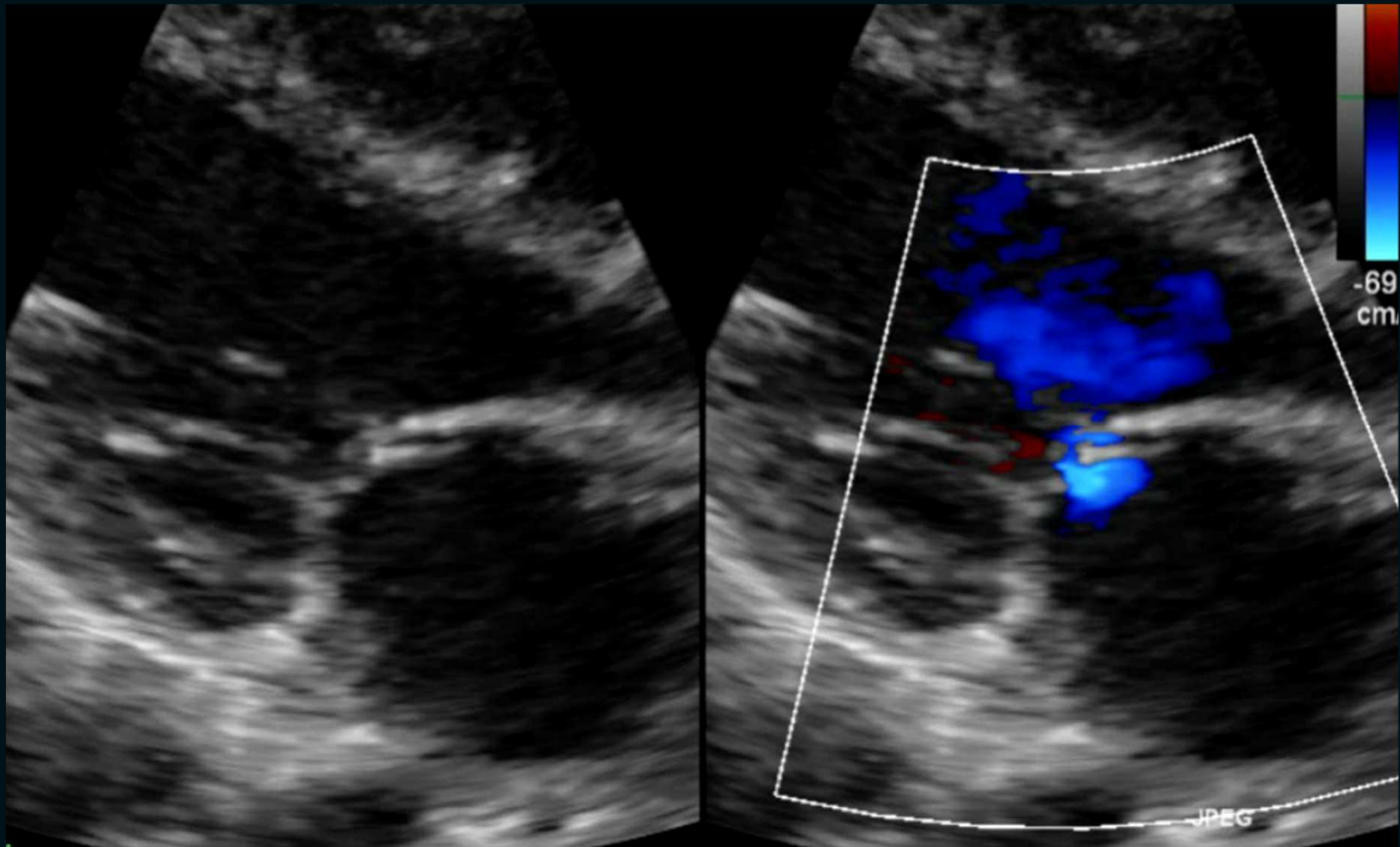
# LAVVR—"Cleft" Closed



# LAVVR—"Cleft" Closed



# LAVVR—"Cleft" Closed



# Left Ventricular Outflow Tract Obstruction



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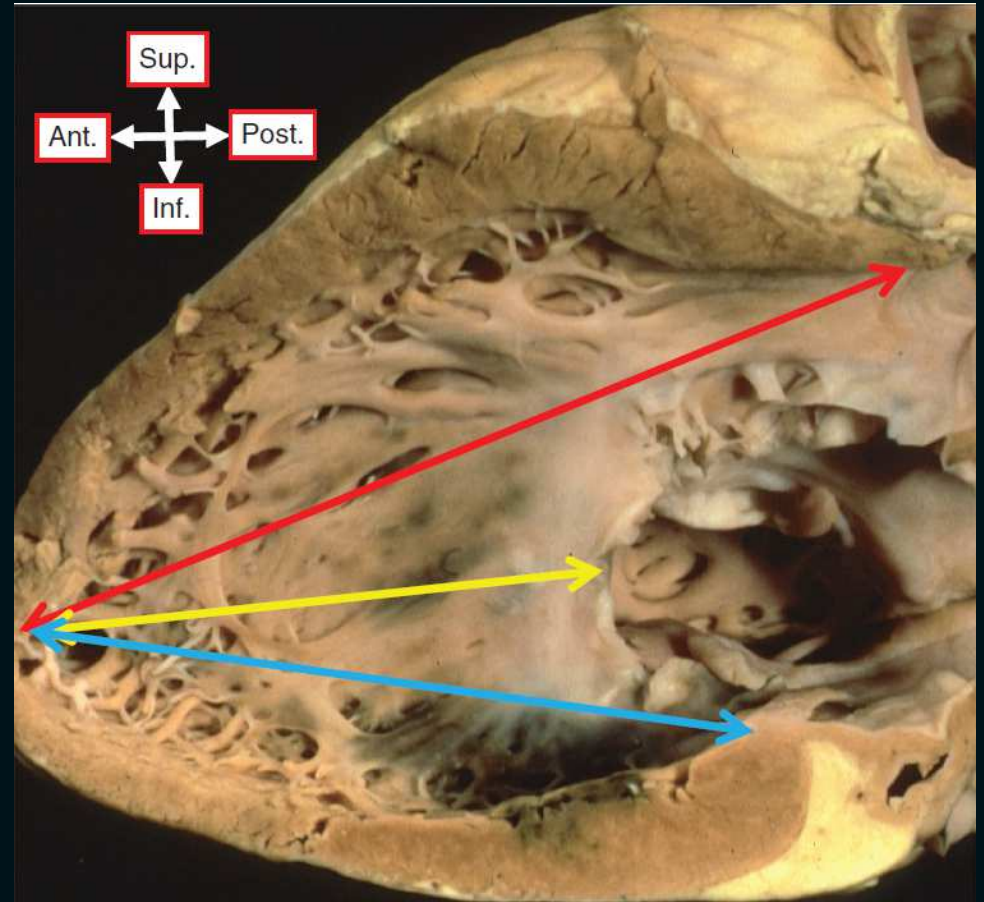
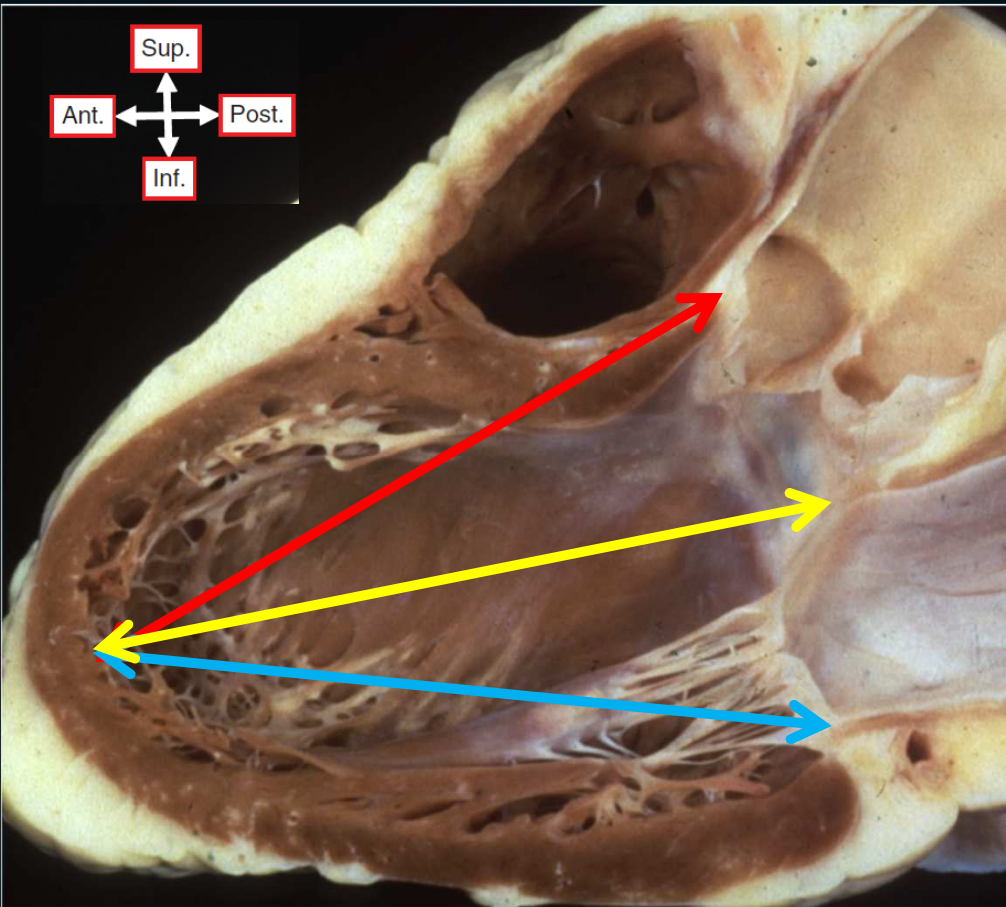
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# LV Outflow Tract Obstruction

- ▶ Between 1-6% require surgery for LVOTO after primary repair
  - ▶ 5-12 yr after AVSD repair
- ▶ Often progressive
- ▶ Substrate:
  - ▶ “Unwedging” of the aortic root
  - ▶ “Scooped out” muscular septum
  - ▶ Chordal attachments in LVOT



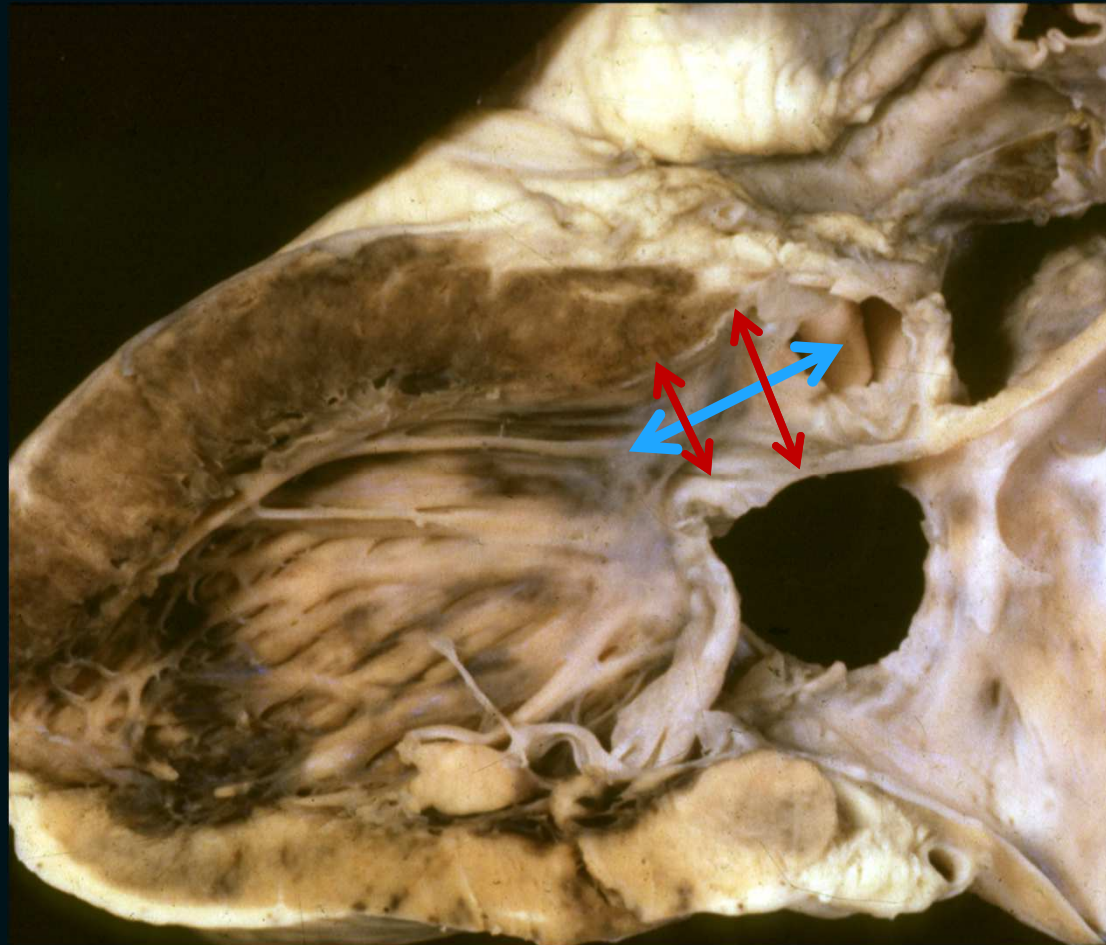
# LVOT Obstruction Substrate



From Anderson, Spicer, Hlavacek, et al. "Wilcox Surgical Anatomy of the Heart", 4<sup>th</sup> Ed. 2013

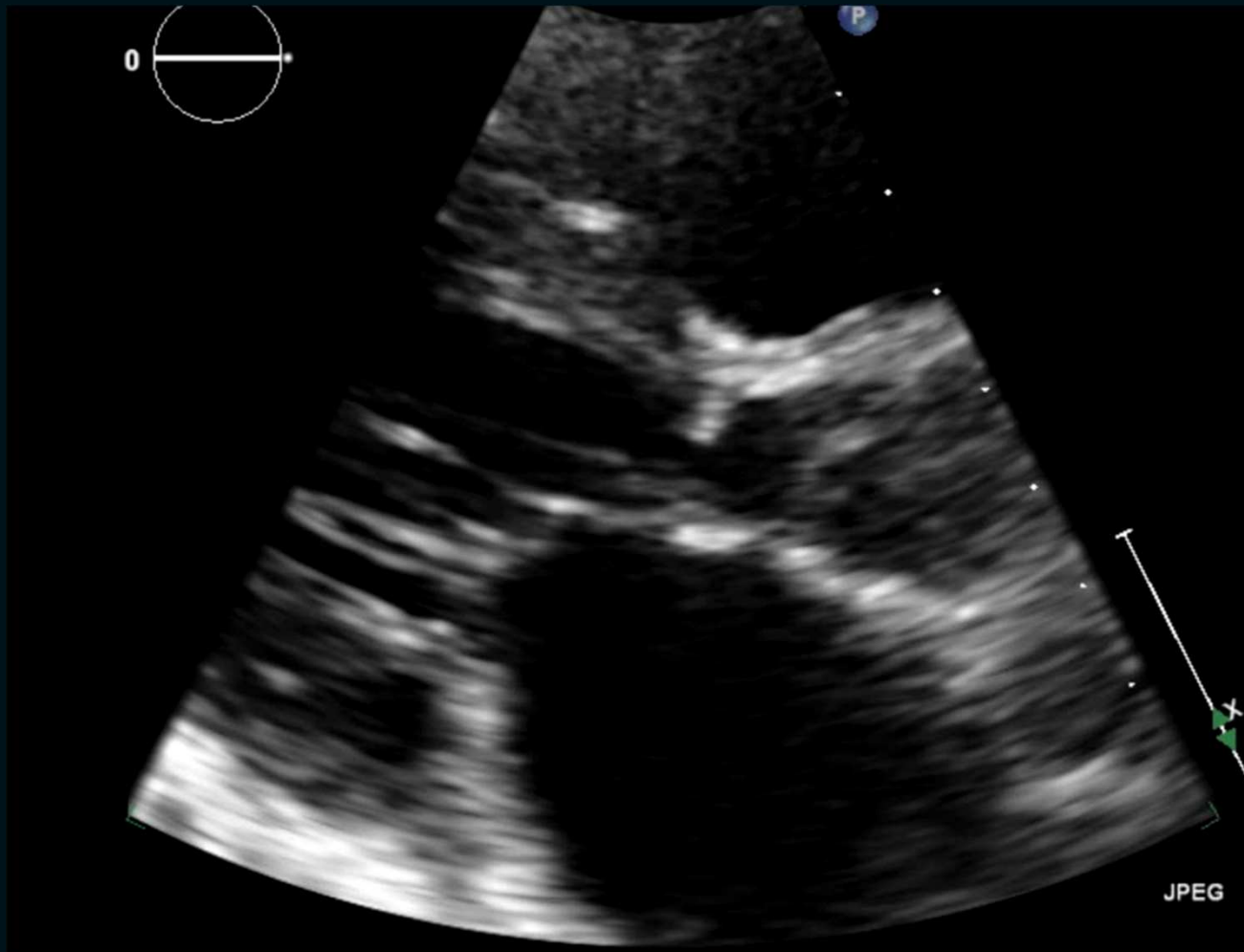


# LVOT Obstruction Substrate

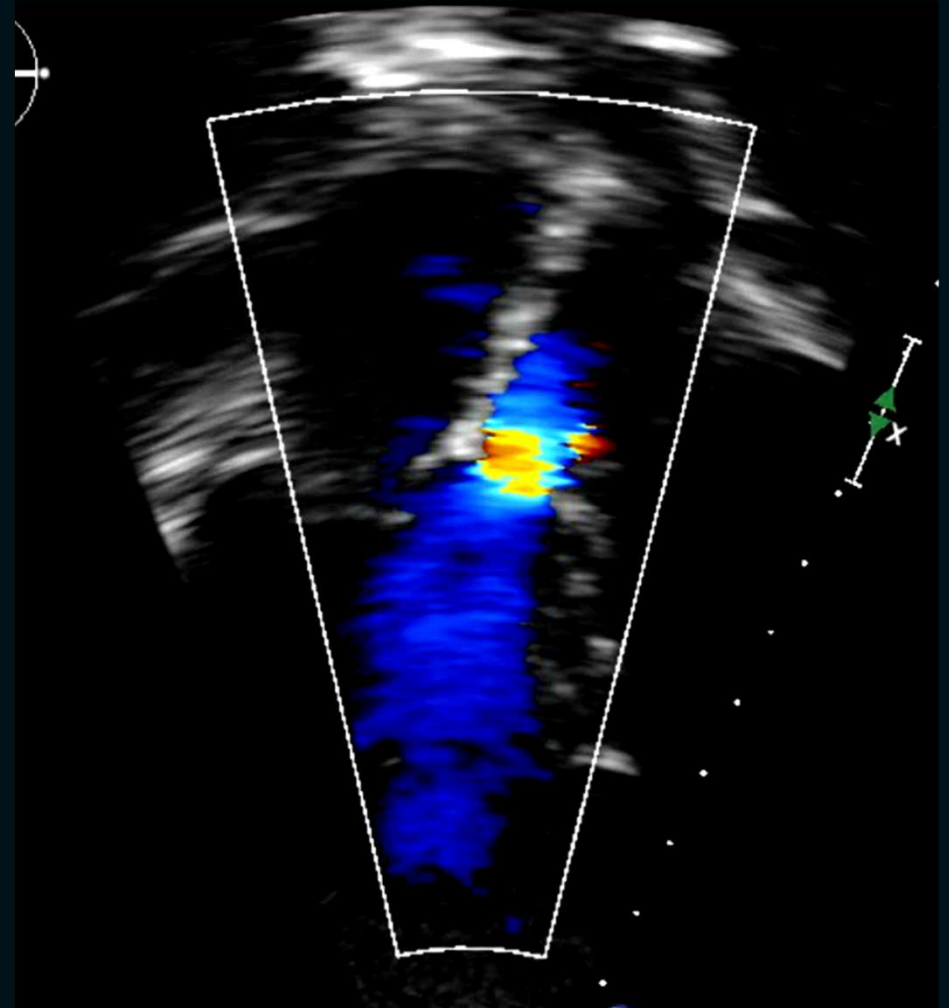
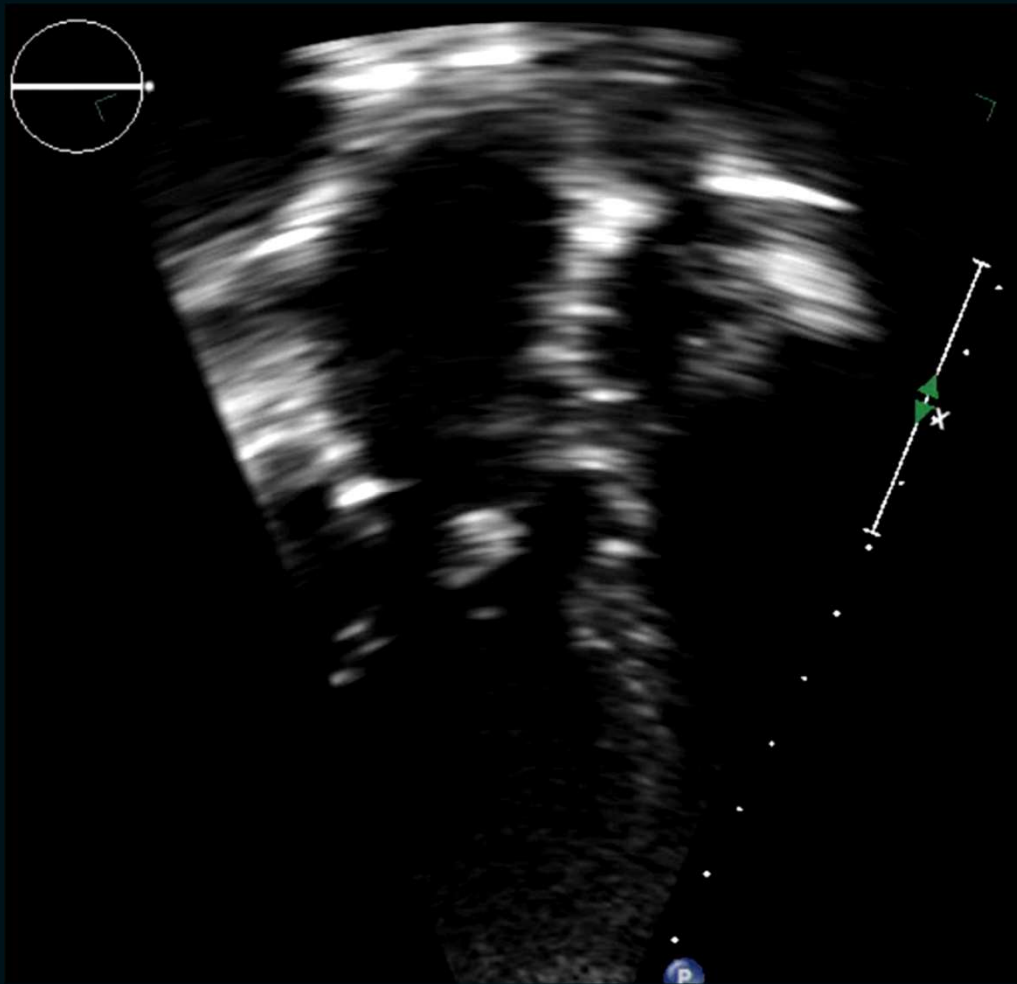


From Anderson, Spicer, Hlavacek, et al. "Wilcox Surgical Anatomy of the Heart", 4<sup>th</sup> Ed. 2013

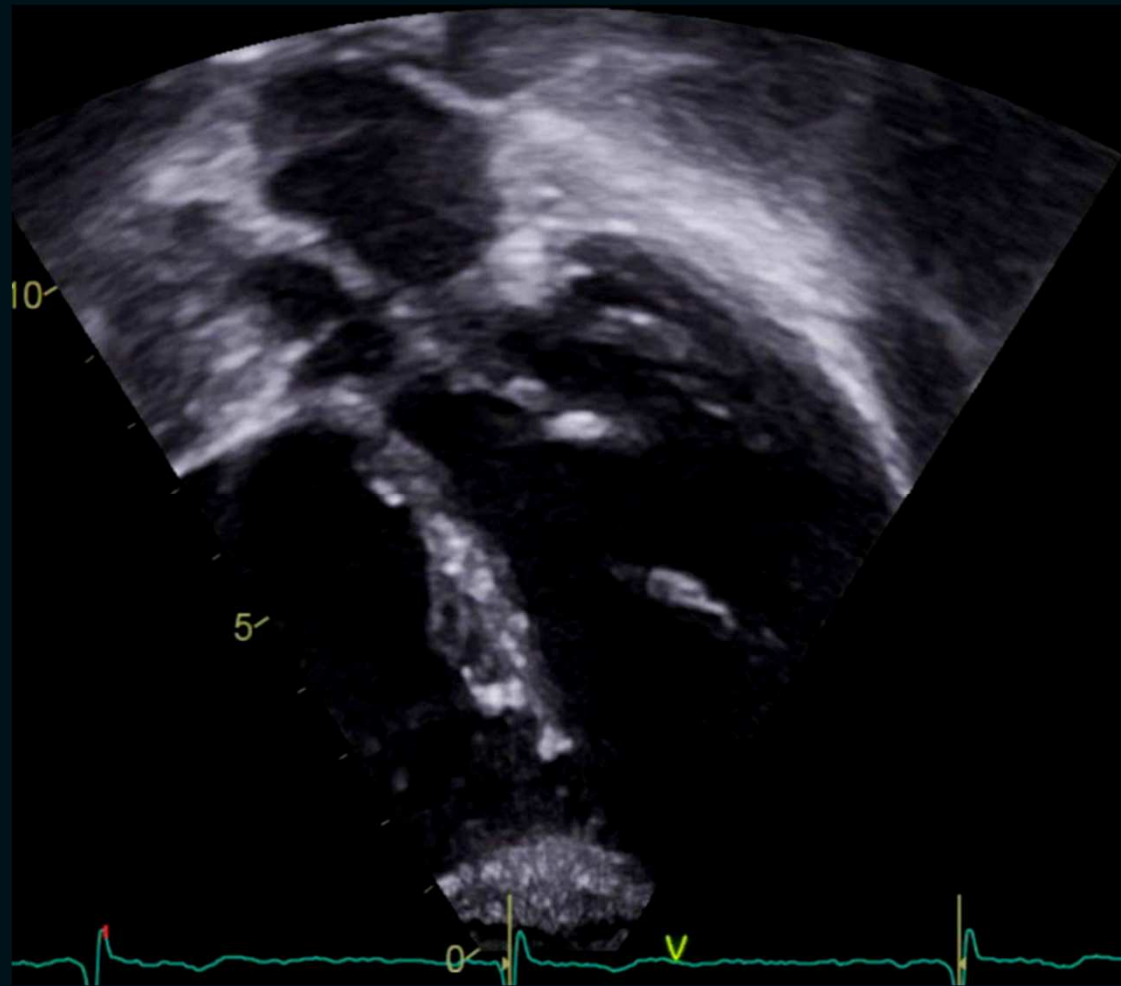
# LVOTO-Discrete Membrane



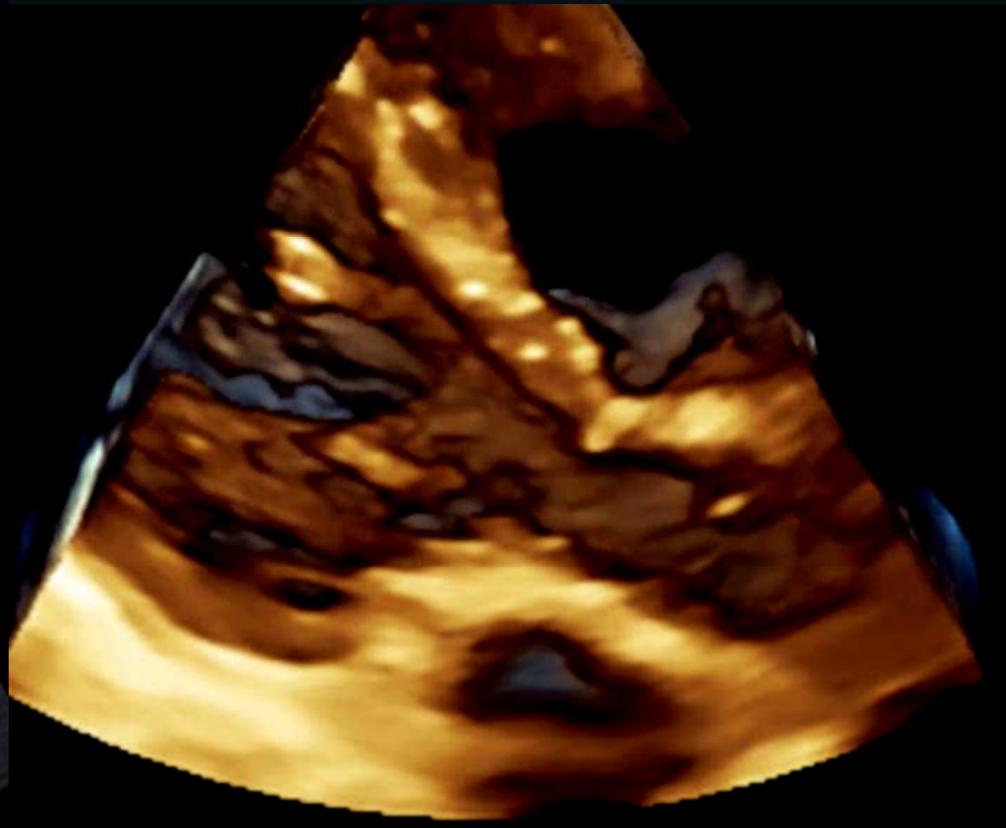
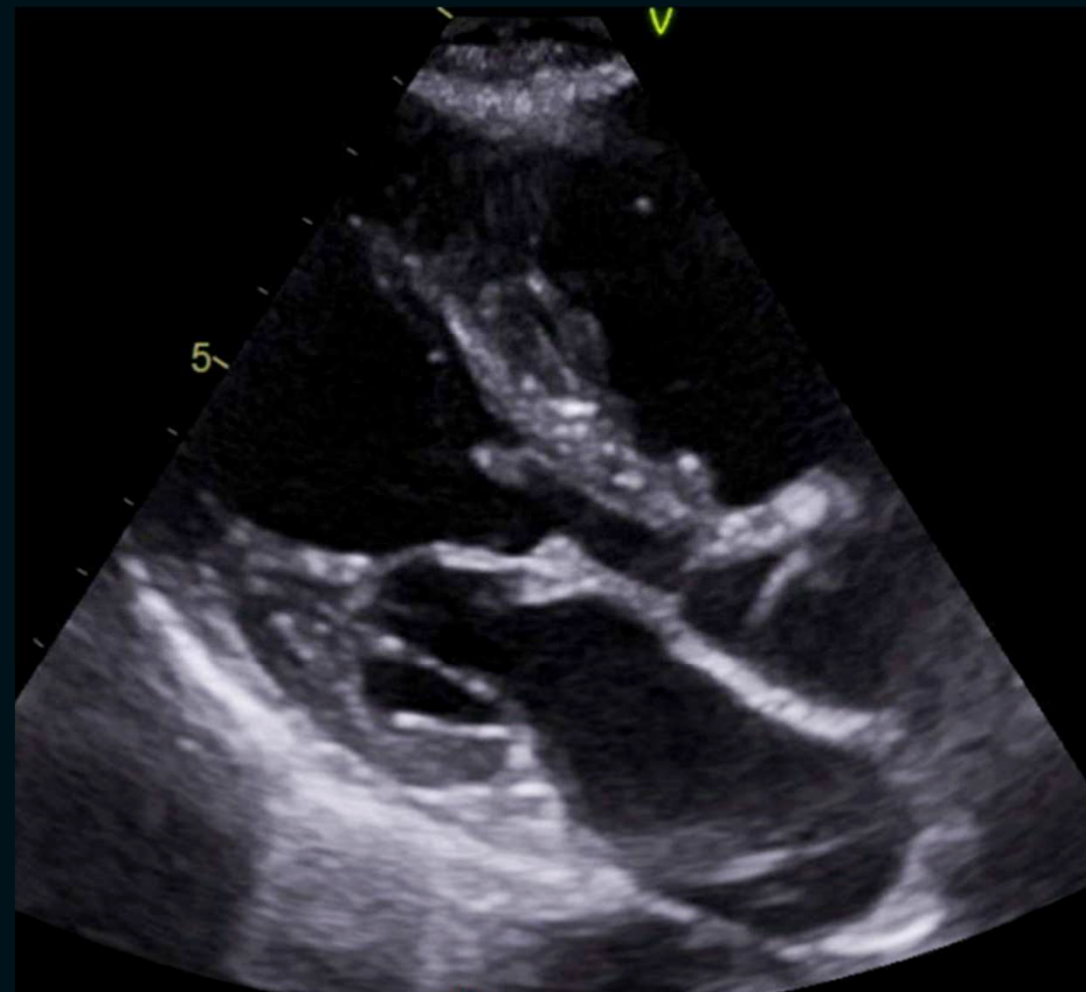
# LVOTO-Discrete Membrane



# LVOTO-Chordae



# LVOTO-Chordae

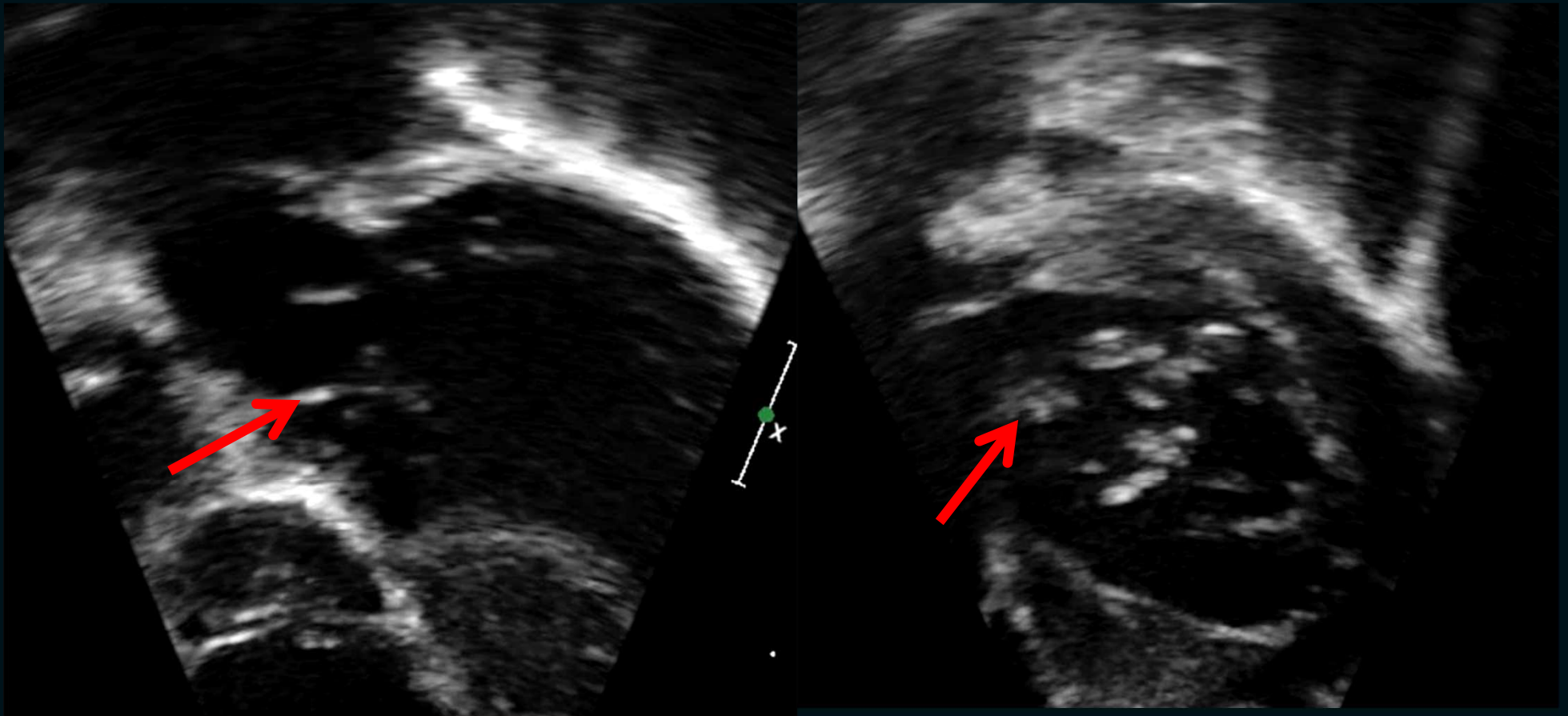




# LVOTO-Chordae

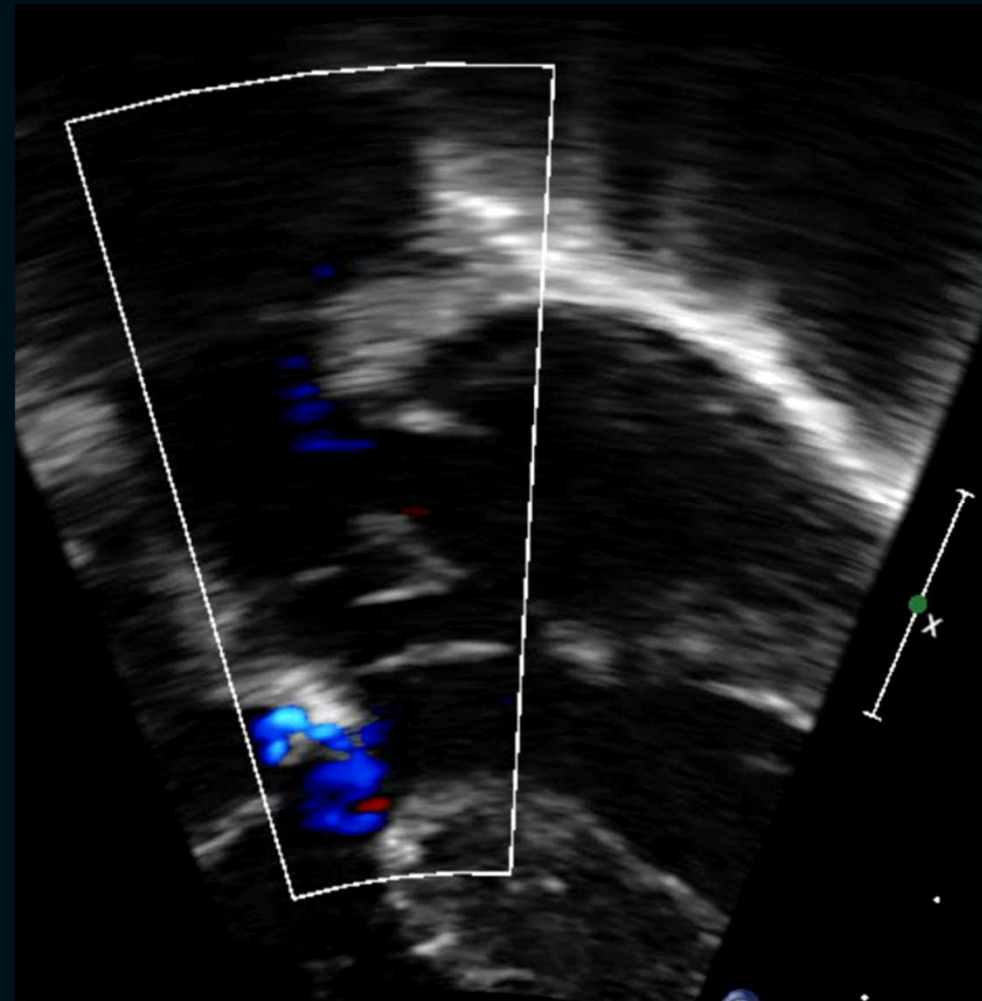
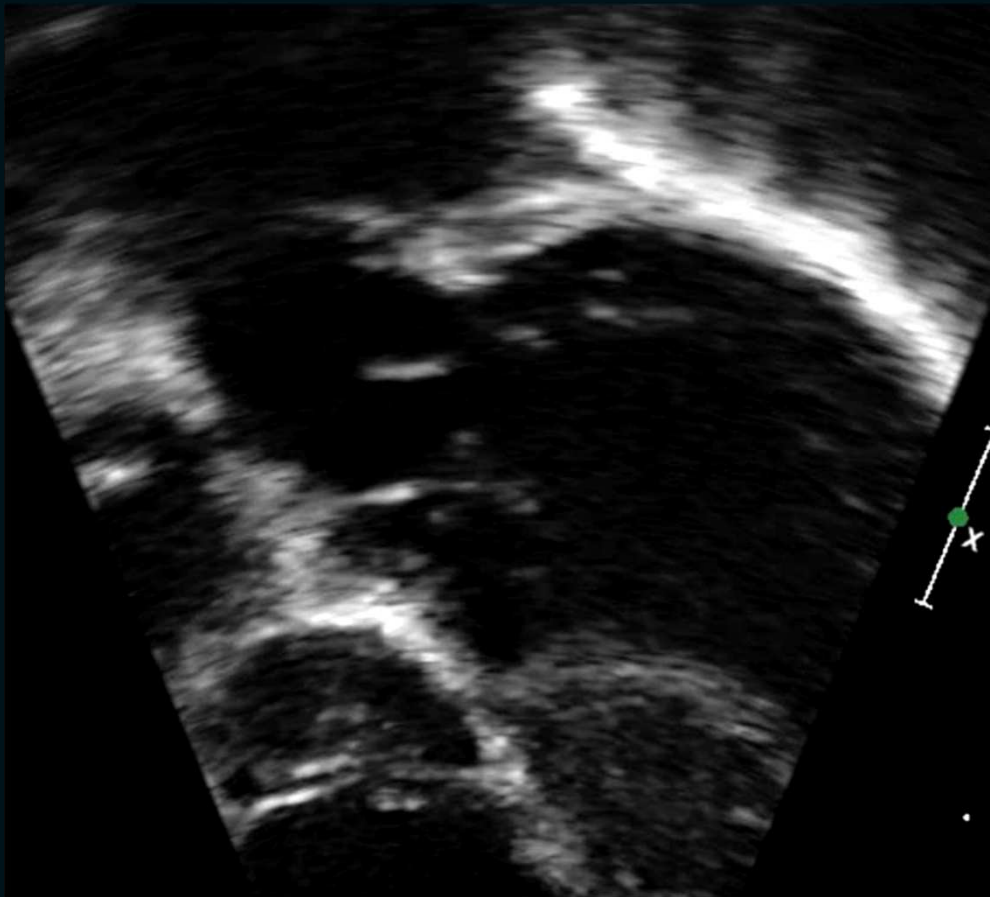


# LVOTO-Chordae and Muscular





# LVOTO-Chordae and Muscular



# Conclusion

- ▶ Residual lesions common after AVSD repair
- ▶ Residual ASD/VSD often improve/resolve
- ▶ LAVVR and LVOTO most common issues requiring intervention
  - ▶ LAVVR early, LVOTO later
- ▶ 3D echo can be helpful
- ▶ CMR potentially useful with LAVVR



