AORTIC VALVE AND IT'S WOES

Date: April 27th 2019

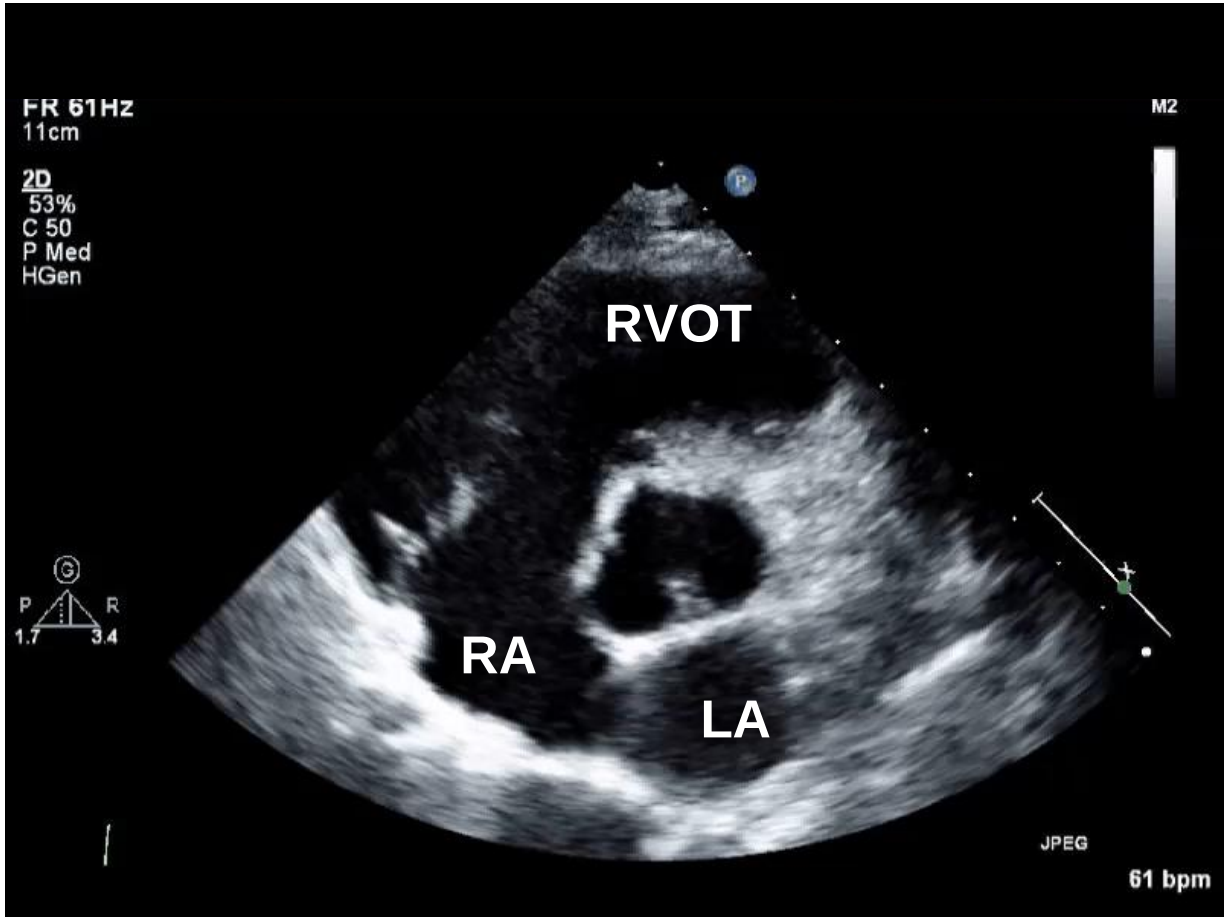
Author: Salima Bhimani
Author Title: PGY-IV, Pediatric Cardiology

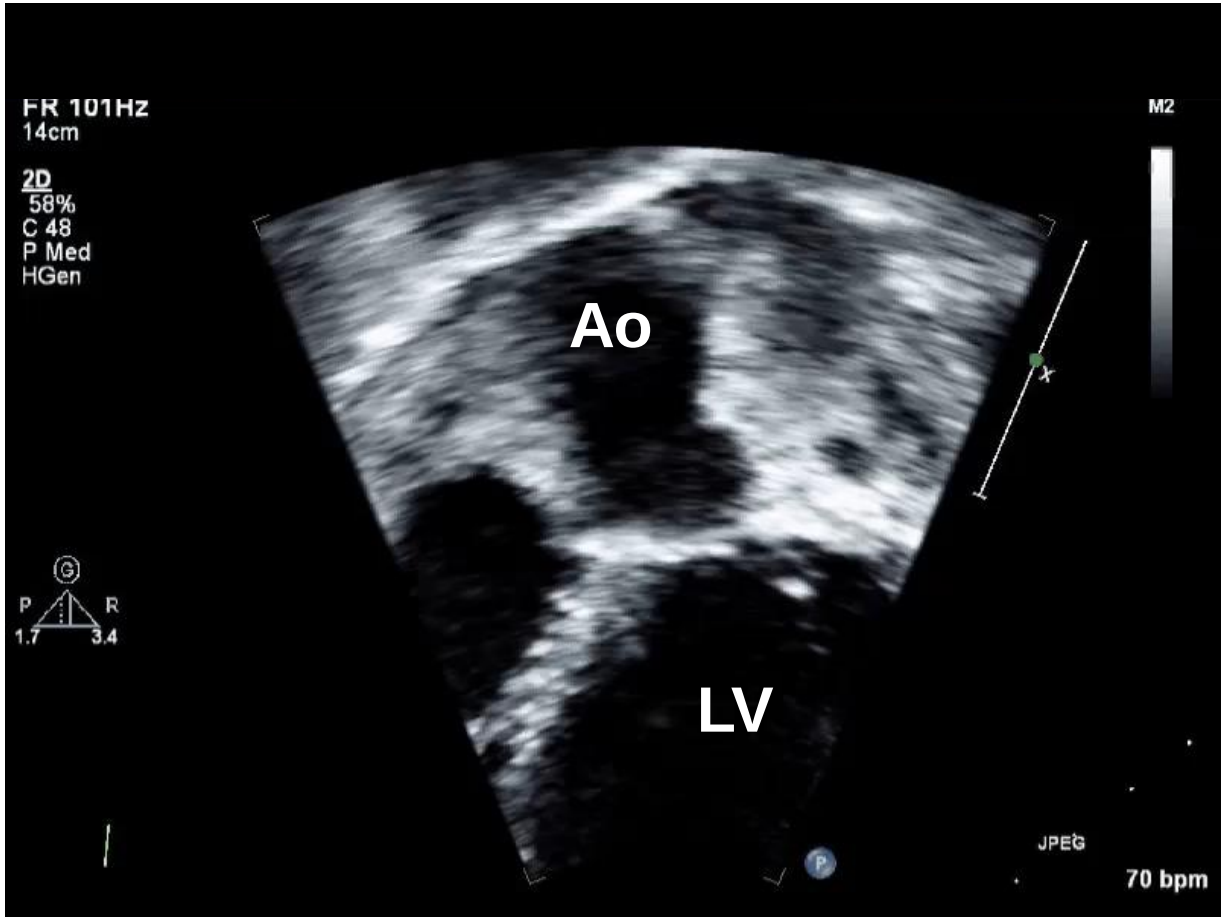
Cleveland Clinic Children's

Clinical Presentation

- 11 y/o male with Osteogenesis Imperfecta and known bicuspid aortic valve with moderate AS and AR, progressive ascending aortic dilatation
- presented with weight loss, night sweats and acute onset of intermittent fever
- Unchanged murmur; new onset of tender hepatosplenomegaly
- Blood cultures were positive for *Streptococcus Mutans*
- Due to clinical suspicion of infective endocarditis, an emergent transthoracic echocardiogram (TTE) was performed

Six months ago…

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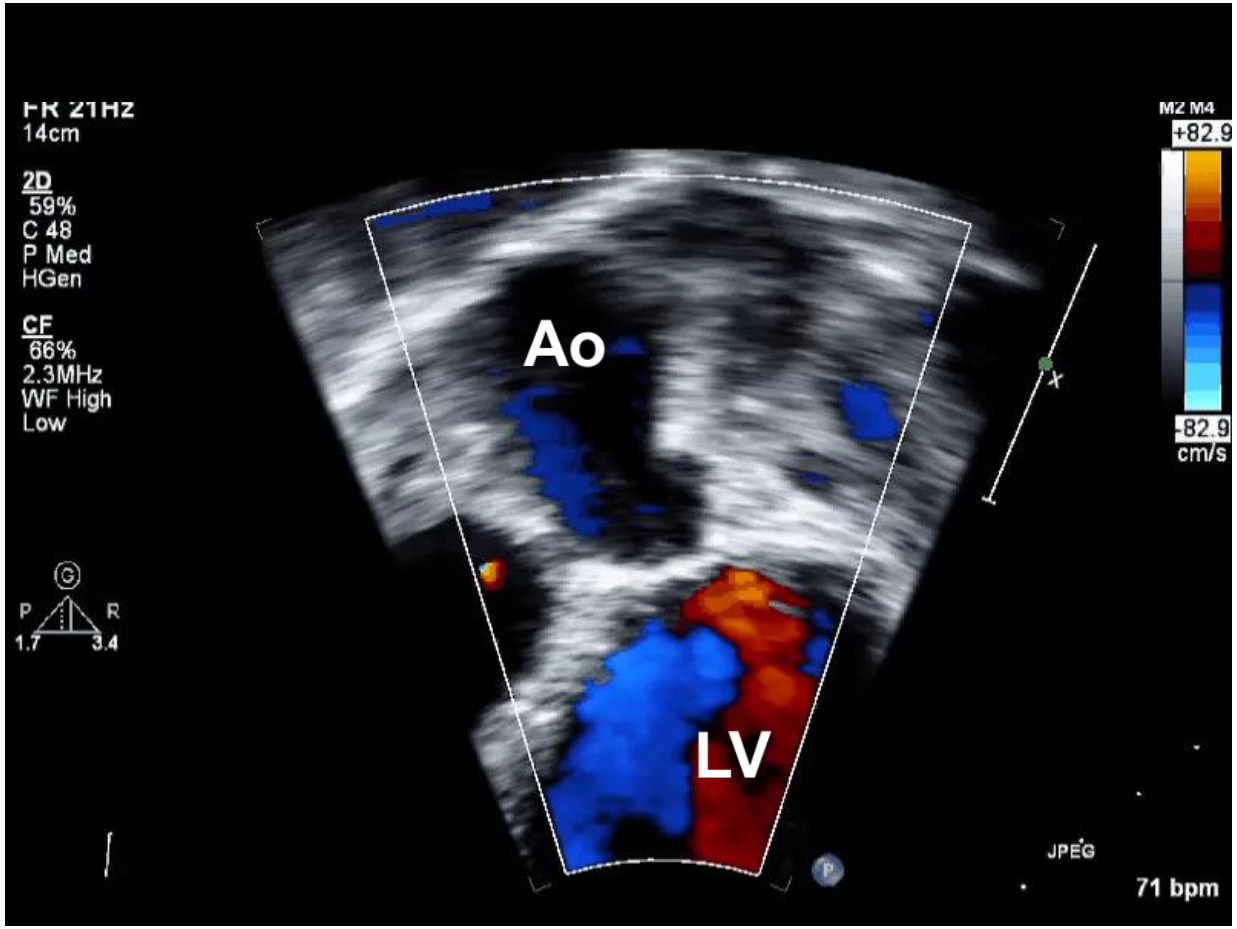
14cm
2D
58%
C 48
P Med
HGen
M2
Ao
LV
P
R
1.7
3.4
JPEG
70 bpm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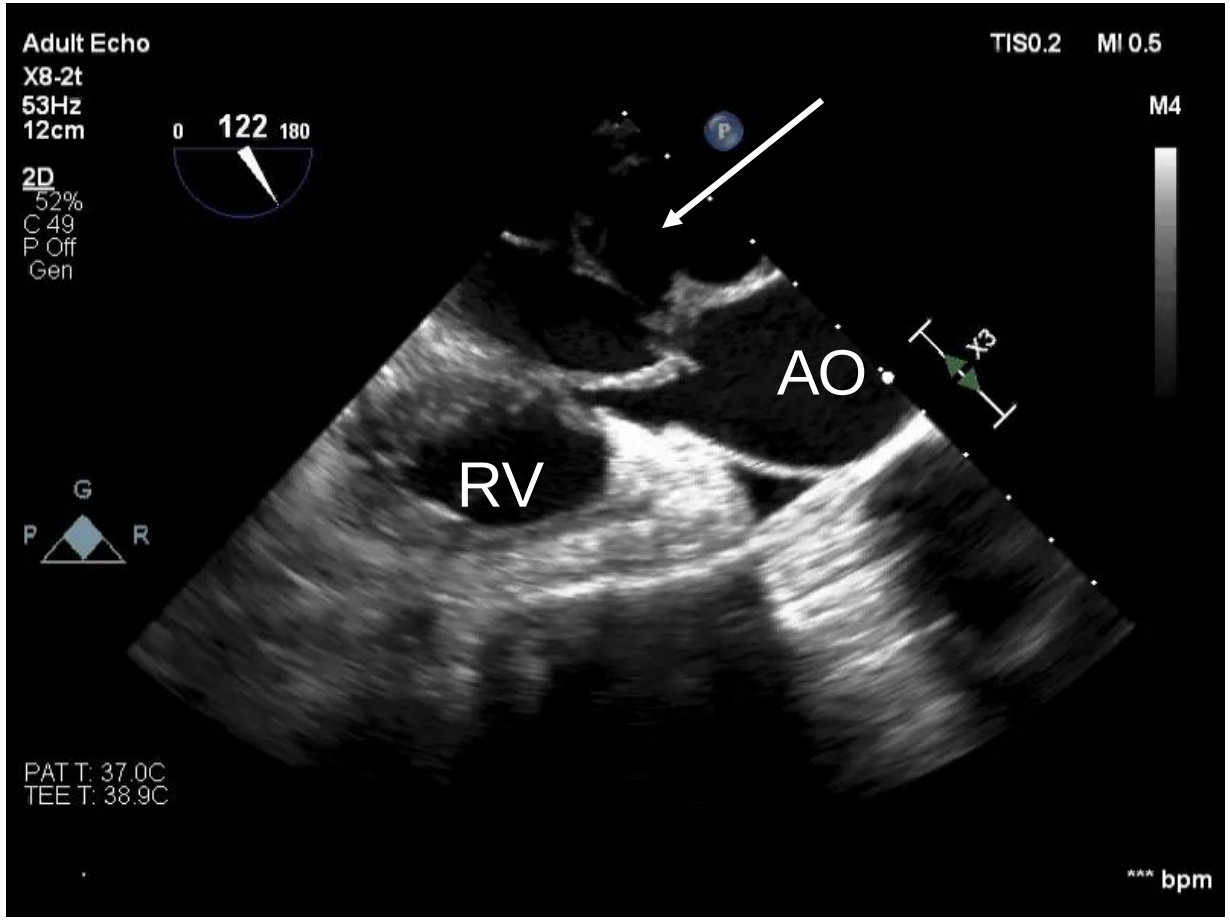
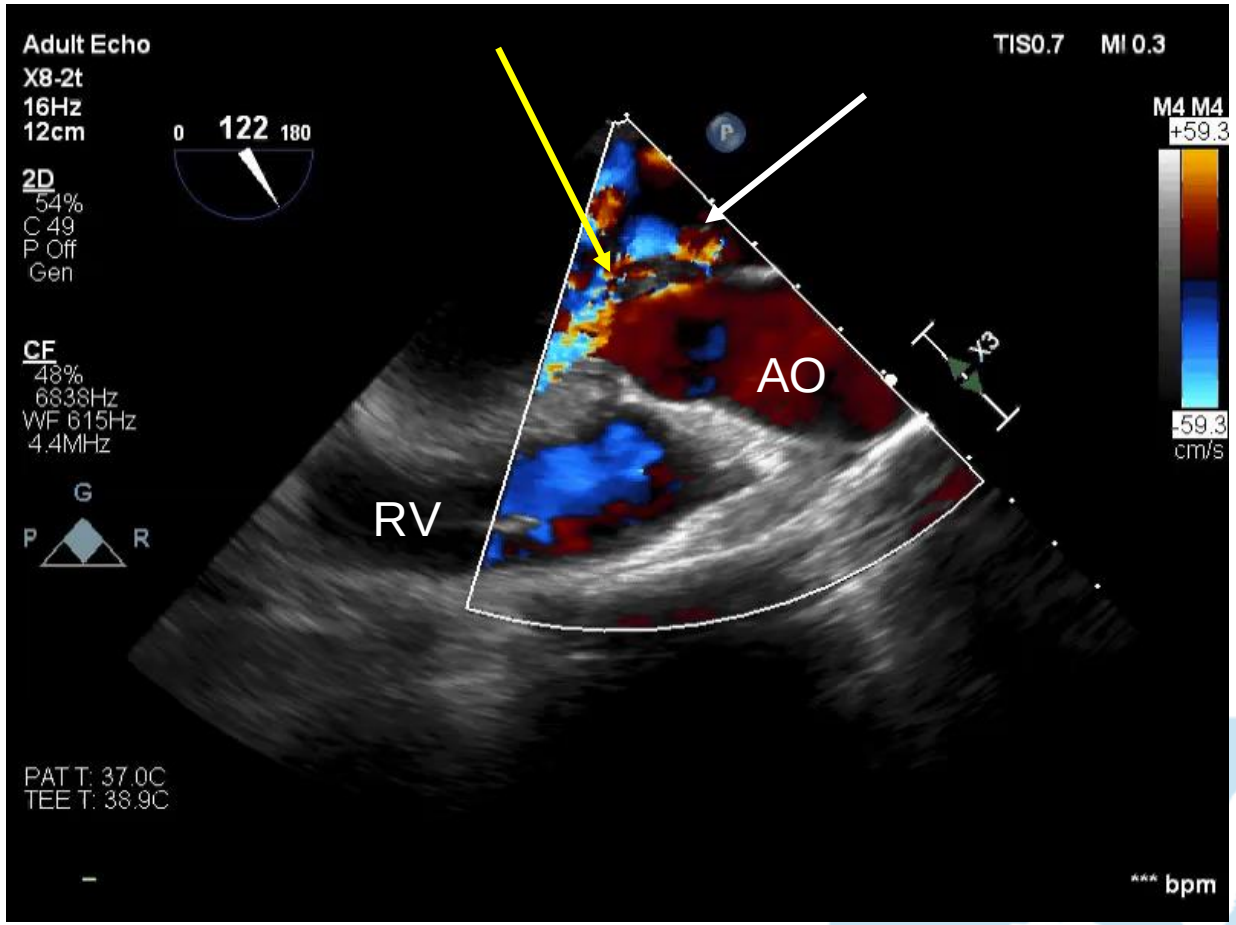
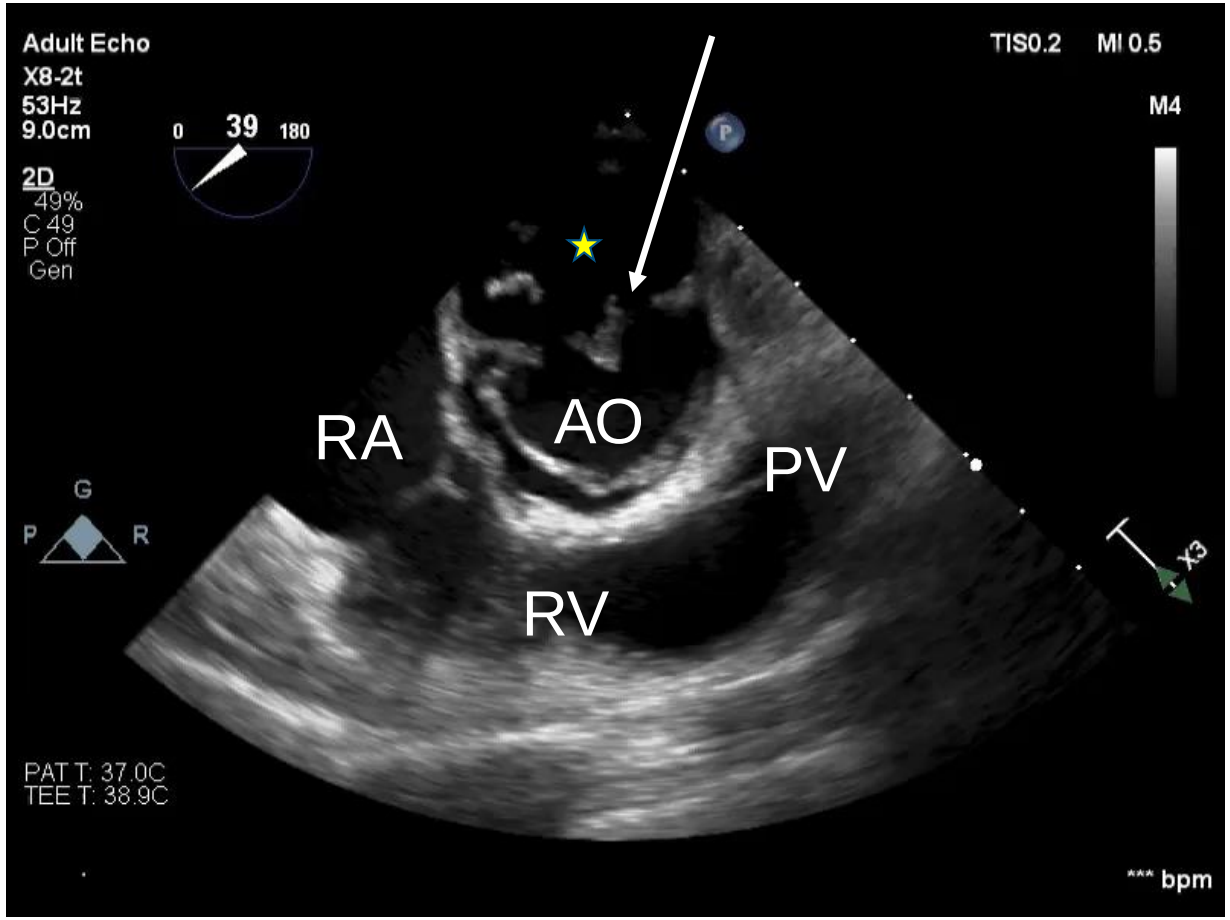
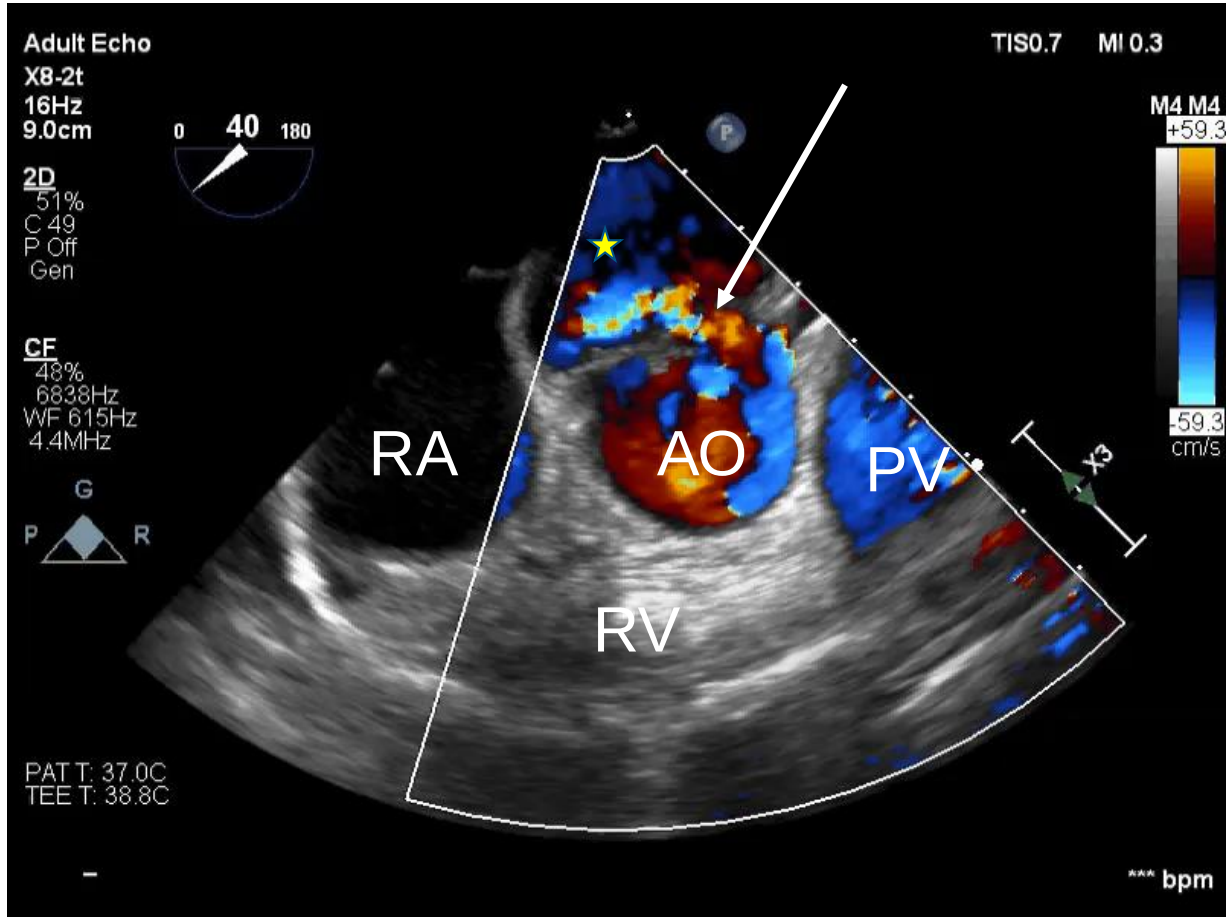
14cm
2D
59%
C 48
P Med
HGen
CF
66%
2.3MHz
WF High
Low
M2 M4
+82.9
-82.9
cm/s
Ao
LV
P
R
1.7
3.4
JPEG
71 bpm

Six months later: PREOP TEE

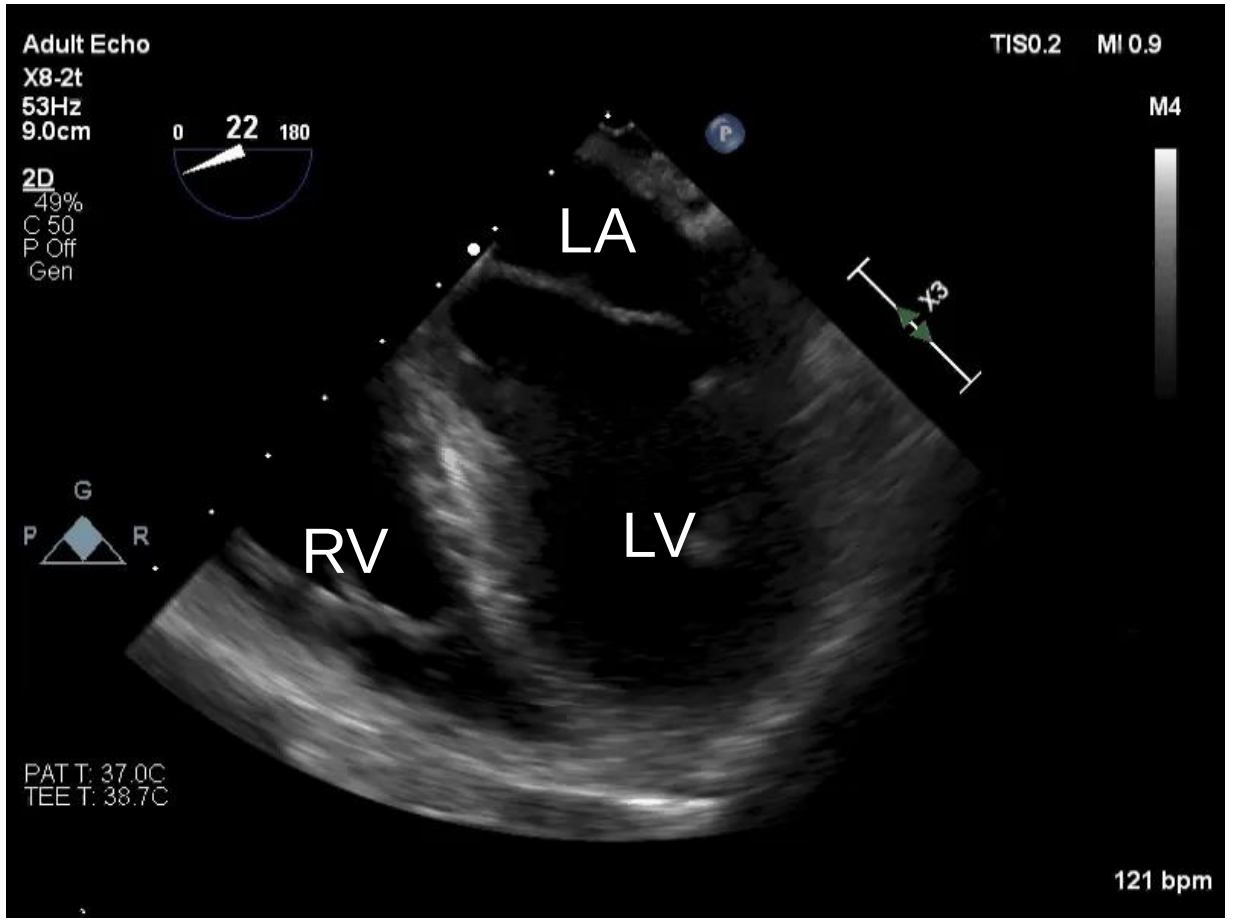




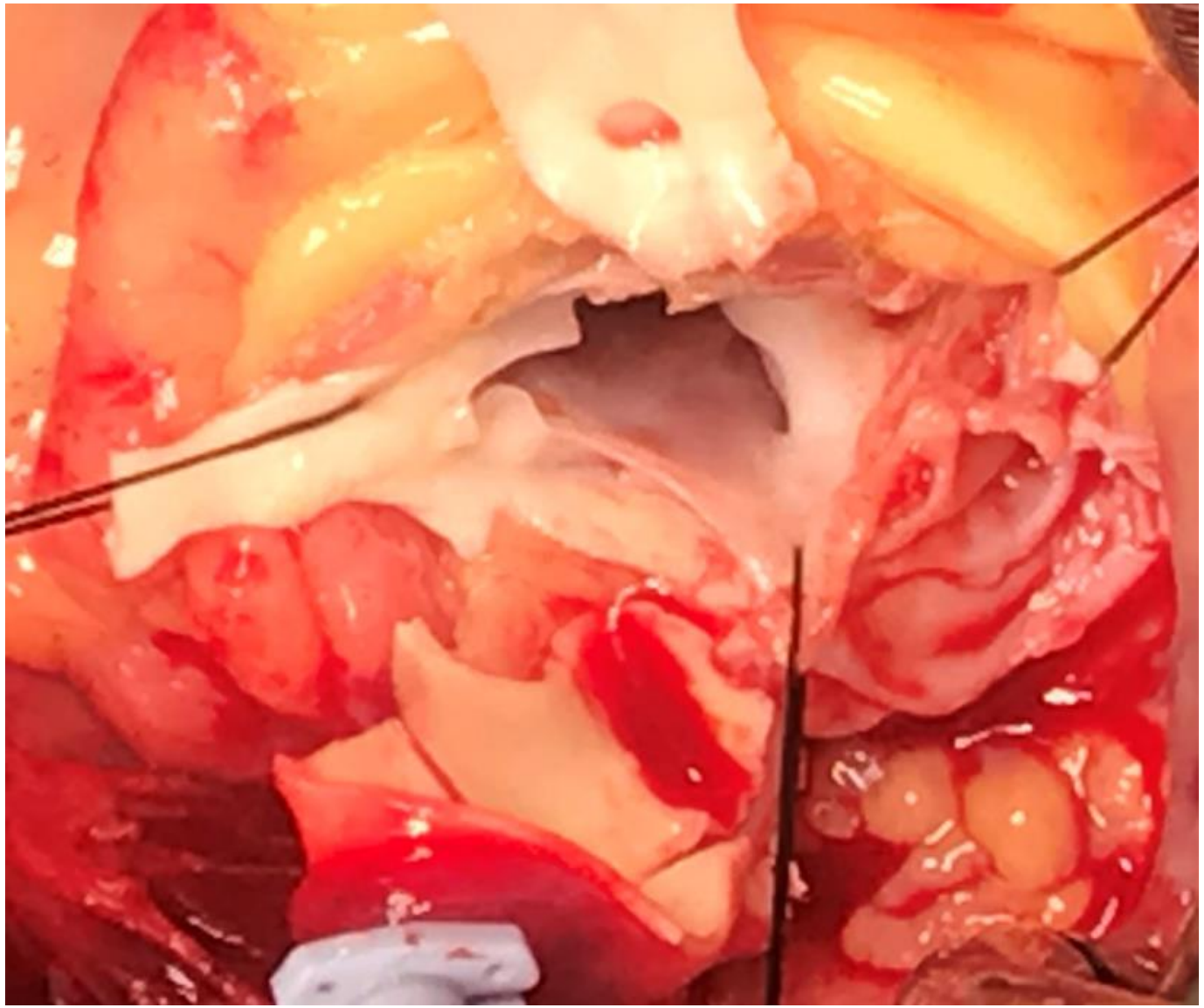

Adult Echo
X8-2t
53Hz
9.0cm
2D
49%
C 49
P Off
Gen
TIS0.2
MI 0.5
M4
RA
AO
PV
RV
PAT T: 37.0C
TEE T: 38.9C
*** bpm


Adult Echo
X8-2t
16Hz
9.0cm
2D
51%
C 49
P Off
Gen
CF
48%
683.8Hz
WF 615Hz
4.4MHz
TIS0.7
MI 0.3
M4 M4
+59.3
-59.3
cm/s
RA
AO
PV
RV
PAT T: 37.0C
TEE T: 38.8C
*** bpm

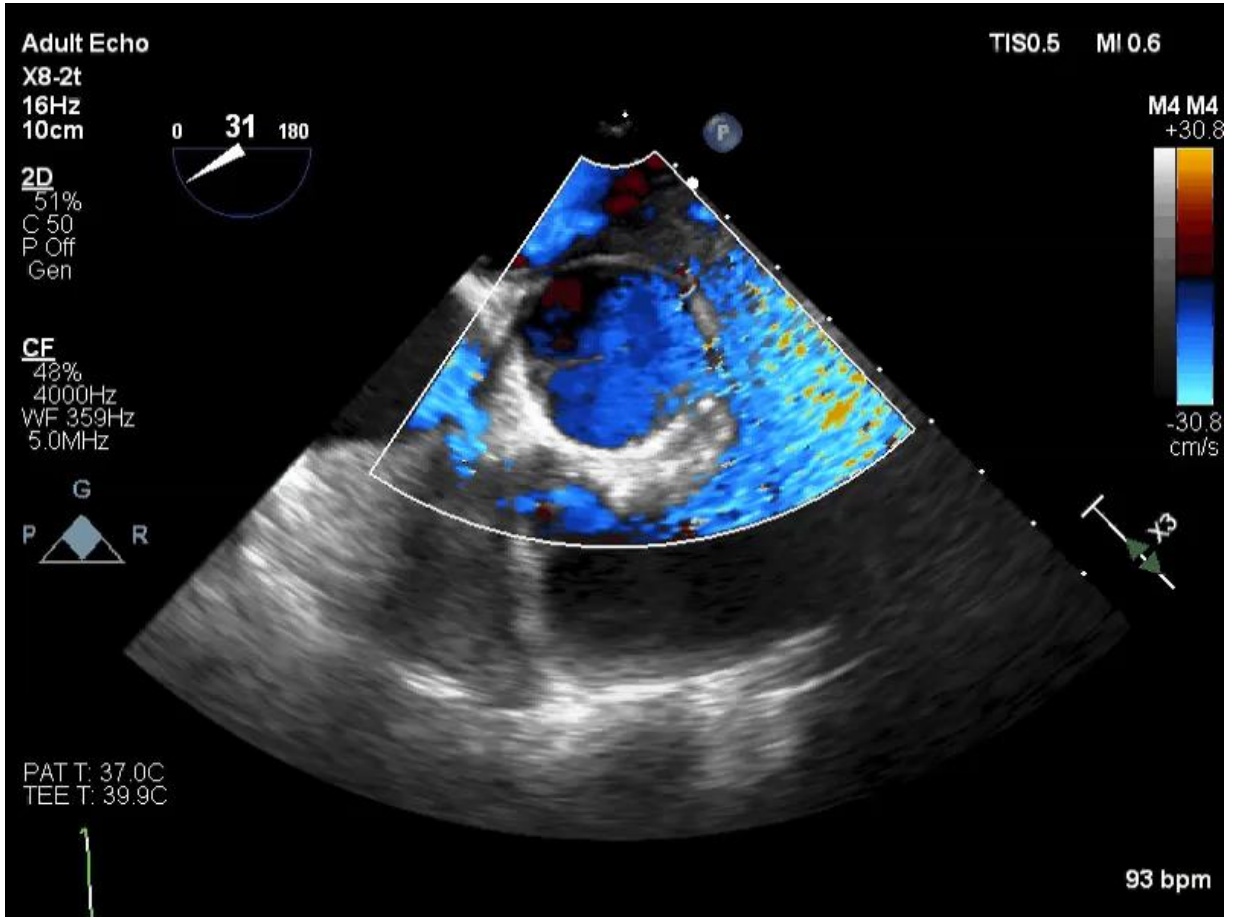
TEE

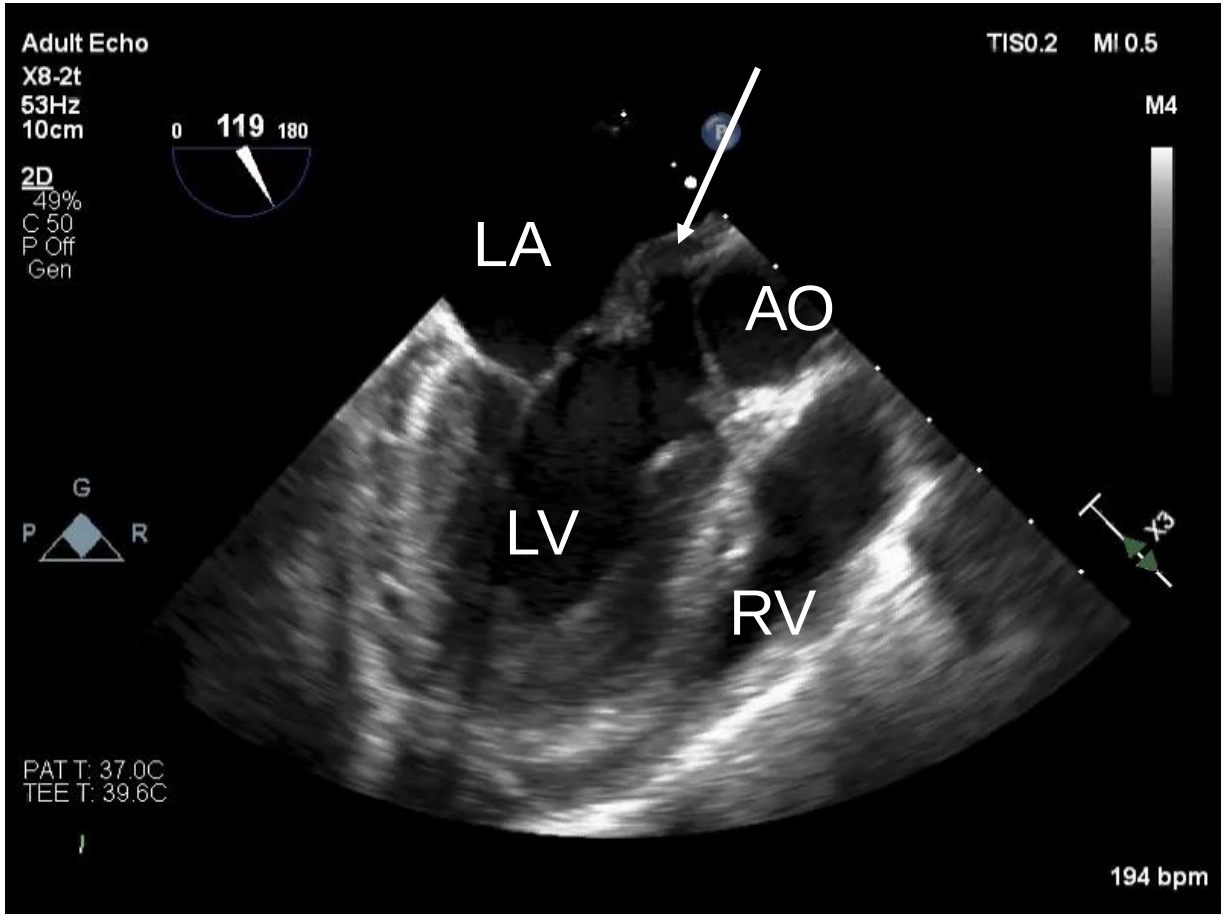

Adult Echo
X8-2t
53Hz
9.0cm
2D
49%
C 50
P Off
Gen
0
22
180
TIS0.2
MI 0.9
M4
LA
X3
G
P
R
RV
LV
PAT T: 37.0C
TEE T: 38.7C
121 bpm

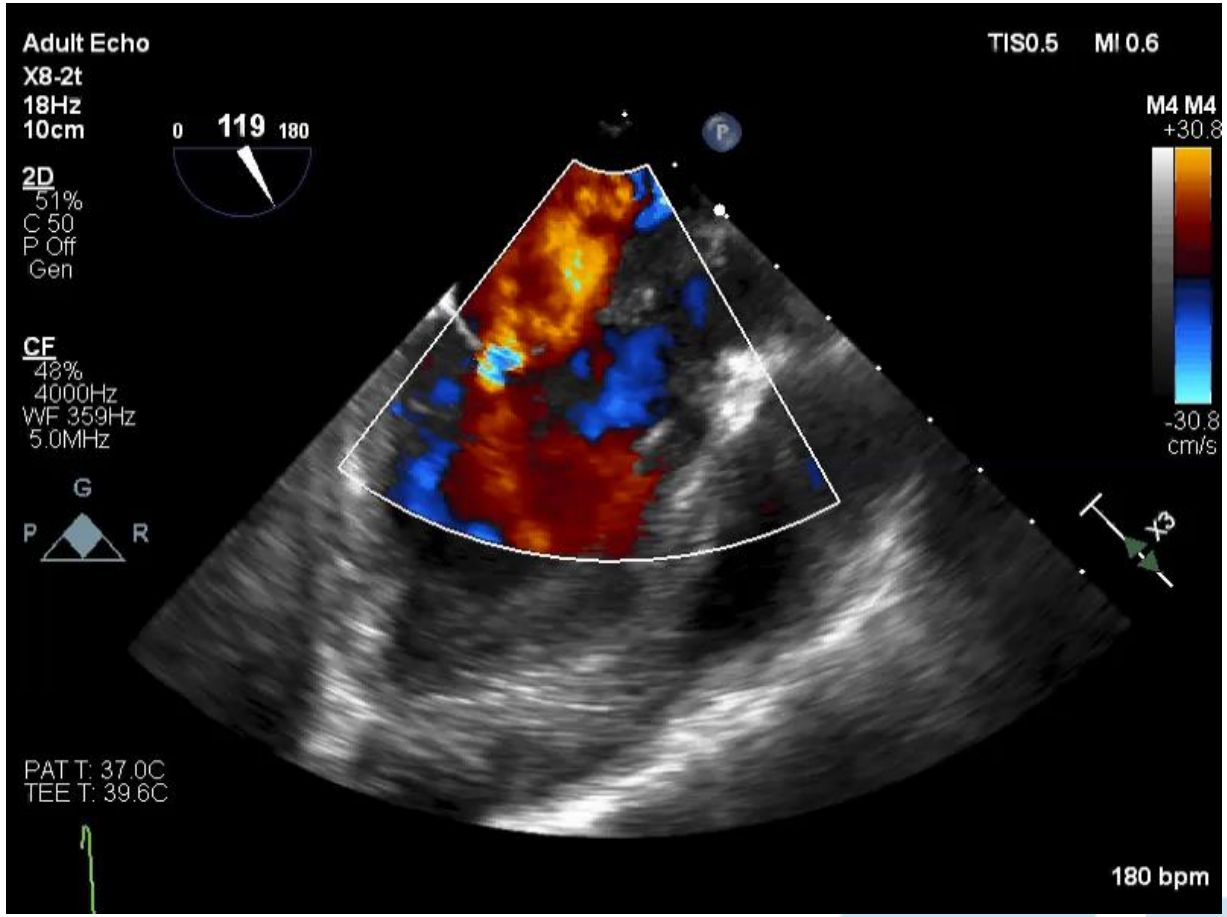
THE REAL DEAL



ALL FIXED


Adult Echo
X8-2t
16Hz
10cm
0
31
180
2D
51%
C 50
P Off
Gen
CF
48%
4000Hz
WF 359Hz
5.0MHz
G
P
R
TIS0.5
MI 0.6
M4 M4
+30.8
-30.8
cm/s
X3
PAT T: 37.0C
TEE T: 39.9C
93 bpm

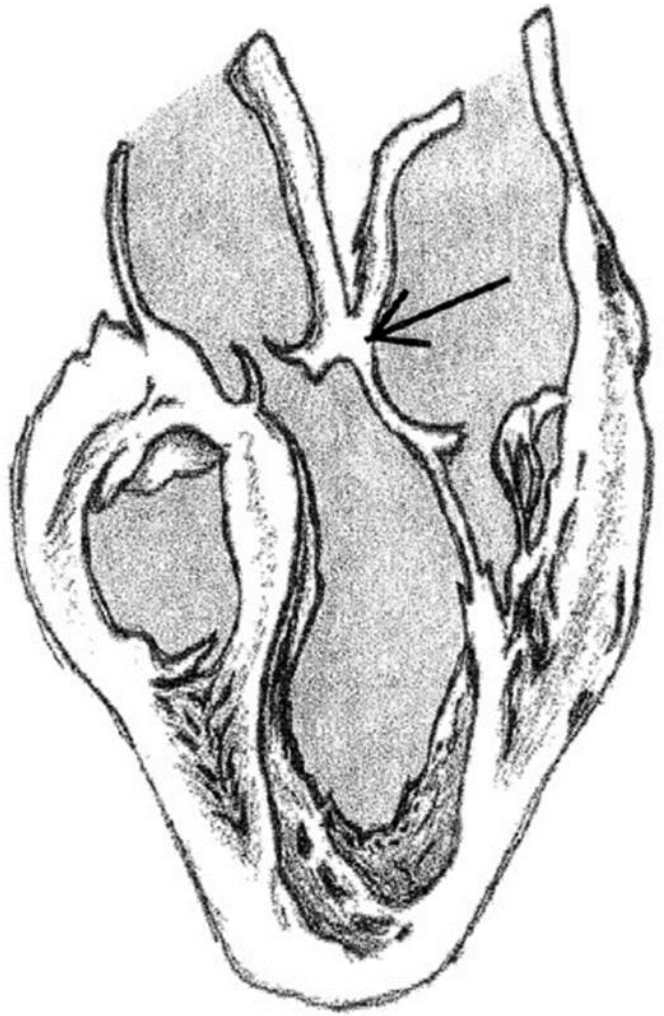

Adult Echo
X8-2t
53Hz
10cm
2D
49%
C 50
P Off
Gen
TIS0.2
MI 0.5
M4
LA
AO
LV
RV
PAT T: 37.0C
TEE T: 39.6C
194 bpm


Adult Echo
X8-2t
18Hz
10cm
2D
51%
C 50
P Off
Gen
CF
48%
4000Hz
WF 359Hz
5.0MHz
TIS0.5
MI 0.6
M4 M4
+30.8
-30.8
cm/s
PAT T: 37.0C
TEE T: 39.6C
180 bpm

P-MAIVF

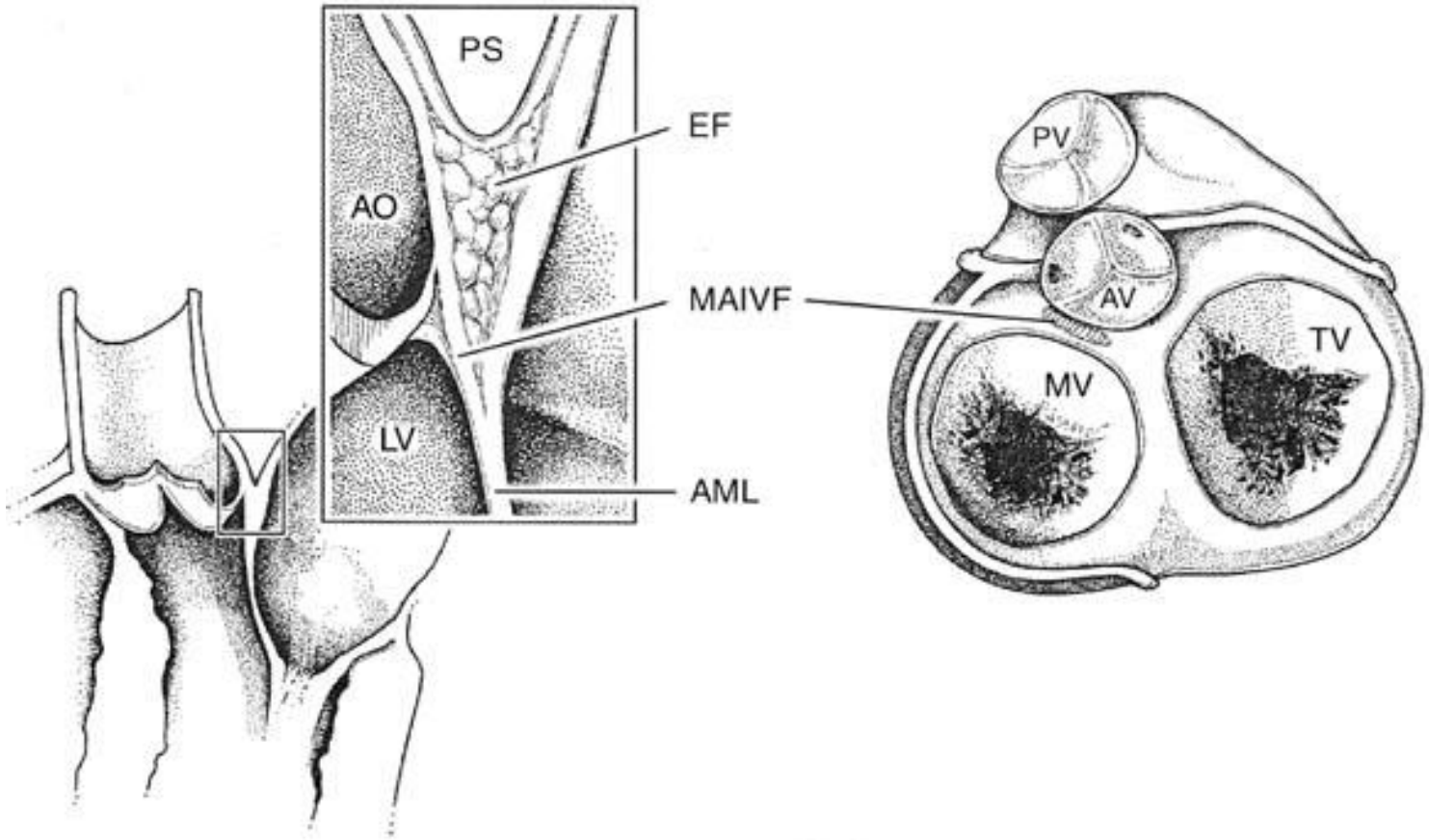
- Inter annular zone between the mitral and aortic valves and its communication with the left ventricular outflow tract between the left coronary or noncoronary aortic cusp and the anterior leaflet of the mitral valve.

- Associations: Infective endocarditis and surgical trauma

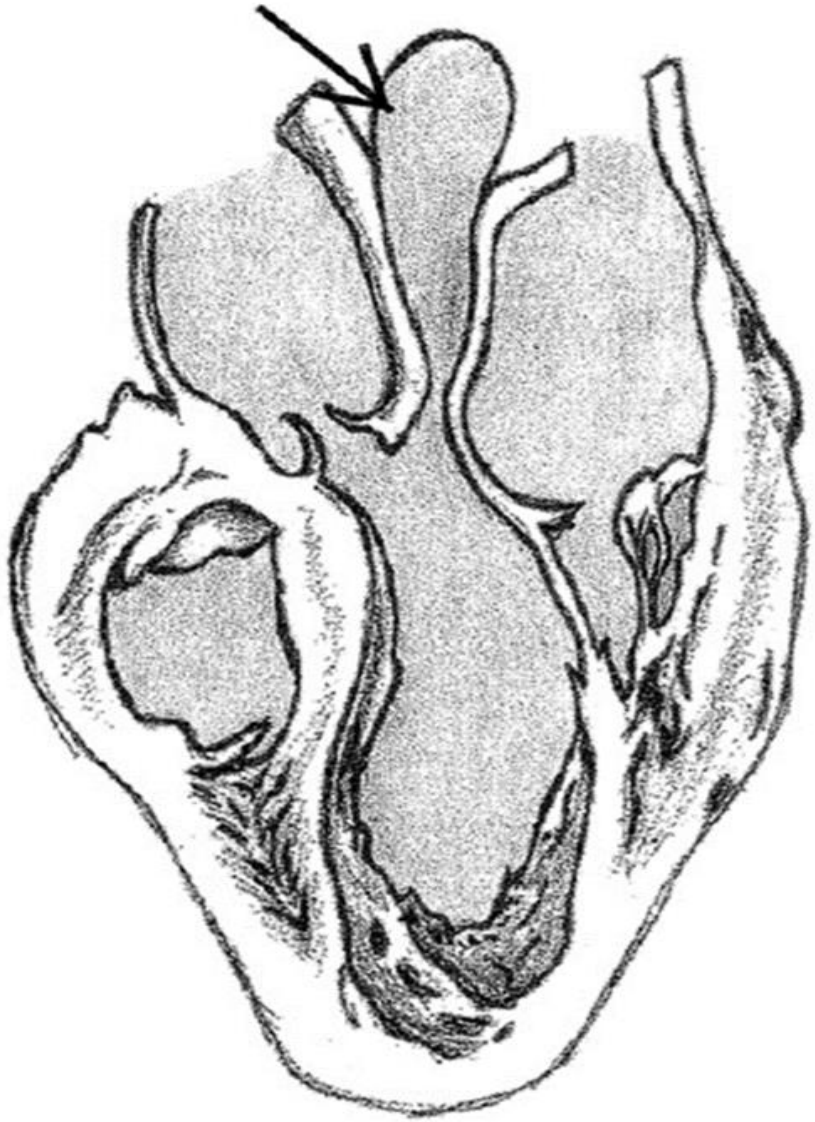


A

Sudhakar et al, Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (MAIVF): A comprehensive review. J Am Soc Echocardiogr. 2010 Oct;23(10):1009-18

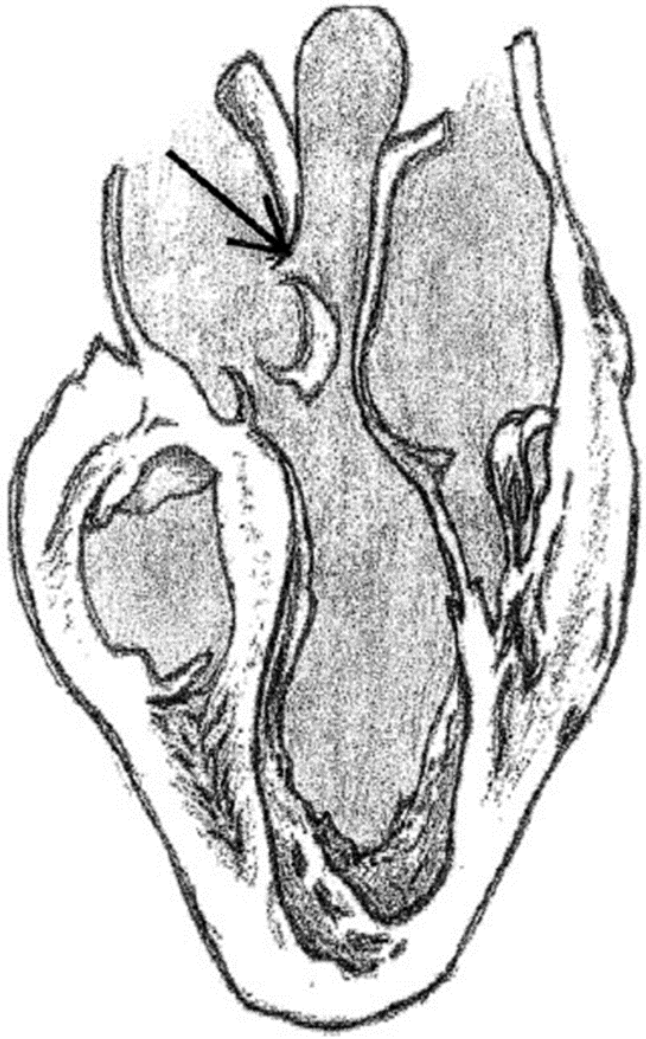


Journal of the American Society of Echocardiography 2002 15, 743-745DOI: (10.1067/mje.2002.118909)



B

Sudhakar et al, Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (MAIVF): A comprehensive review. J Am Soc Echocardiogr. 2010 Oct;23(10):1009-18



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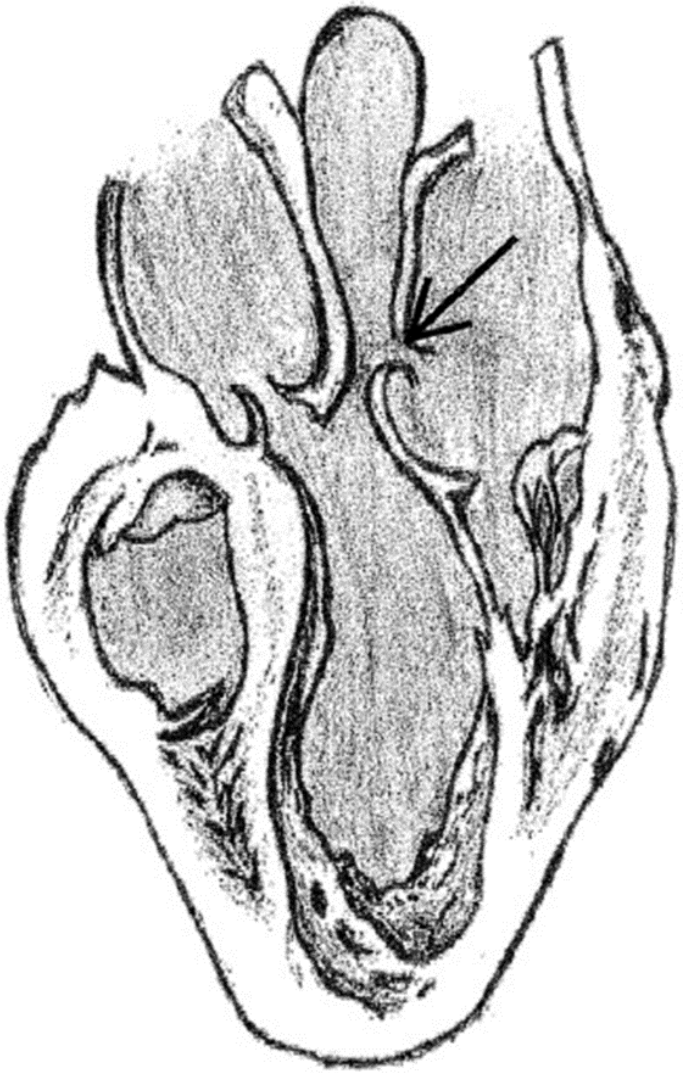


D

Sudhakar et al, Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (MAIVF): A comprehensive review. J Am Soc Echocardiogr. 2010 Oct;23(10):1009-18

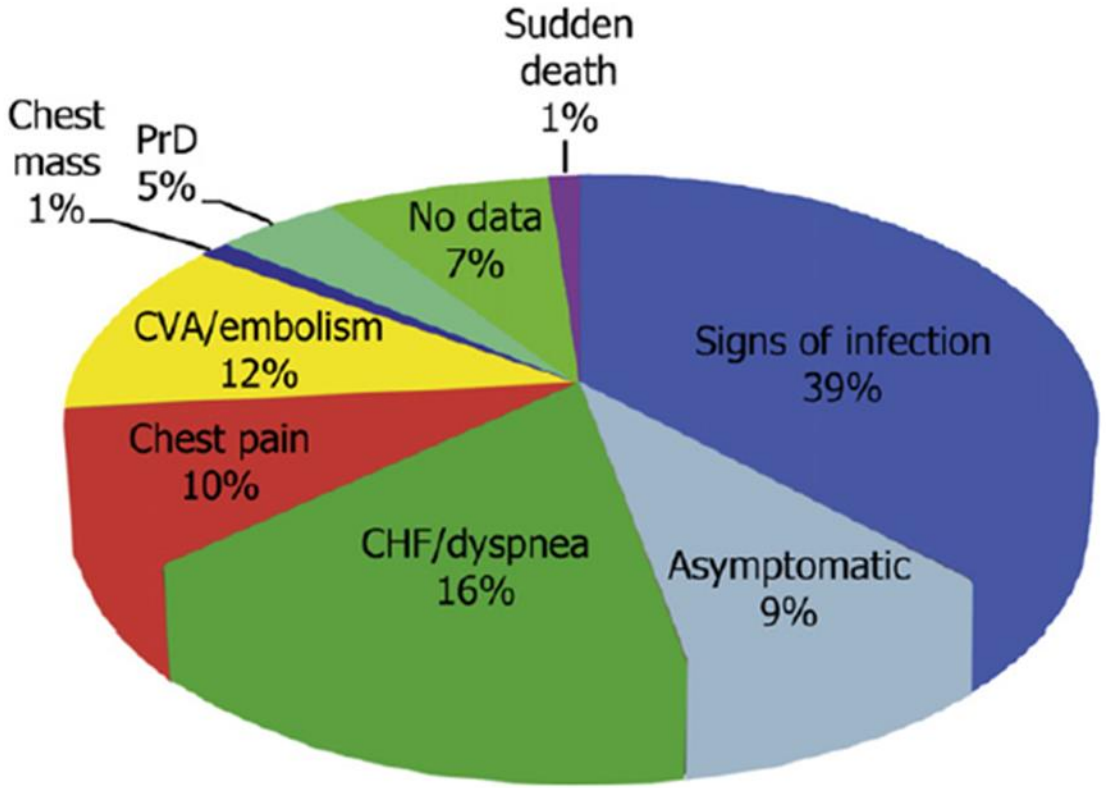


Figure 3 Clinical presentations of reported patients with P-MAIVF. *CHF*, Congestive heart failure; *CVA*, cerebrovascular accident; *PrD*, prosthetic valve dysfunction.

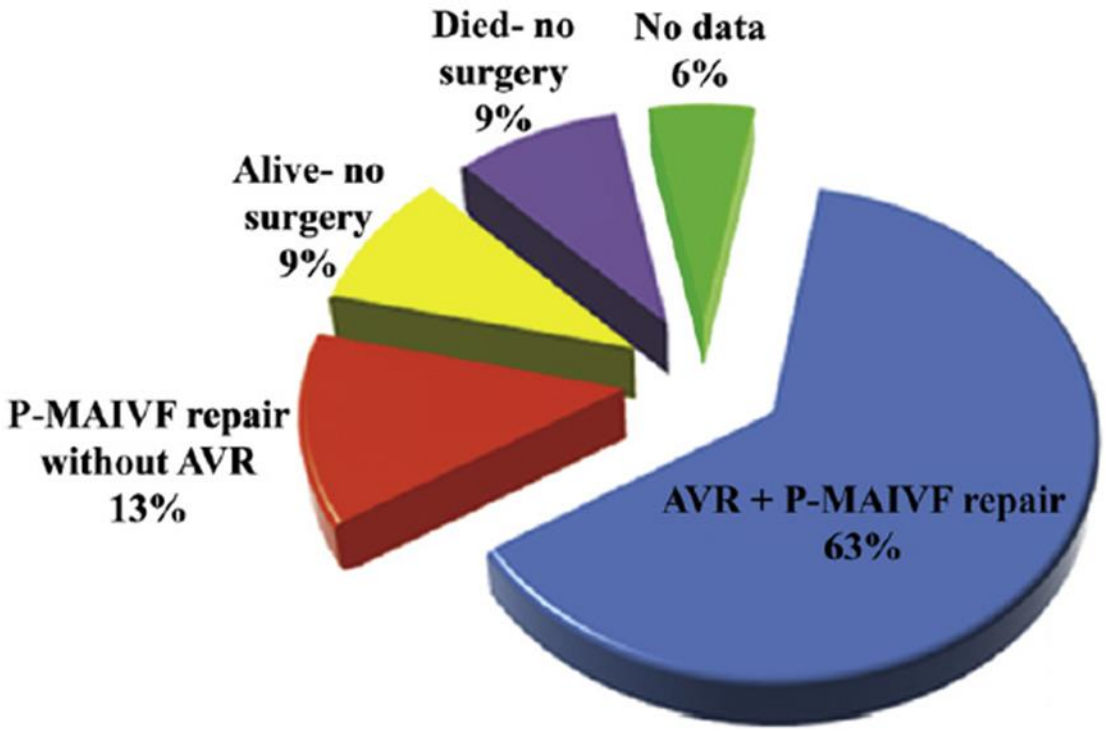


Figure 4 Outcomes of reported patients with P-MAIVF. *AVR*, Aortic valve replacement.

Sudhakar et al, Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (MAIVF): A comprehensive review. J Am Soc Echocardiogr. 2010 Oct;23(10):1009-18

HIGH RISK FEATURES

Table 4 High-risk features for progression of P-MAIVF and development of complications
Active endocarditis
P-MAIVF > 3 cm
Bicuspid aortic valve
Aortic regurgitation
Presence of fistula to cardiac chamber or aorta
Thrombus in P-MAIVF
Compression of adjacent structures (coronary or pulmonary artery)

Sudhakar et al, Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (MAIVF): A comprehensive review. J Am Soc Echocardiogr. 2010 Oct;23(10):1009-18

COMPLICATIONS

Table 3 Complications of P-MAIVF
Compression
Coronary artery: angina, thrombosis (myocardial infarction), dissection, aneurysm
Pulmonary artery: pulmonary hypertension
Mitral valve: mitral regurgitation
Fistula formation
Aorta
Left atrium
Rupture
Pericardial tamponade, death
Thrombosis
Transient ischemic attack, cerebrovascular accident
Infection
Endocarditis, fistula formation
Heart failure

Sudhakar et al, Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (MAIVF): A comprehensive review. J Am Soc Echocardiogr. 2010 Oct;23(10):1009-18

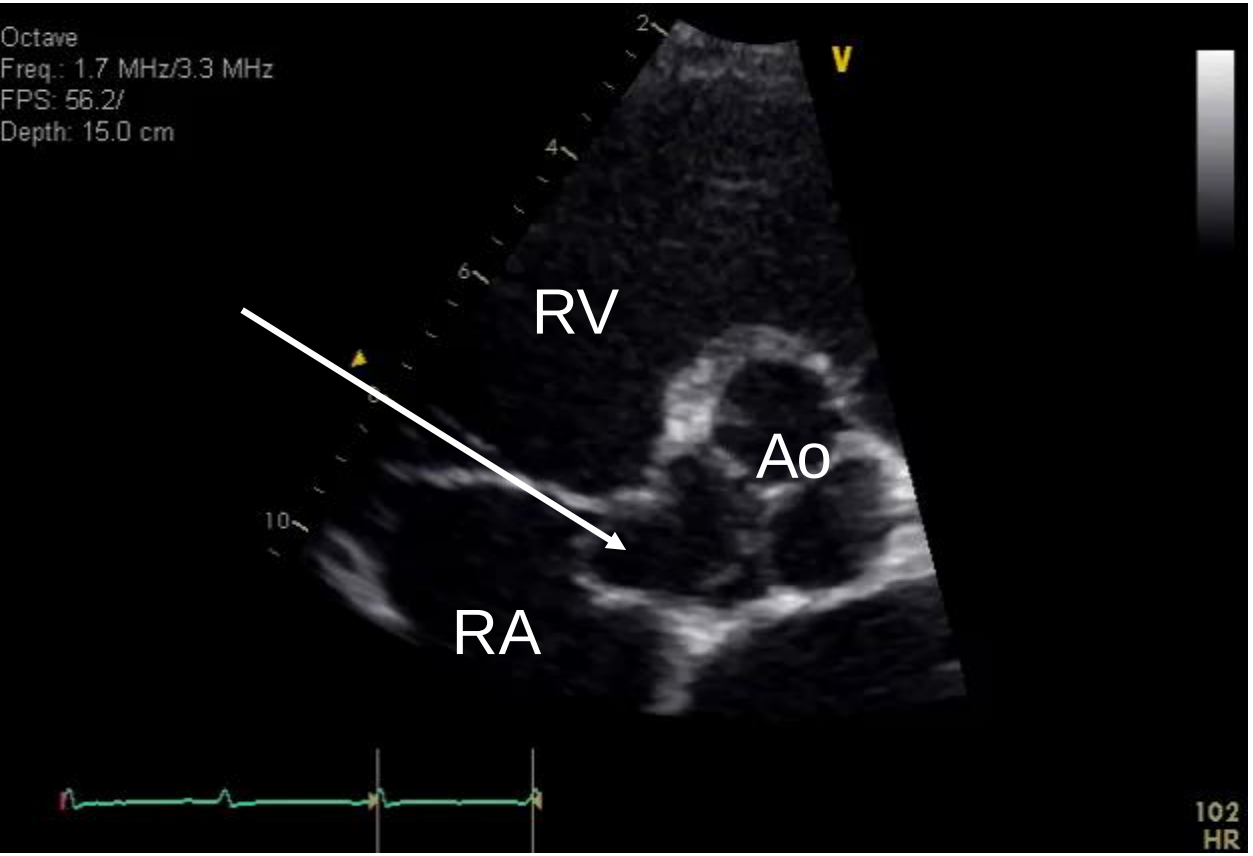
ECHOCARDIOGRAPHIC FEATURES

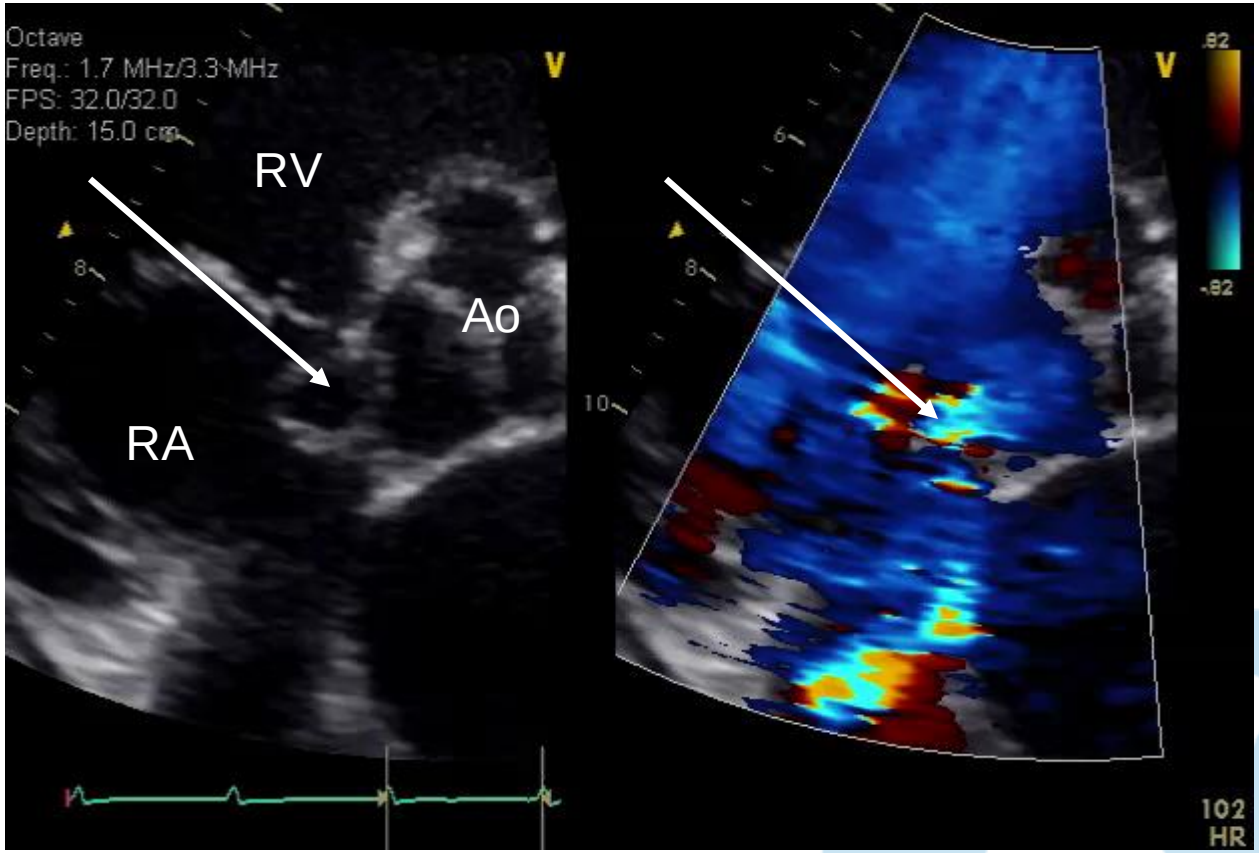
- Visualization of the echo-free space with systolic expansion and diastolic collapse of the pseudoaneurysm

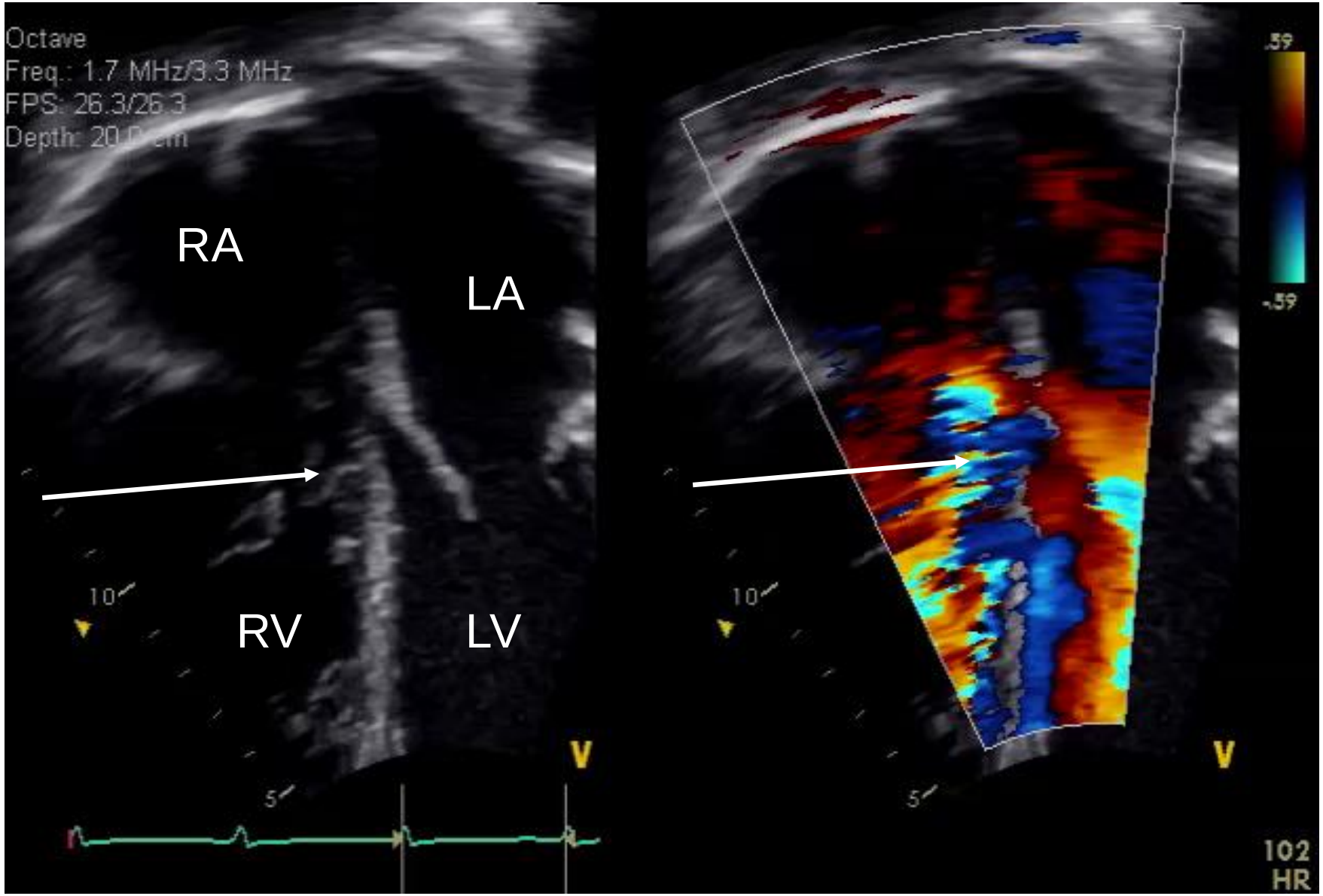
- During systole: the high LV pressure increases the blood flow into the pseudoaneurysm, and during diastole, the blood flows back into the LVOT

Sudhakar et al, Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (MAIVF): A comprehensive review. J Am Soc Echocardiogr. 2010 Oct;23(10):1009-18

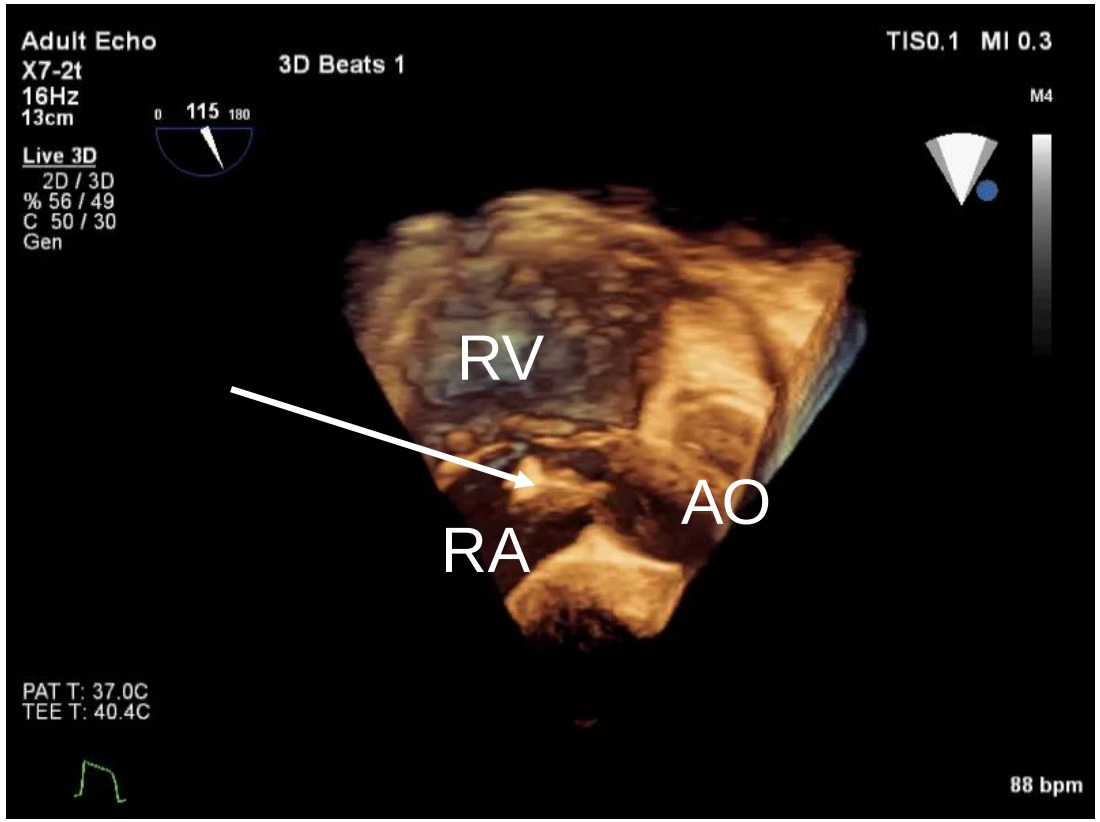
OTHER CONSIDERATIONS - SOVA

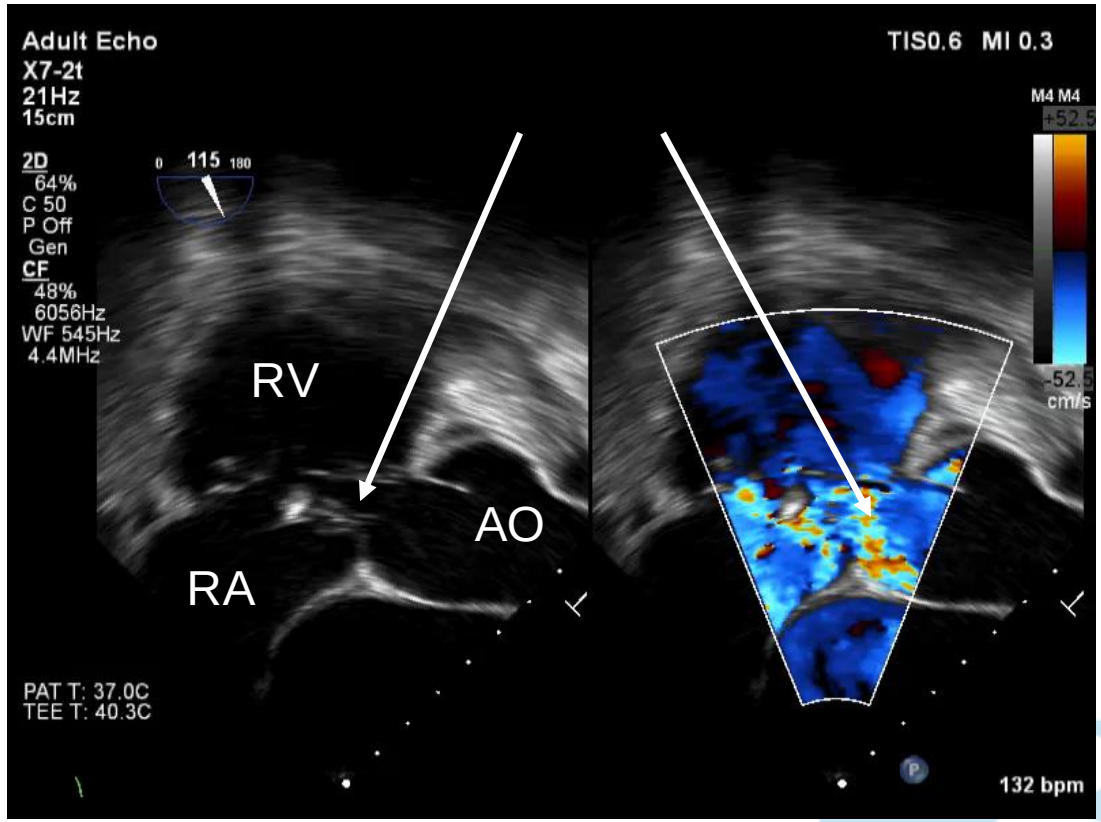





Octave
Freq.: 1.7 MHz/3.3 MHz
FPS: 26.3/26.3
RA
LA
RV
LV
.59
-.59
102
HR

Ruptured SOVA





- A SOV… …eakness affectin…
- Echo: s… …OVA that expand…
- An ane… …acent cavity

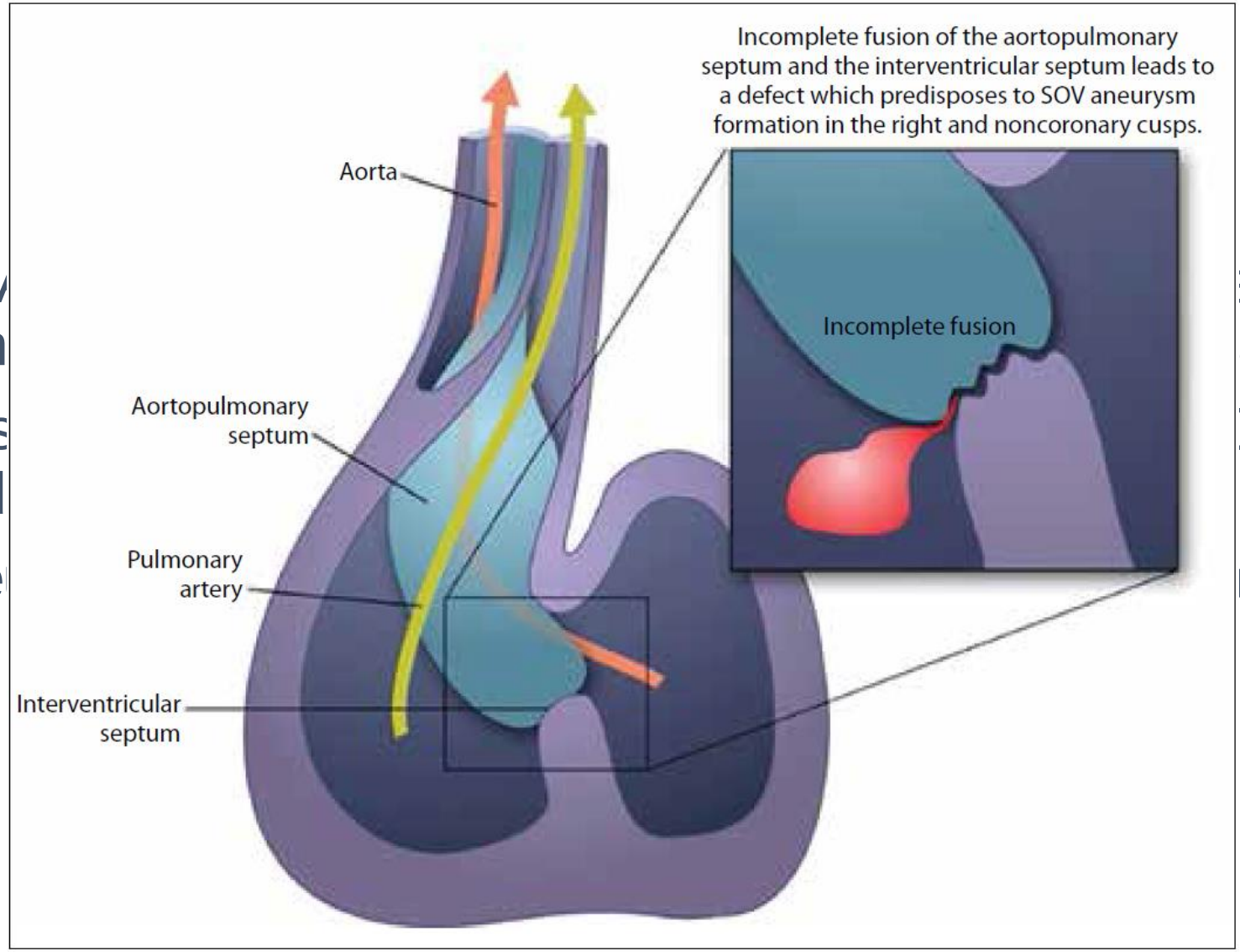


Figure 2. Illustration of the heart and root of aorta showing the pathogenesis for congenital SOV aneurysm.

Mitral-Aortic Intervalvul… …G, Tsakalis K, Alexopoulos N, Barbetseas J.J Cardiovasc Ultrasound. 2015 Dec;23(4):257-61

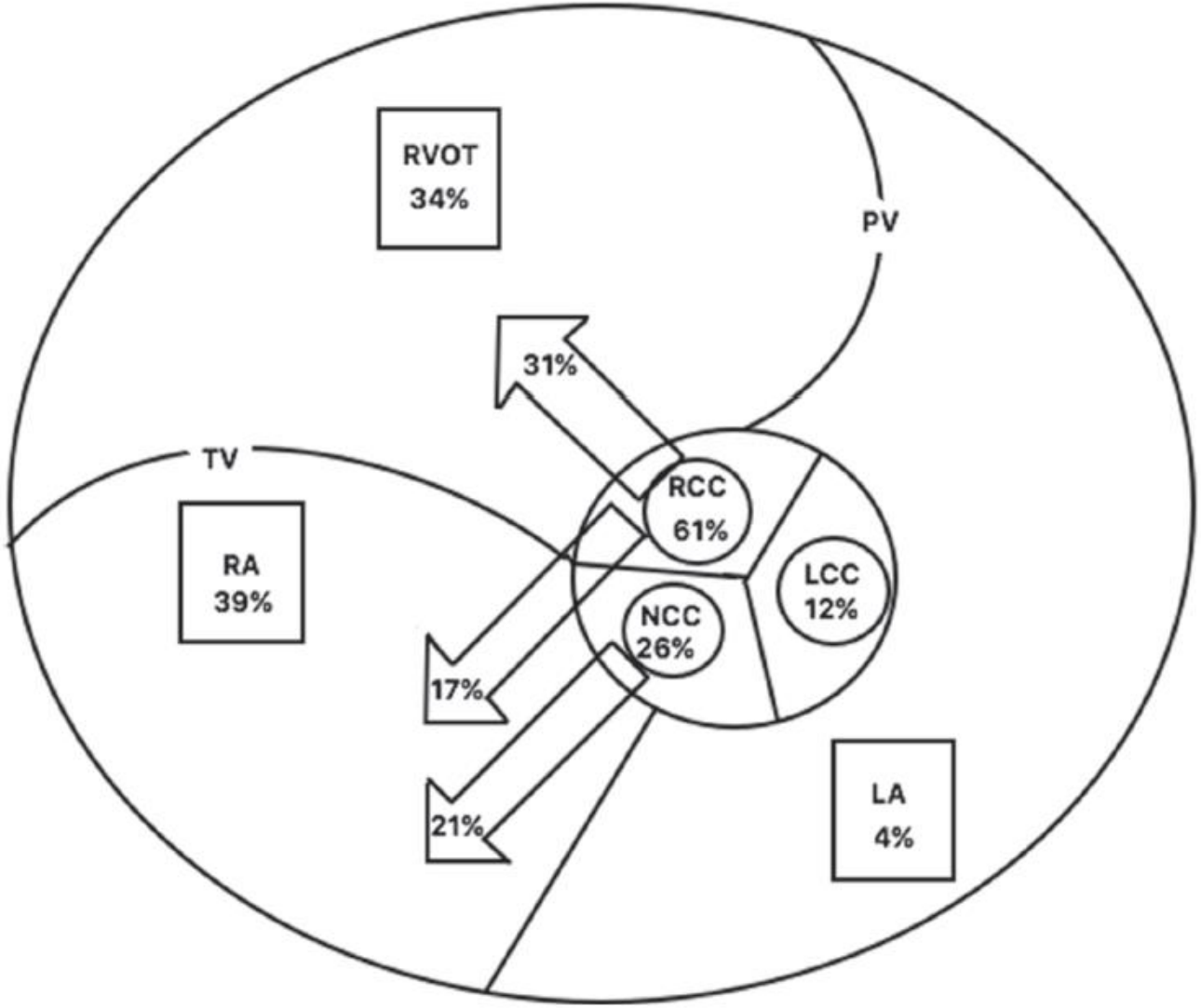
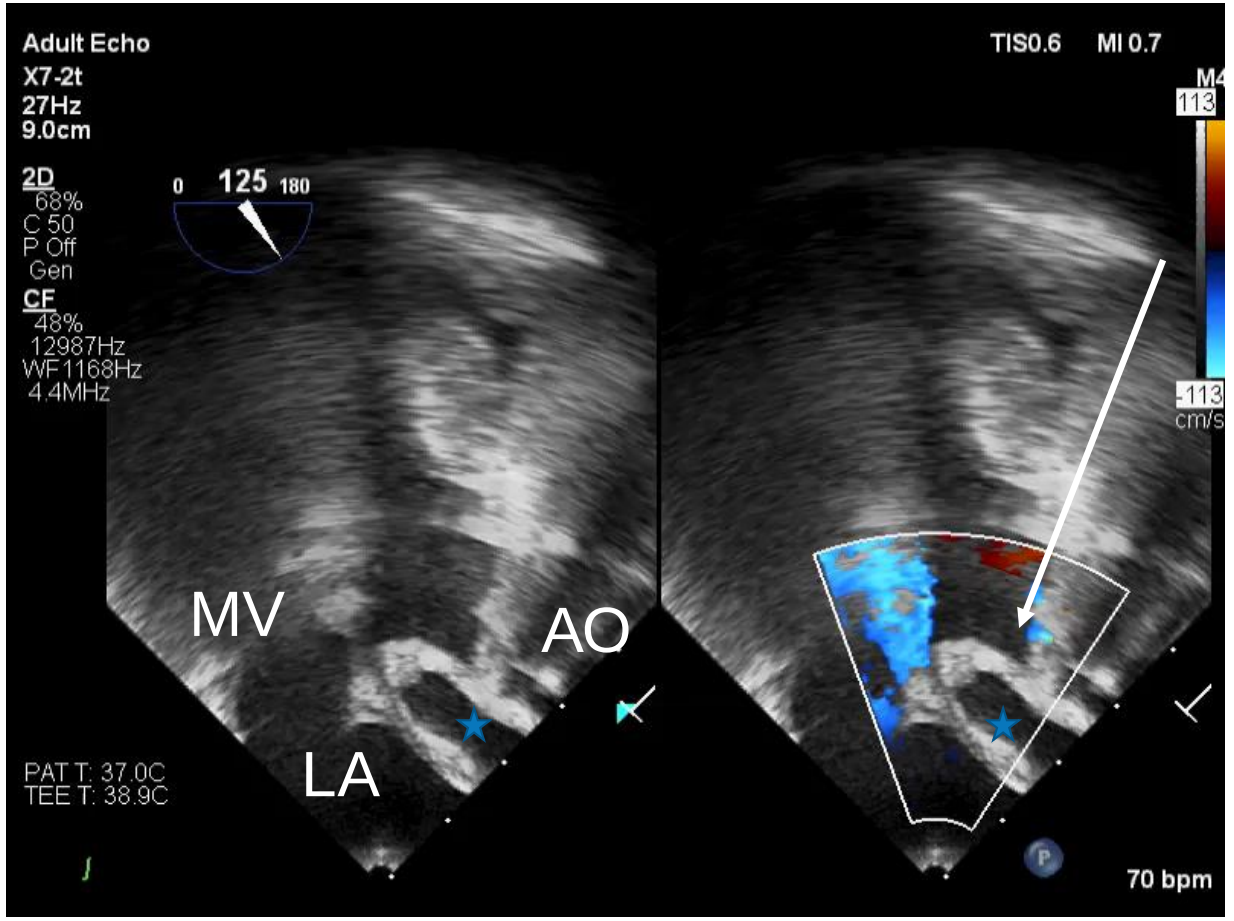
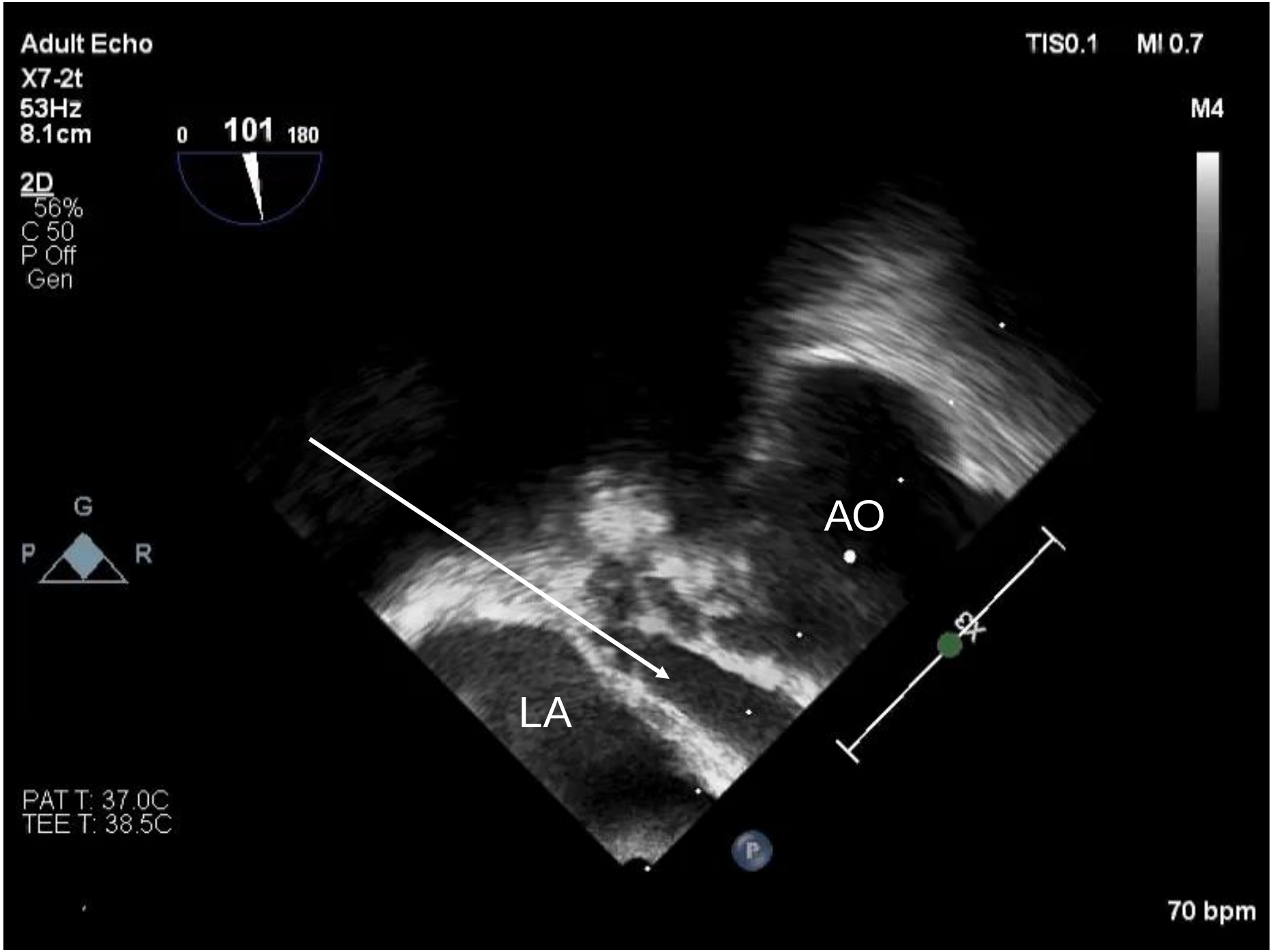


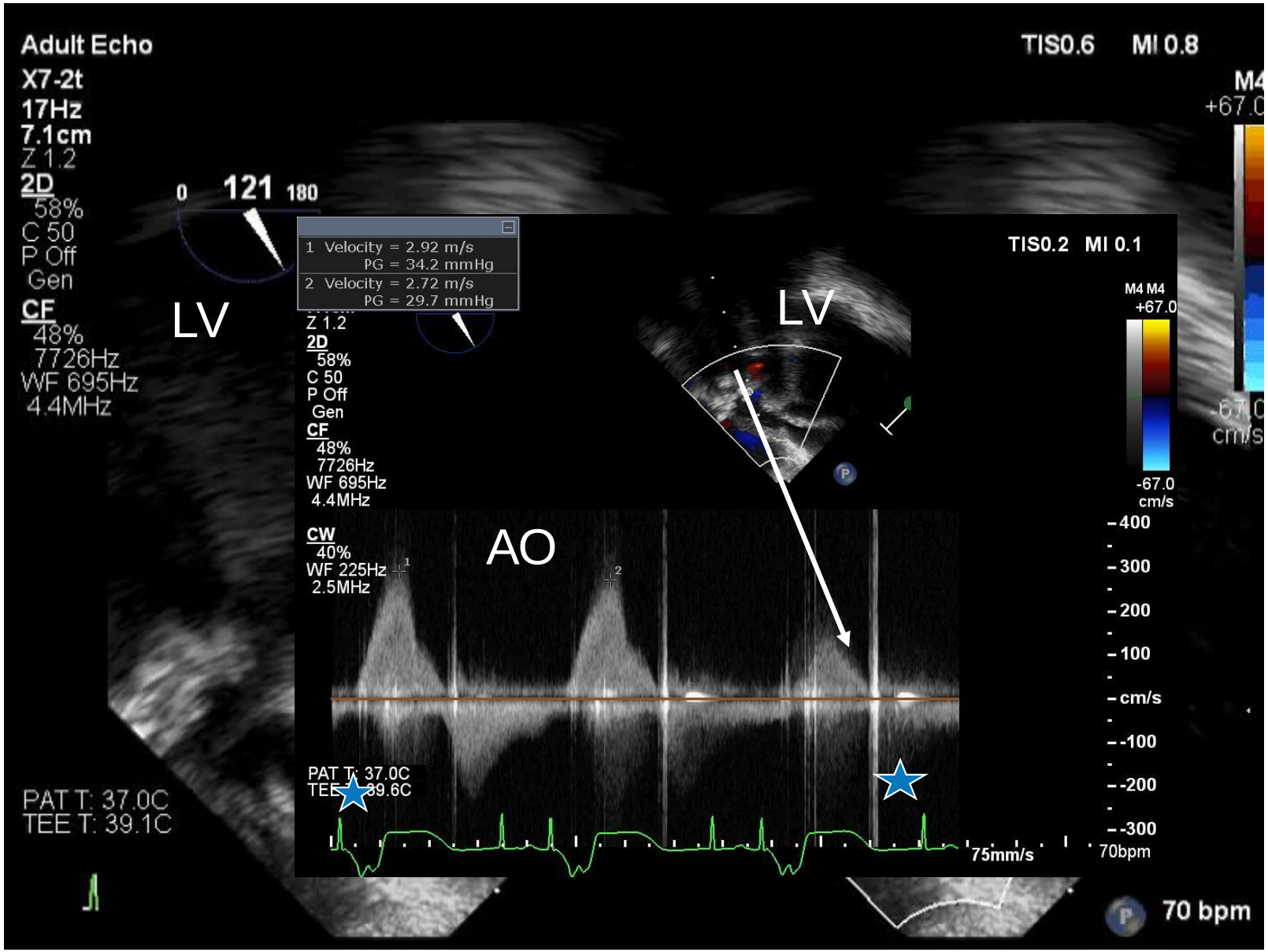
Fig. 1. Anatomical diagram of most common origins (○), pathways (⇑) and endpoints (□) of RSoVA. RCC: right coronary cusp, NCC: non-coronary cusp, LCC: left coronary cusp, RVOT: right ventricular outlet tract, RA: right atrium, LA: left atrium, PV: pulmonic valve, TV: tricuspid valve. For further information please refer to Tables 2 and 3.

T.S. Marroush et al. / Heart & Lung 47 (2018) 131–135

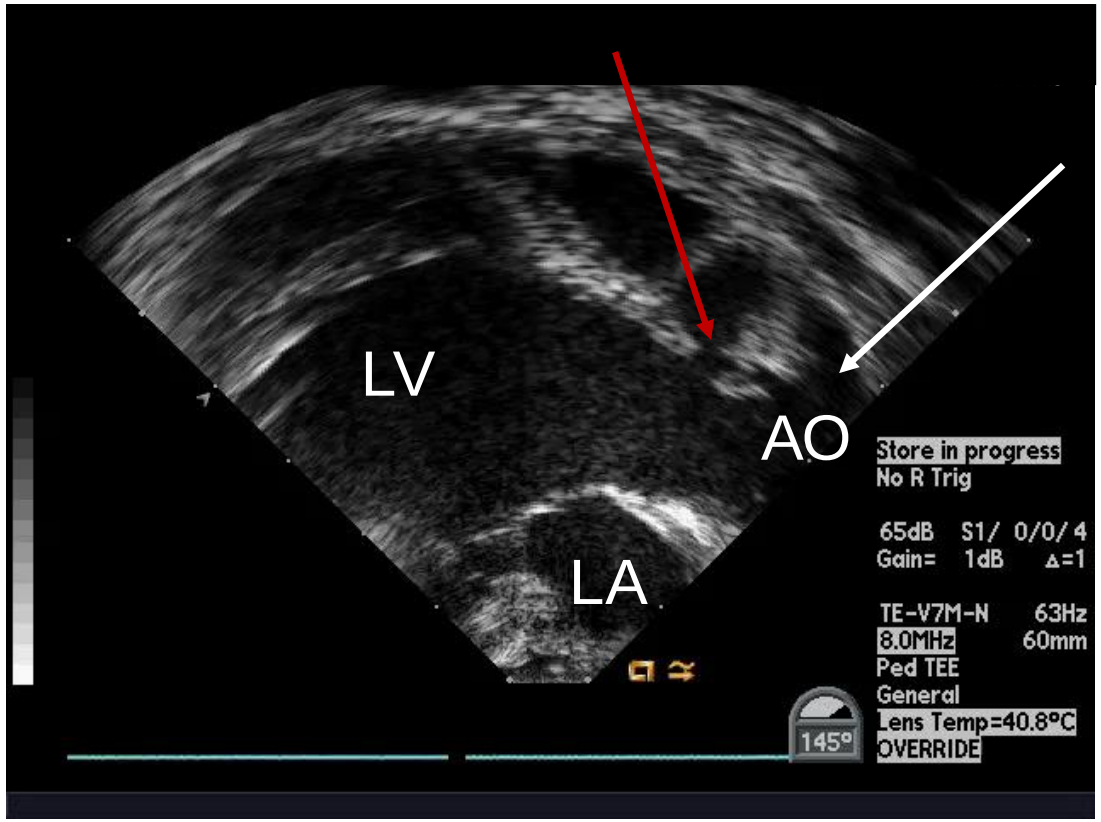
Surgically created patch

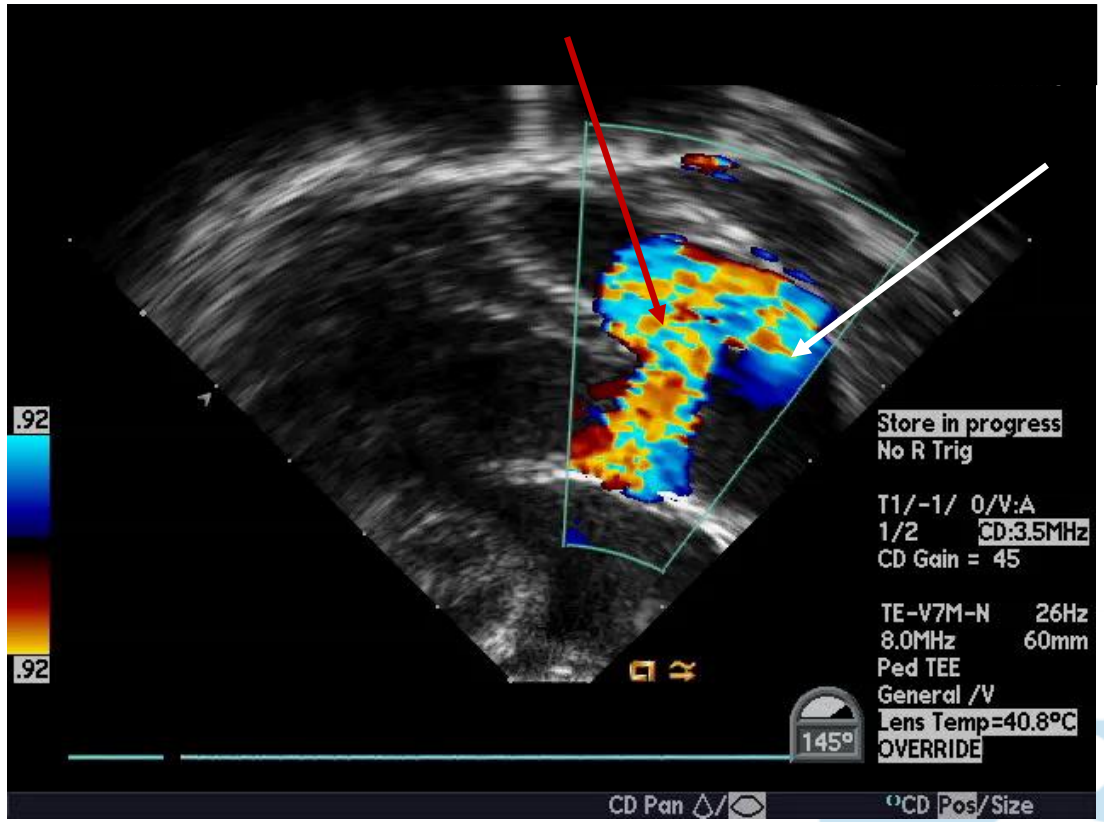



Adult Echo
X7-2t
53Hz
8.1cm
0
101
180
2D
56%
C 50
P Off
Gen
TIS0.1
MI 0.7
M4
G
P
R
AO
LA
PAT T: 37.0C
TEE T: 38.5C
70 bpm

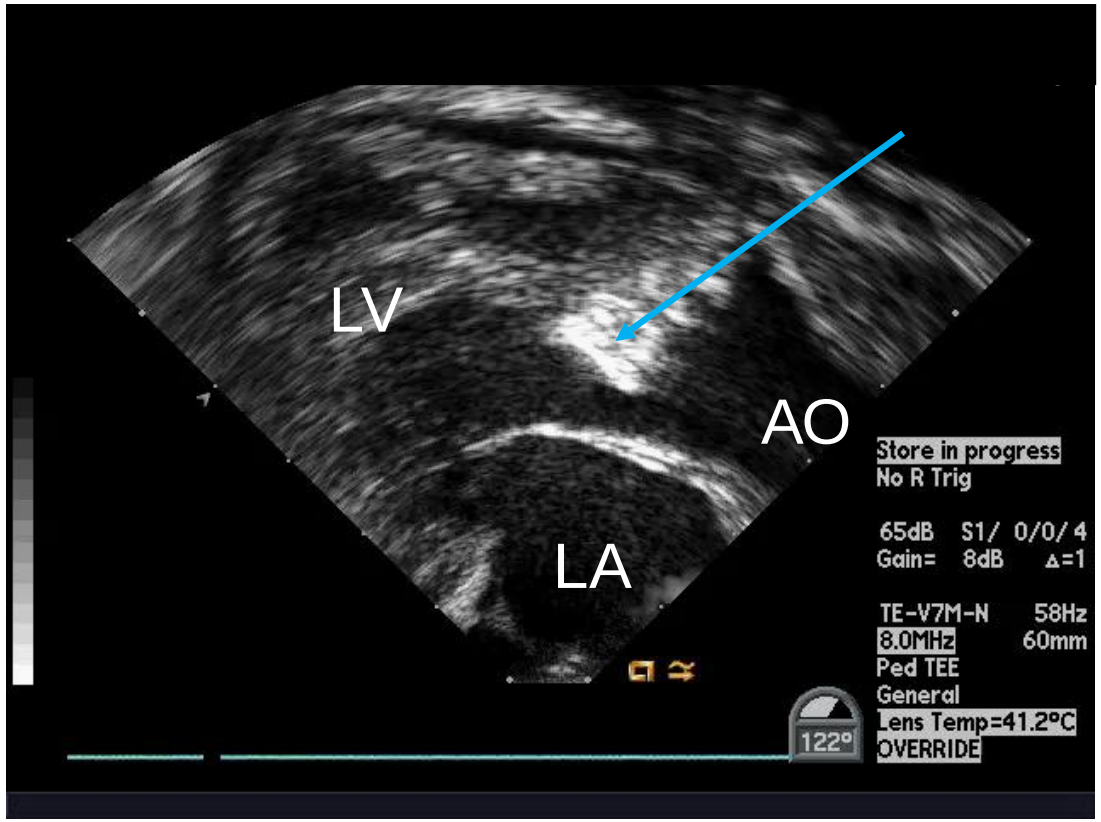

Adult Echo
X7-2t
17Hz
7.1cm
Z 1.2
2D
58%
C 50
P Off
Gen
CF
48%
7726Hz
WF 695Hz
4.4MHz
TIS0.6 MI 0.8
0 121 180
LV
1 Velocity = 2.92 m/s
PG = 34.2 mmHg
2 Velocity = 2.72 m/s
PG = 29.7 mmHg
TIS0.2 MI 0.1
LV
M4 M4
+67.0
-67.0
cm/s
CW
40%
WF 225Hz
2.5MHz
AO
400
300
200
100
cm/s
-100
-200
-300
75mm/s
70bpm
PAT T: 37.0C
TEE T: 39.1C
70 bpm

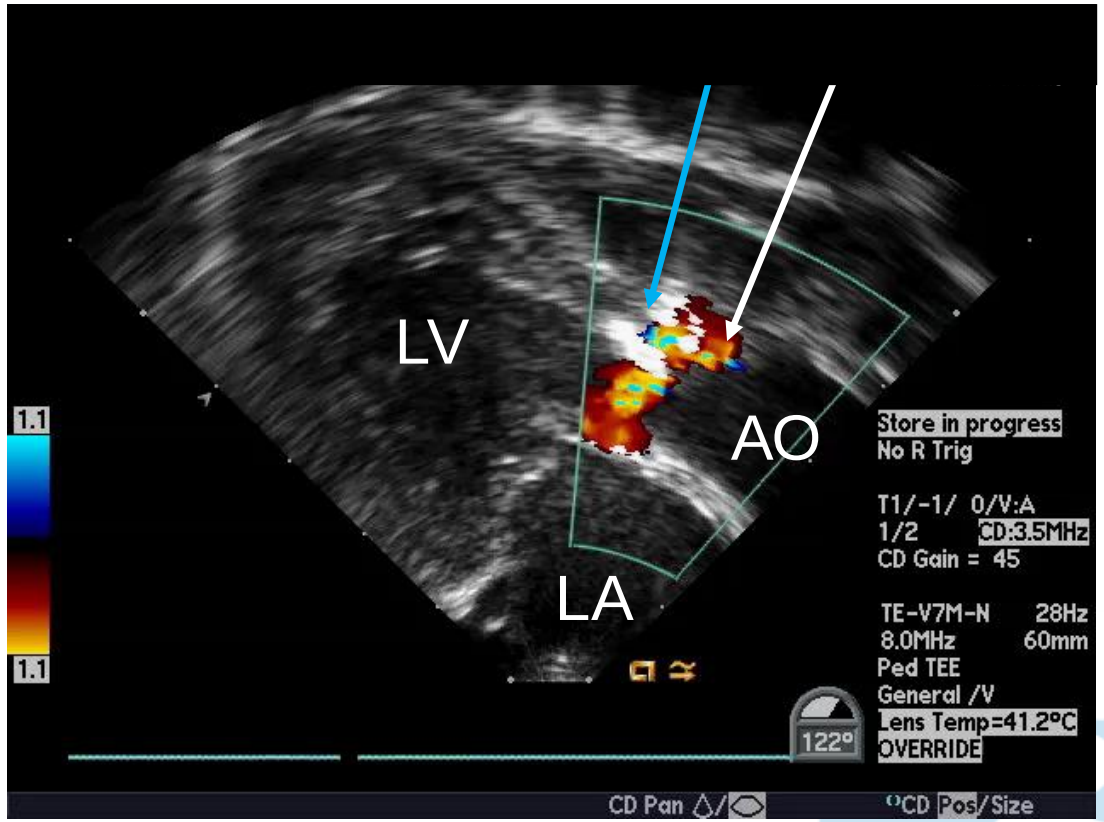
Aortic to LV tunnel



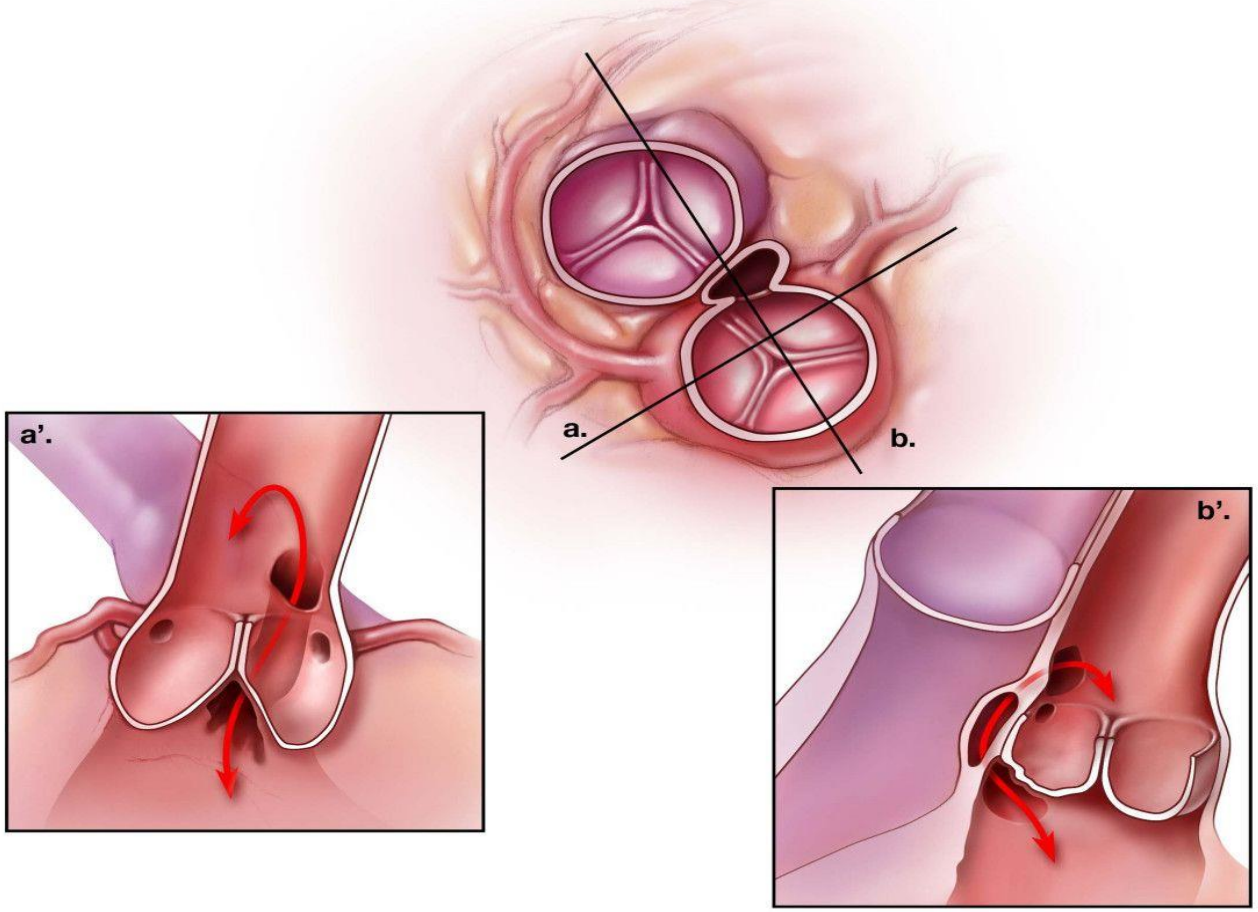


Device closure of aortic to LV tunnel

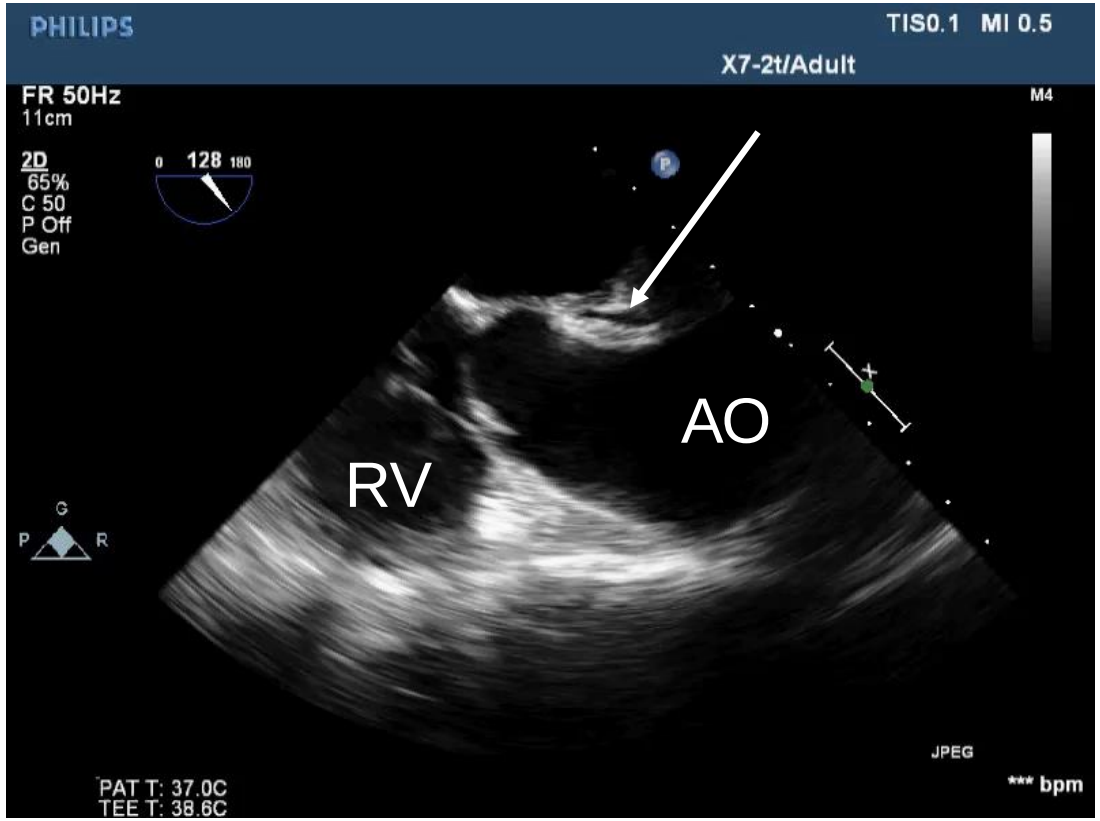


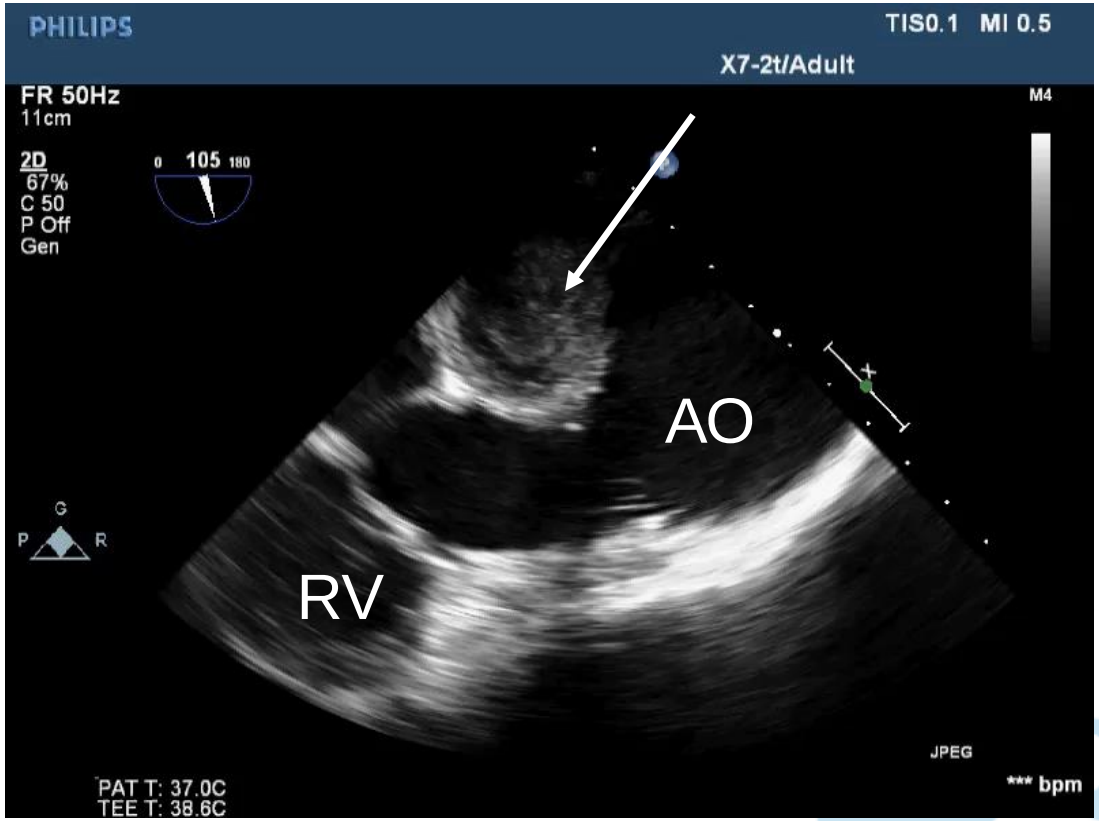


Aortic to LV tunnel

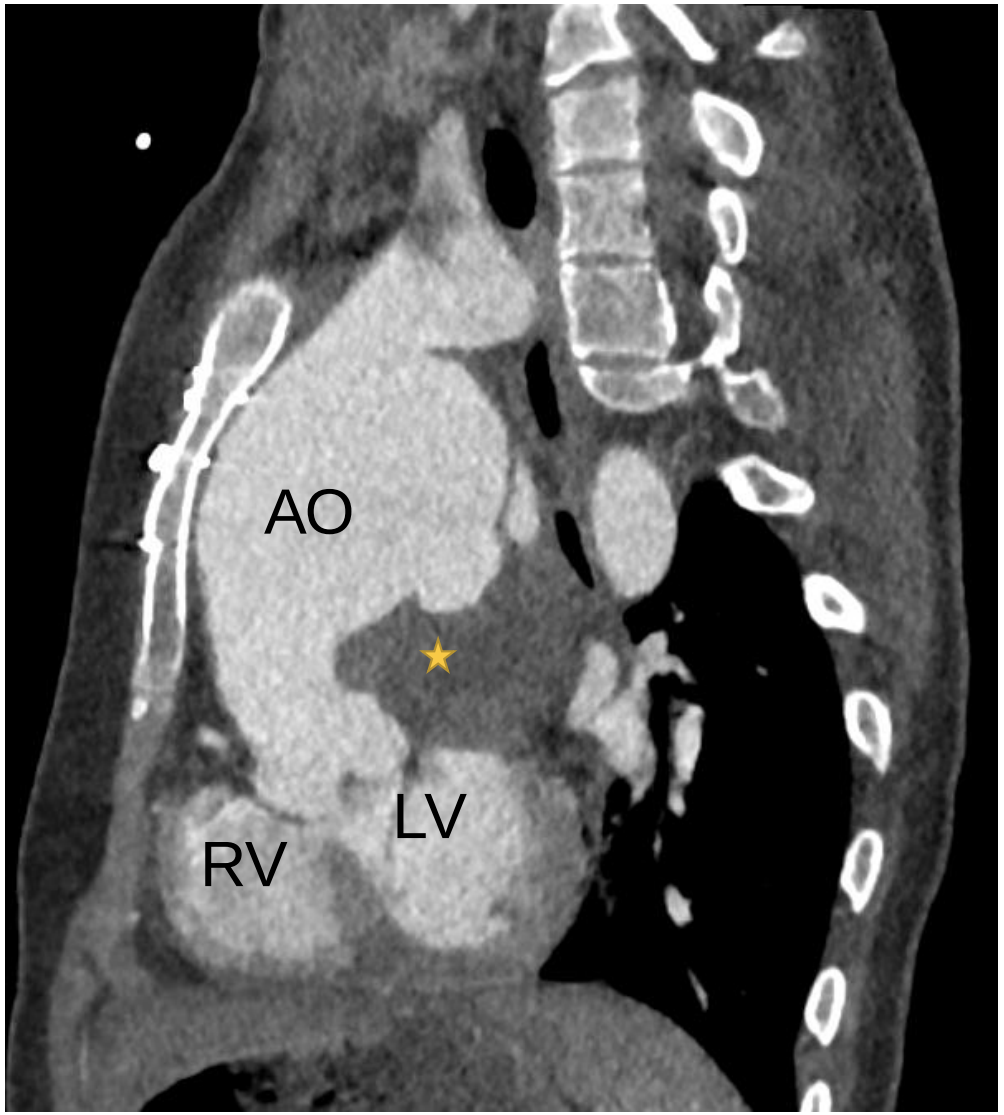


McKay R. Orphanet J Rare Dis.2007 Oct 8;2:41.


PHILIPS
TIS0.1 MI 0.5
X7-2t/Adult
FR 50Hz
11cm
M4
2D
65%
C 50
P Off
Gen
0 128 180
AO
RV
JPEG
PAT T: 37.0C
TEE T: 38.6C
*** bpm


PHILIPS
TIS0.1 MI 0.5
X7-2t/Adult
FR 50Hz
11cm
M4
2D
67%
C 50
P Off
Gen
0 105 180
AO
RV
JPEG
PAT T: 37.0C
TEE T: 38.6C
*** bpm

CTA



Contained aortic rupture

- A contained aortic root rupture–usually associated with regional manipulations and peri annular hematoma–may be extremely difficult to discriminate from MAIVF-P by echocardiography

Mitral-Aortic Intervalvular Fibrosa Pseudoaneurysm.Bonou M, Papadimitraki ED, Vaina S, Kelepeshis G, Tsakalis K, Alexopoulos N, Barbetseas J.J Cardiovasc Ultrasound. 2015 Dec;23(4):257-61

Pearls

- A space between the aortic valve and MV is abnormal and should be interrogated further
- Patients with BAV are prone to endocarditis
- New onset murmur should prompt concern for a fistulous communication in a previously healthy patient
- TTE and TEE are initial useful modalities; CT and MRI provide excellent delineation

Come, seek, for search is the foundation of fortune: every success depends upon focusing the heart.

Rumi

Cleveland Clinic Children's