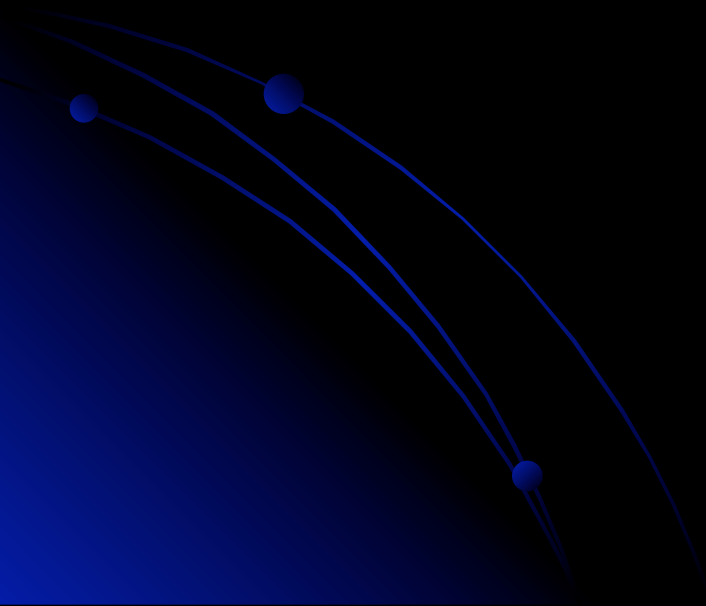


Case 1

**A 23-year-old man with a
heart murmur**

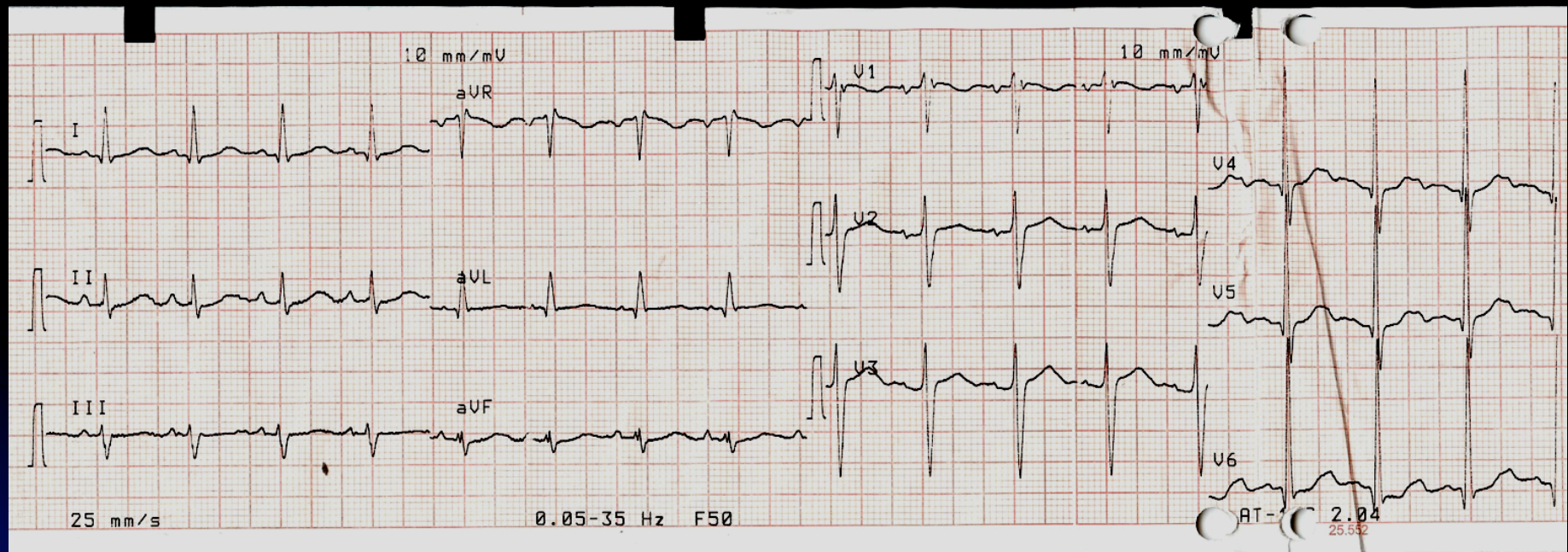


- A 23-year-old labourer of south-asian ethnic background, experienced a sharp central chest pain, which commenced 20 minutes after climbing 2 floors of stairs, while carrying a 50 kg heavy cement bag.
- The chest pain subsided within 30 minutes, but was followed by progressive and intense shortness of breath and dizziness, for which he presented.
- No fever and no recent history of infection.
- No past history of any serious illness.

Physical:

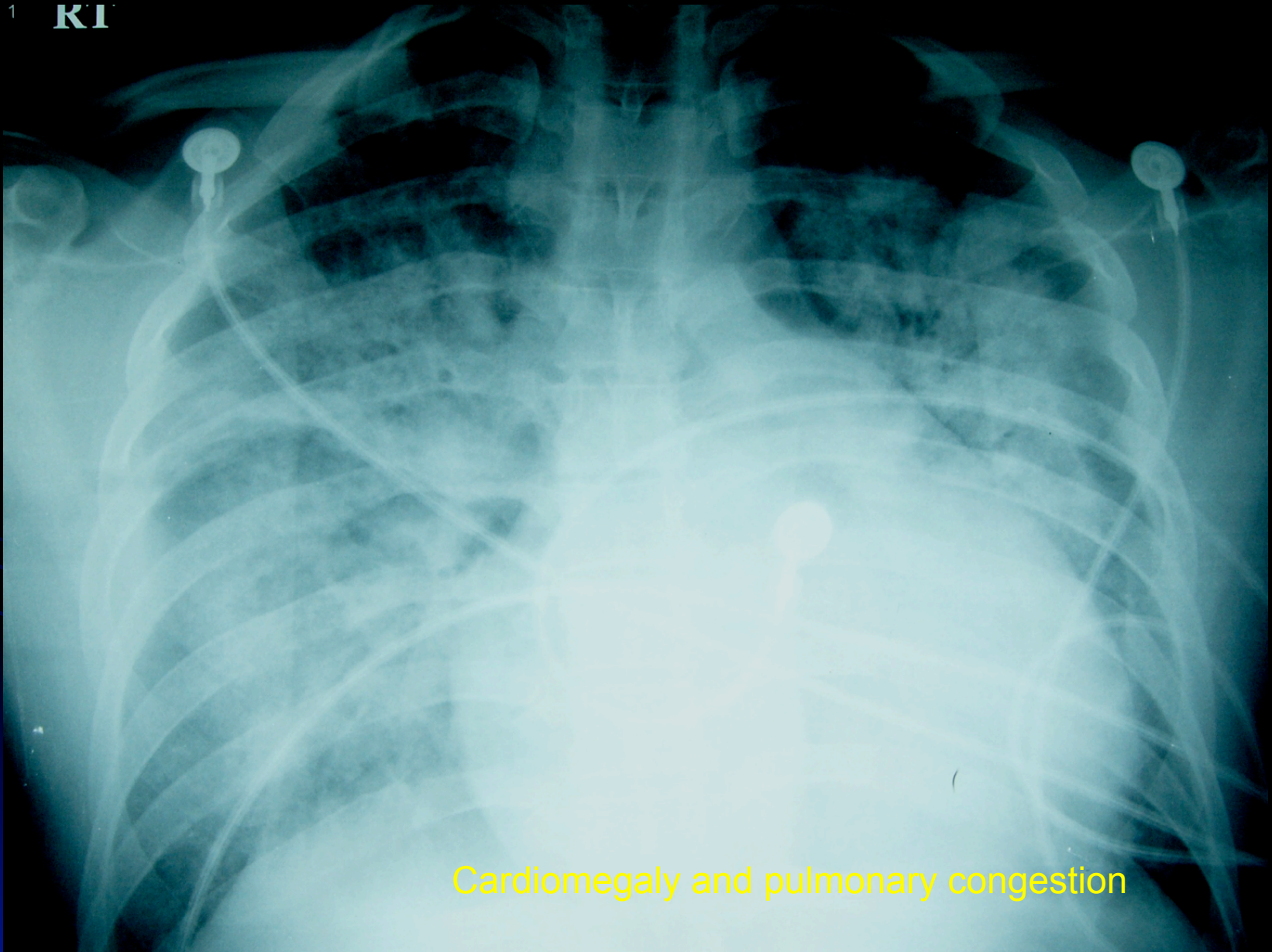
- **SOB. Tachycardic 100 BPM. BP 110/50 mmHg. Normal temperature. Transdermal O2 sat. 91%.**
- **Marked pulsation of jugular veins.**
- **A palpable thrill the sternum.**
- **4/6 loud systolic-diastolic „machinery“ murmur over left sternal border. No S3 or S4.**
- **Chest clear.**
- **Peripheral pulses were bounding.**
- **Abdomen normal.**

ECG: Sinus Rythm, tachycardia 101 BPM, incomplete RBBB.




Chest X-Ray:

1 KI



Cardiomegaly and pulmonary congestion

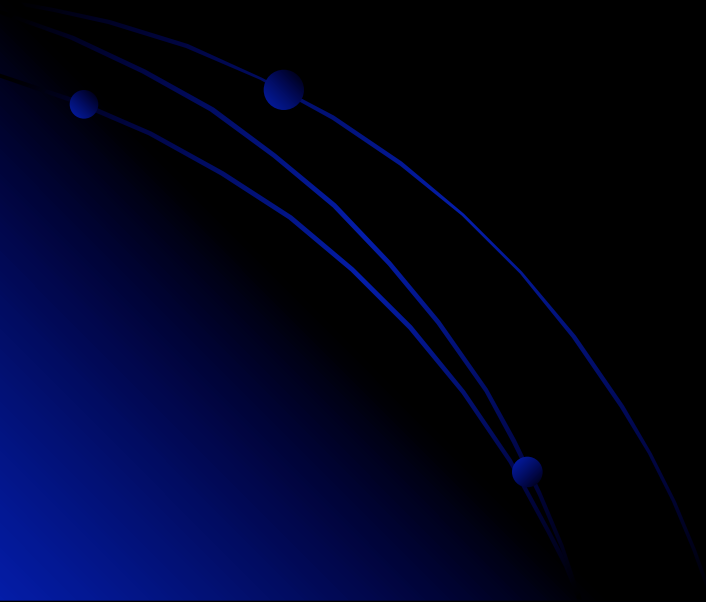
Lab investigations:

- Electrolytes, Creat., liver enzymes, CRP, ESR, PTT, CPK: All normal.
 - CBC: Leucocytosis 18.600 with normal differential WBC. Hg 14 g/dl
 - Troponin: \uparrow 0.3 (N <0.01)
- 

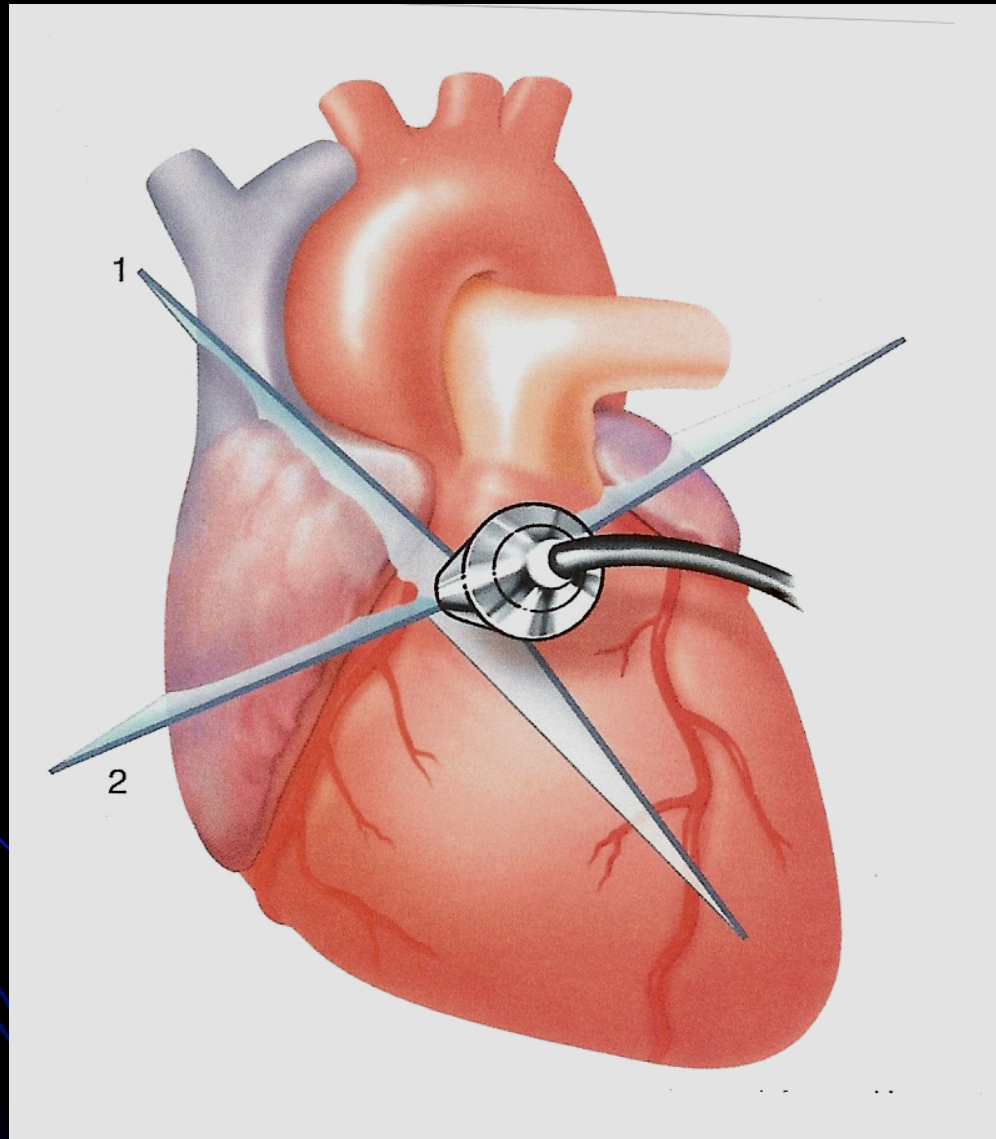
**The patient is in congestive heart failure
but what is the diagnosis?**

- 1. Atrial septal defect (ASD)?**
- 2. Ventricular septal defect (VSD)?**
- 3. Coarctation of the aorta?**
- 4. Aortic regurgitation?**
- 5. Ruptured sinus valsalva aneurysm?
coronary syndrome?**
- 6. Patent ductus arteriosus (PDA)?**
- 7. Acute coronary syndrome?**

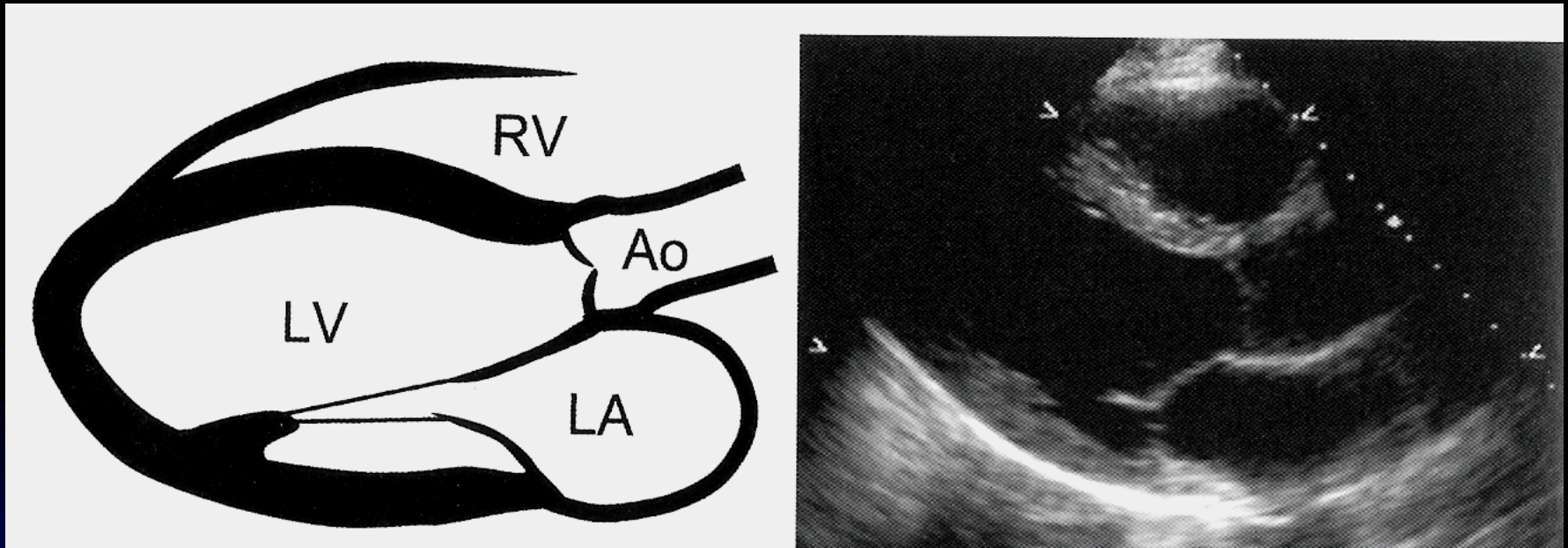
Echocardiogram



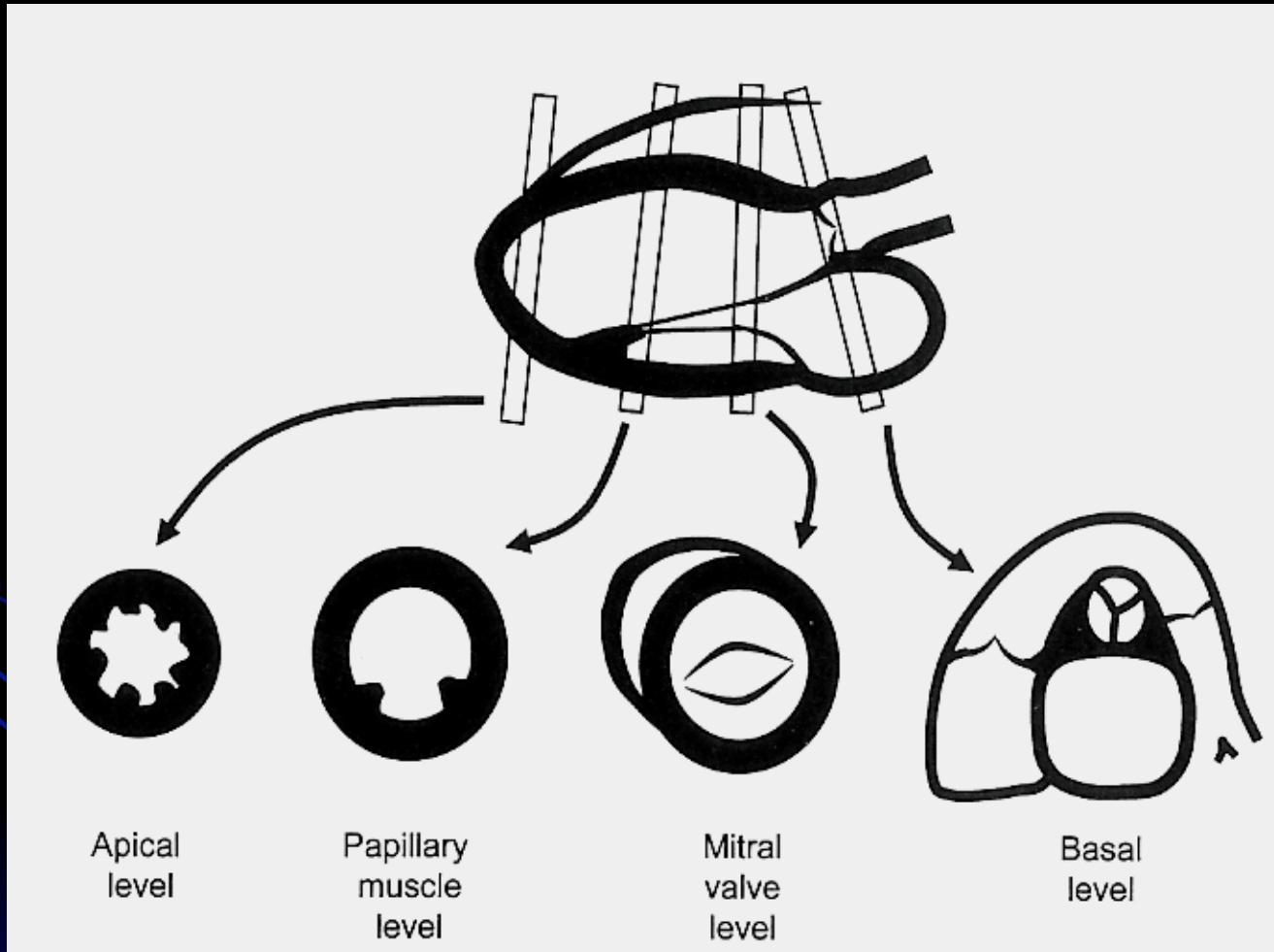
Echocardiographic views:



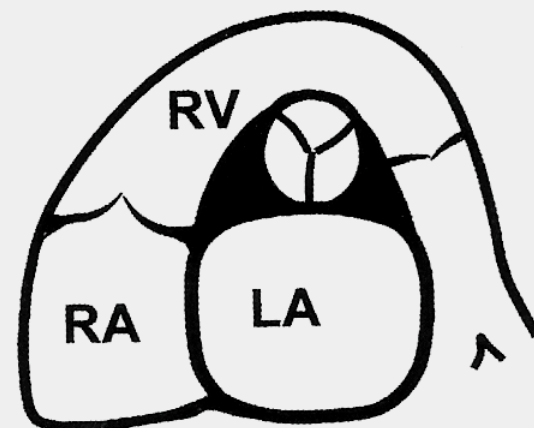
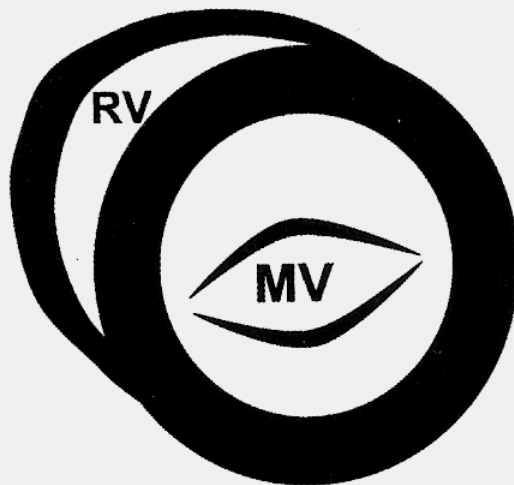
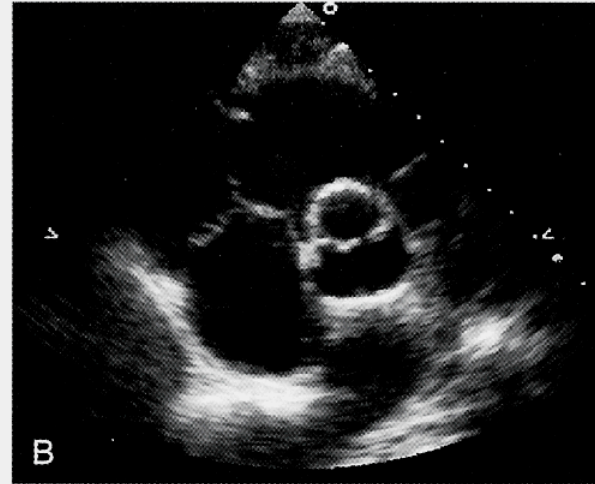
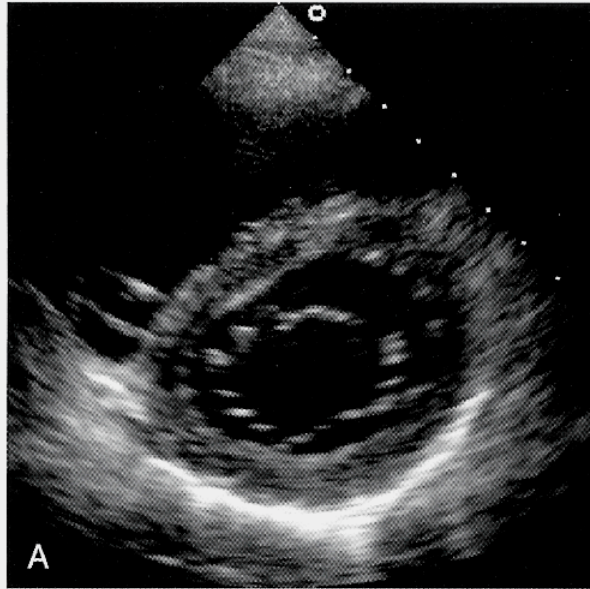
Echocardiogram: Long axis view



Echocardiogram: Short axis view planes



Echocardiogram: Short axis view planes



2D 13 cm
74 f/s
f: 1.7 MHz H
DR: 65 dB
R: 2.0 G: 51

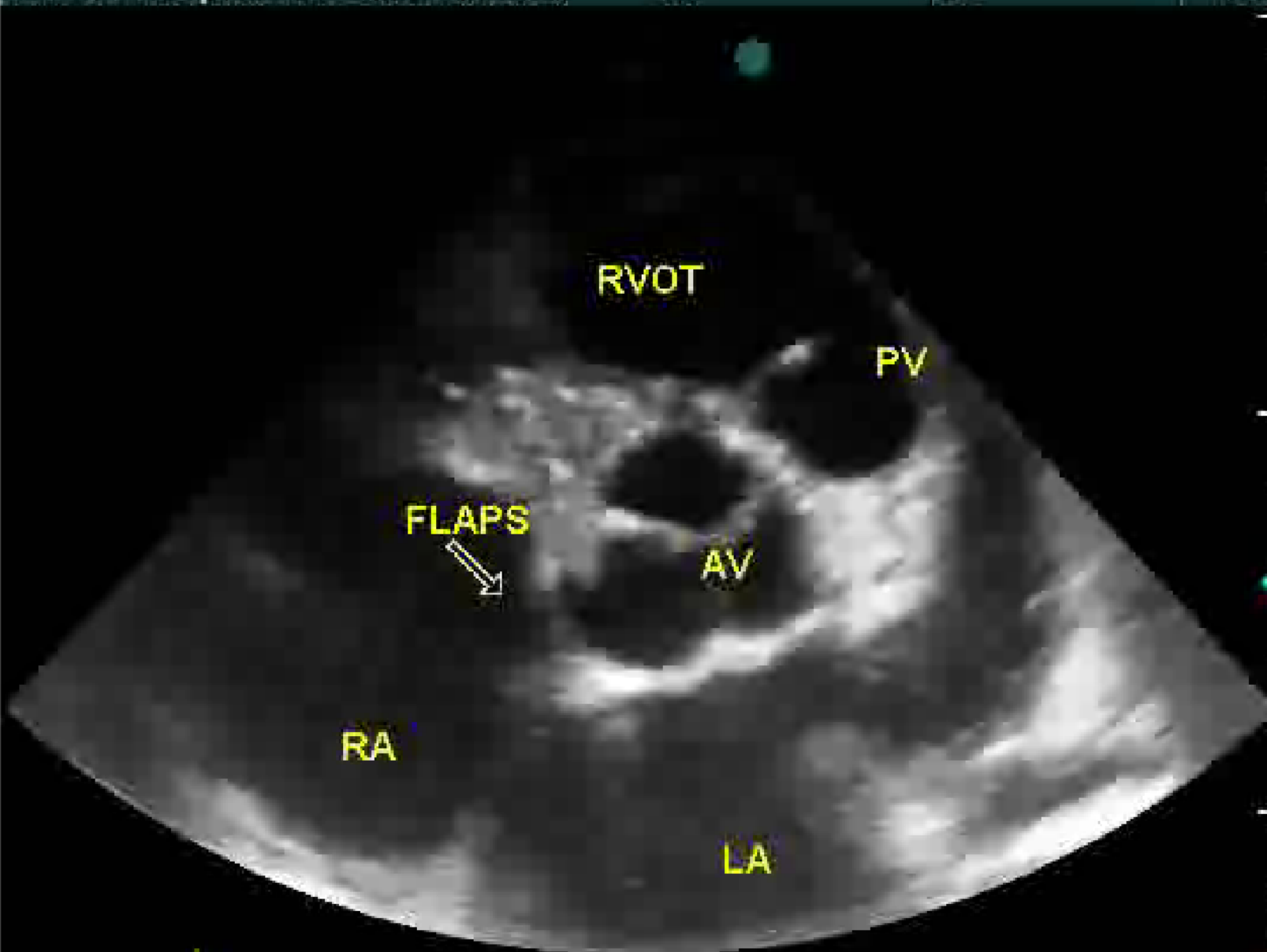


P: 0 dB
Tis: 0.9
MI: 1.1



HR: 101 BPM

2D 12 cm
70 f/s
f: 1.7 MHz H
DR: 65 dB
R: 2.0 G: 47



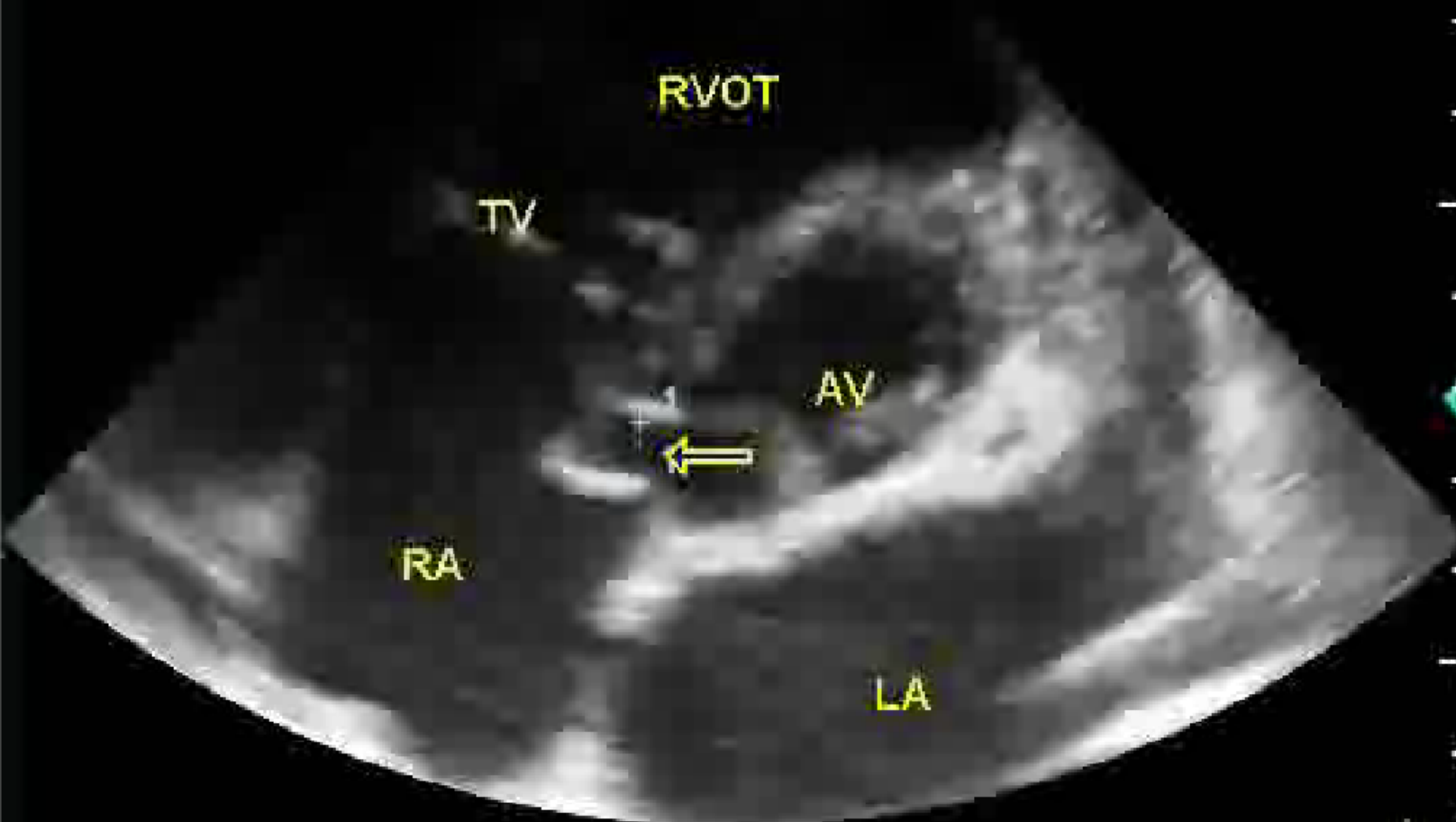
P: 0 dB
Tis: 0.9
MI: 1.2



HR: 45 BPM

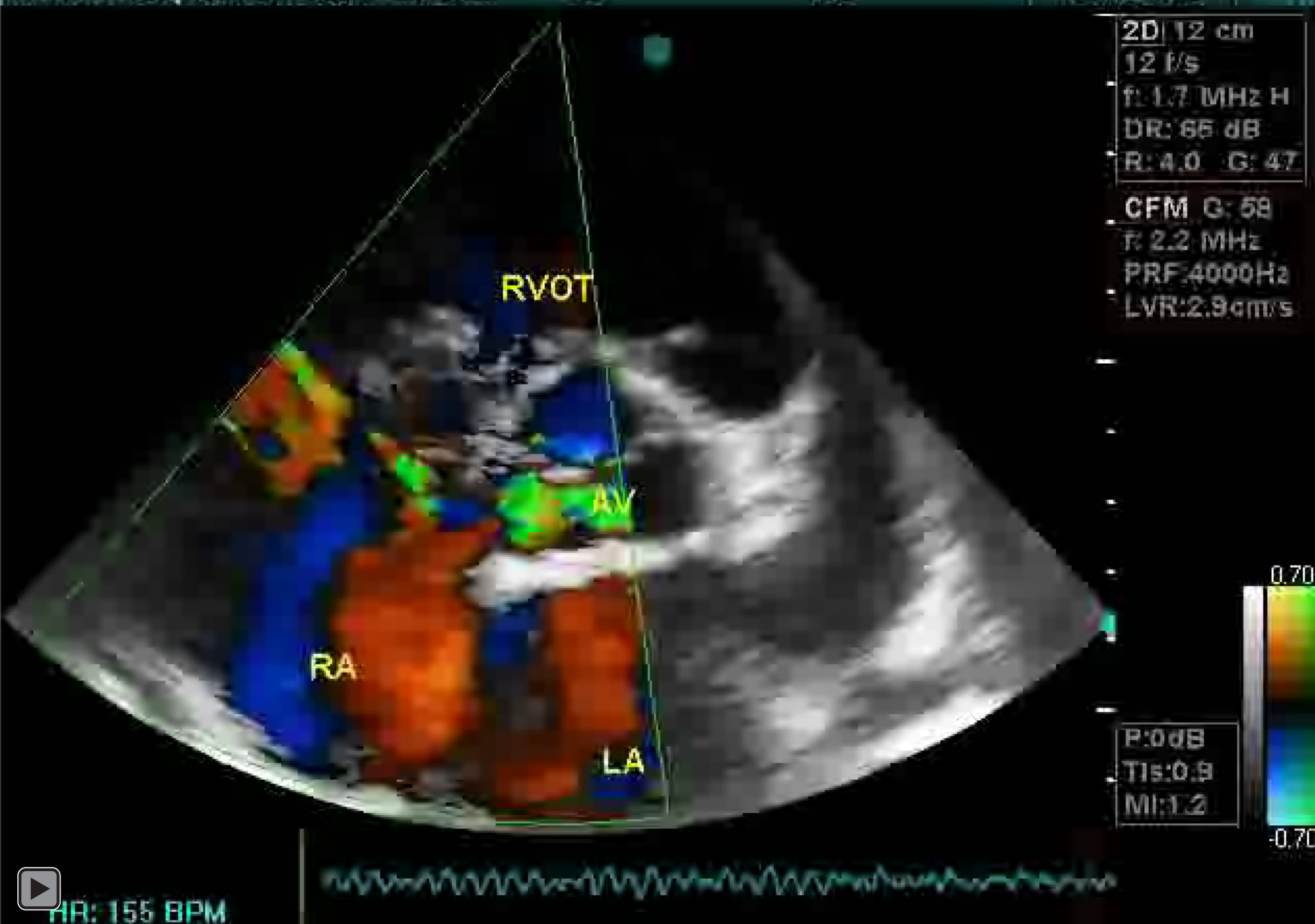
1 Distance= 0.65 cm

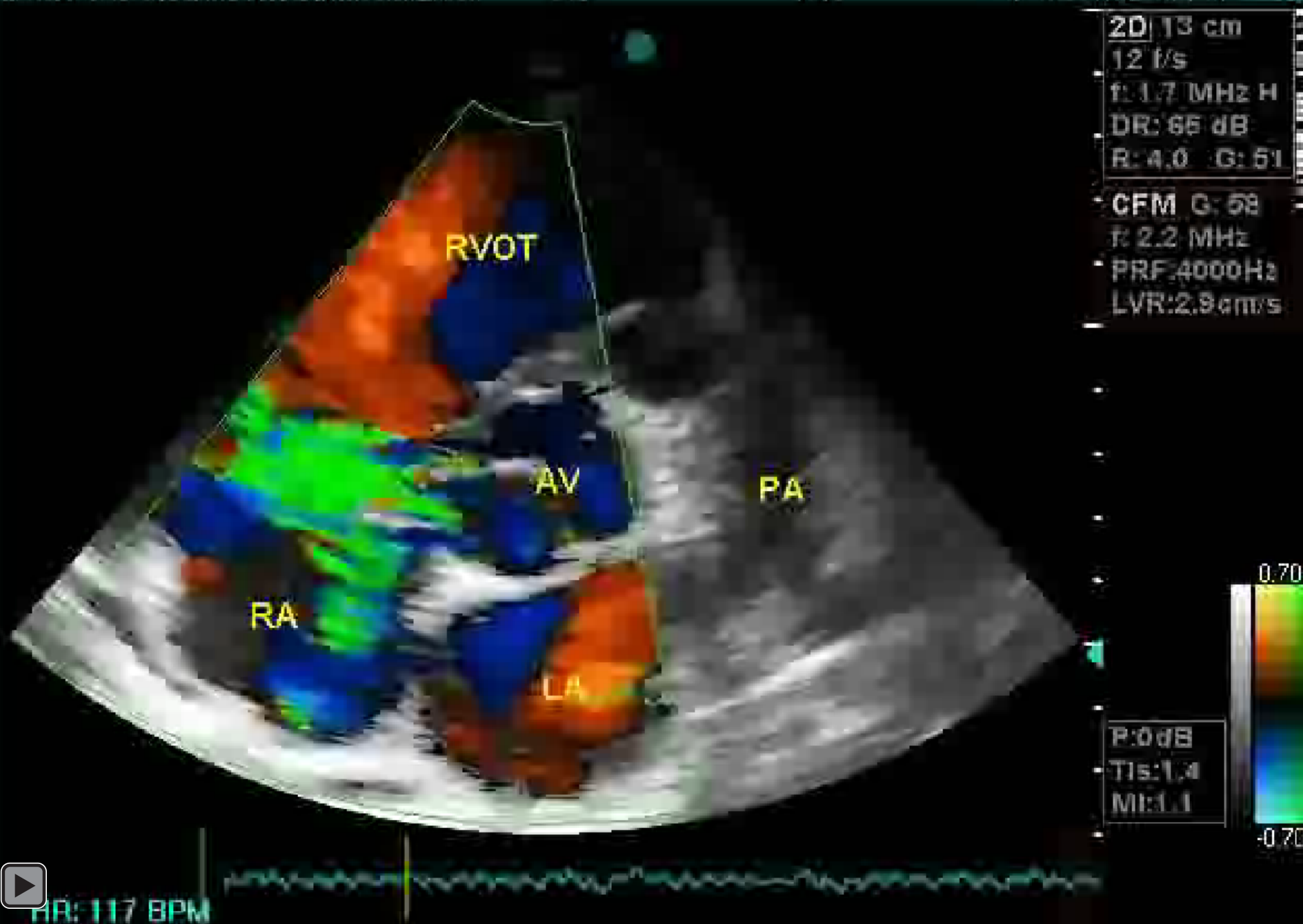
2D 12 cm
70 f/s
f: 1.7 MHz H
DR: 65 dB
R: 2.0 G: 47



P: 0 dB
T: 0.8
M: 1.2

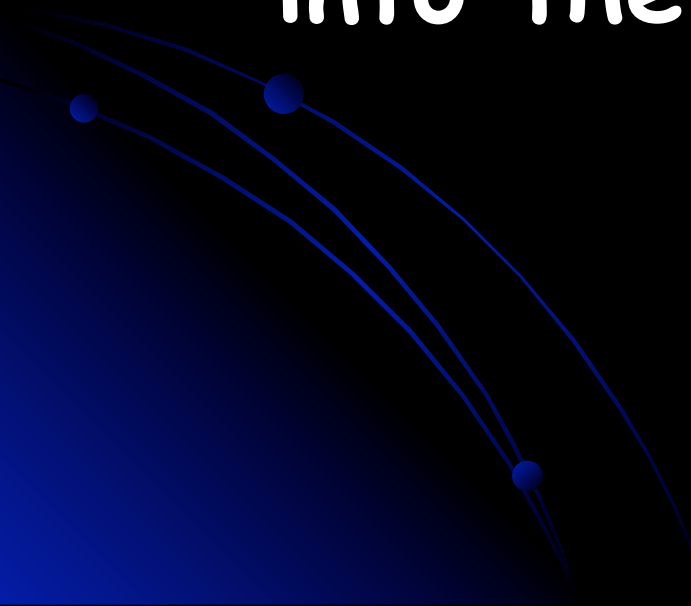






Diagnosis:

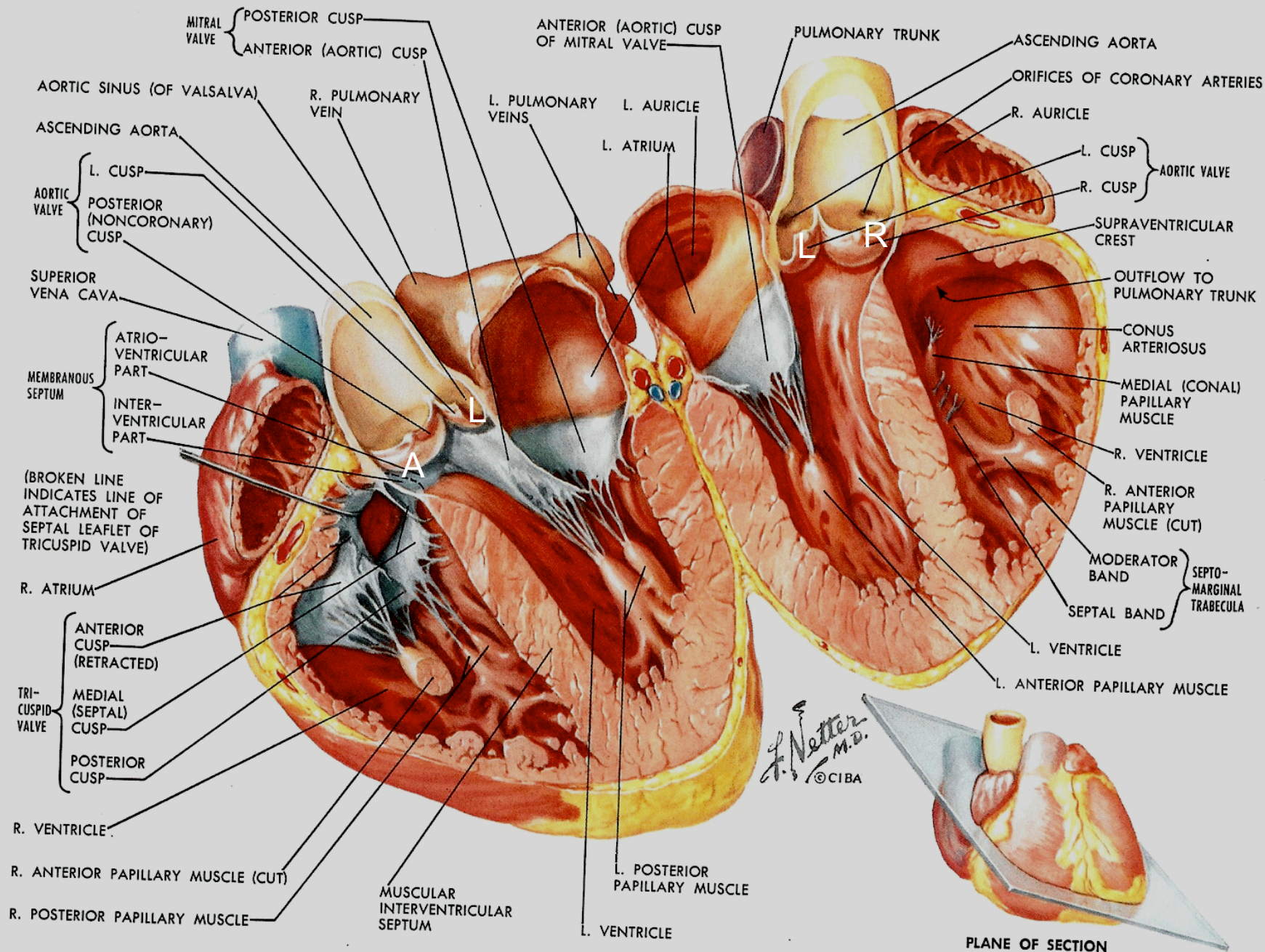
Ruptured aneurysm of the
acoronary sinus of Valsalva
into the right atrium

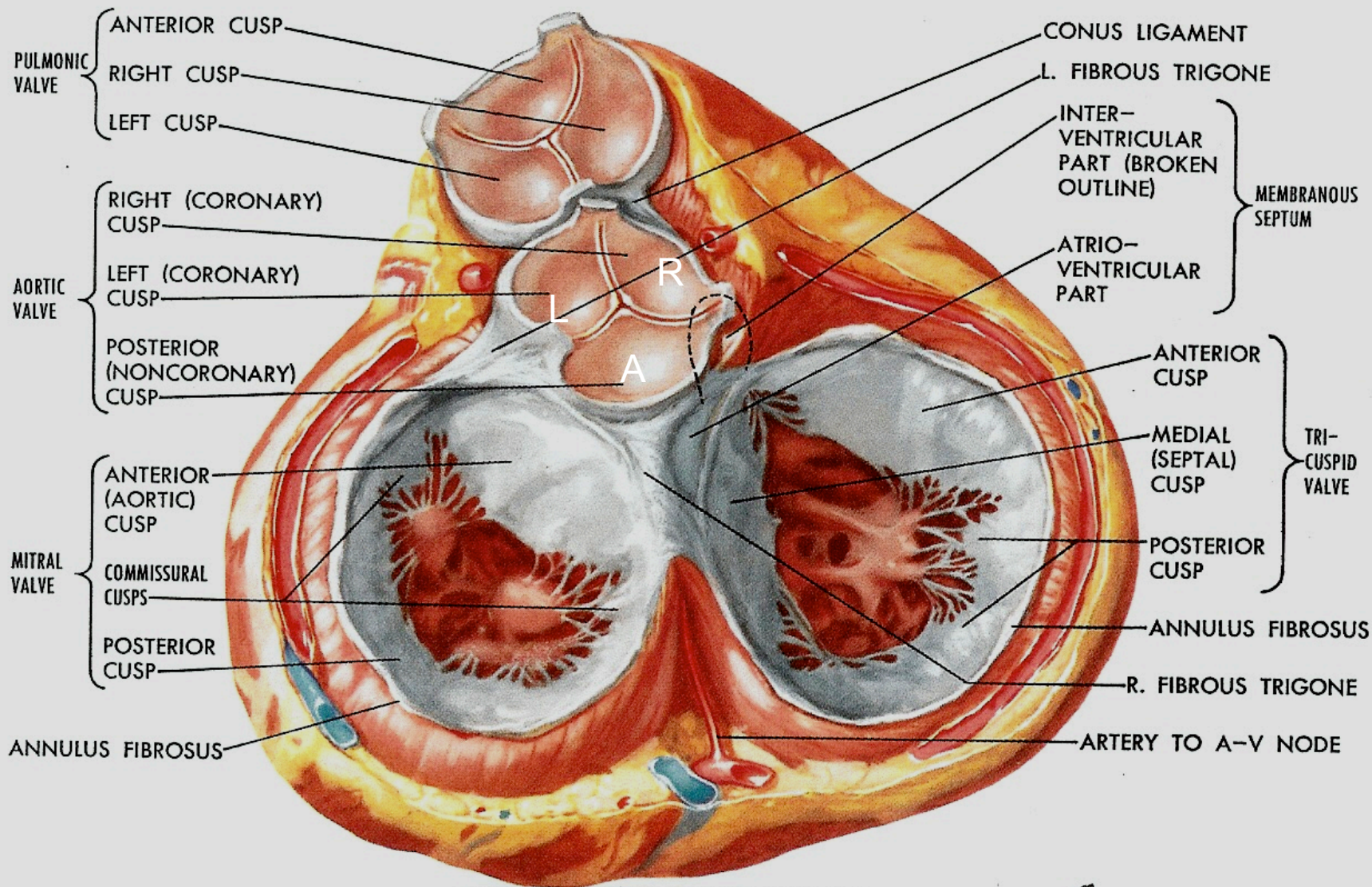


What is a sinus Valsalva aneurysm ?

What are sinuses of Valsalva?

- Normal dilatations of the aortic root immediately above the aortic cusps.
- These 3 sinuses are named according to their relation to the coronary arteries:
 - Left coronary sinus.
 - Right coronary sinus.
 - Acoronary sinus.
- A marked focal bulging of the sinus is called an aneurysm of the sinus Valsalva.





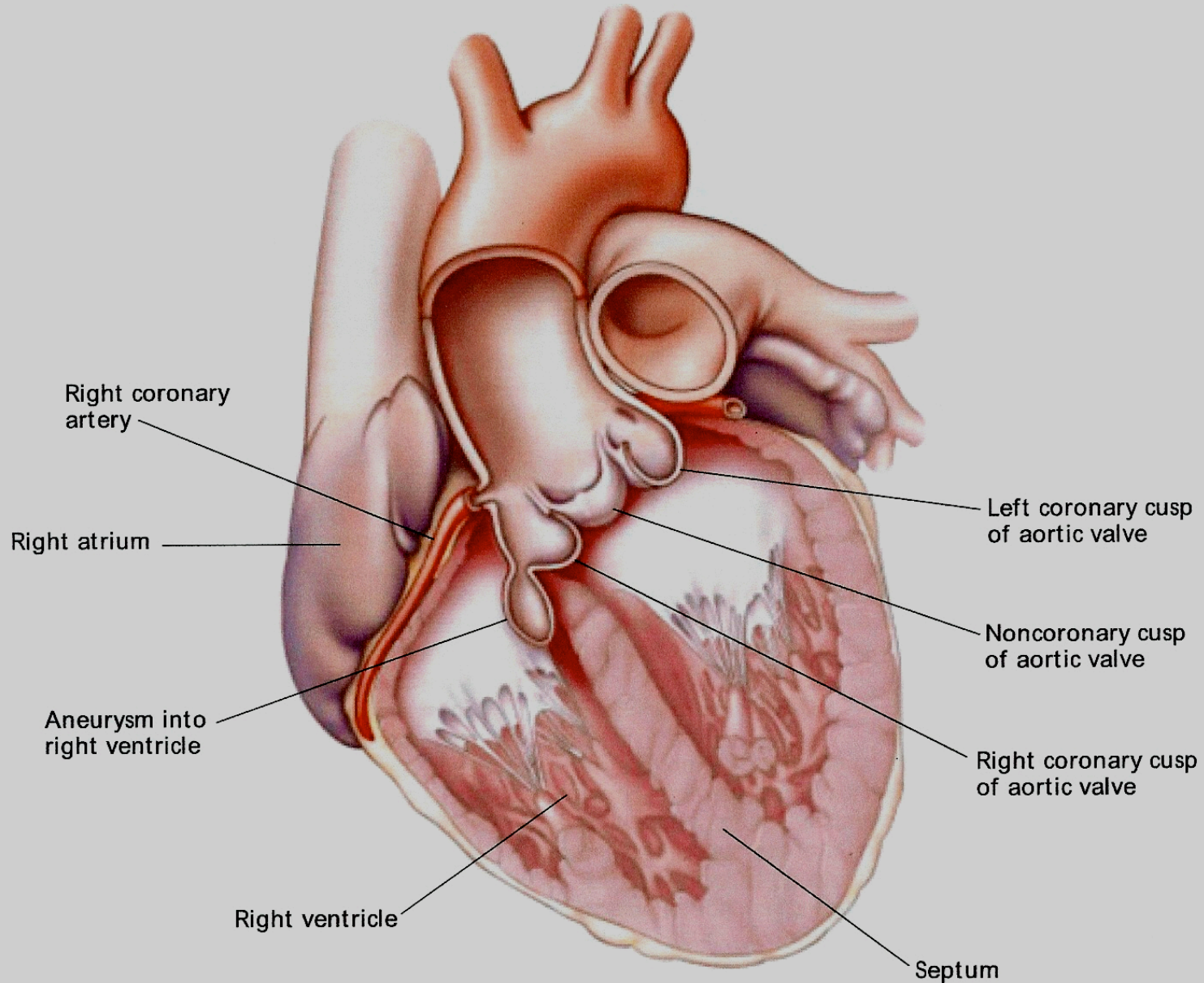
THE HEART IN DIASTOLE: VIEWED FROM BASE WITH ATRIA REMOVED

What is the cause a sinus Valsalva aneurysm ?

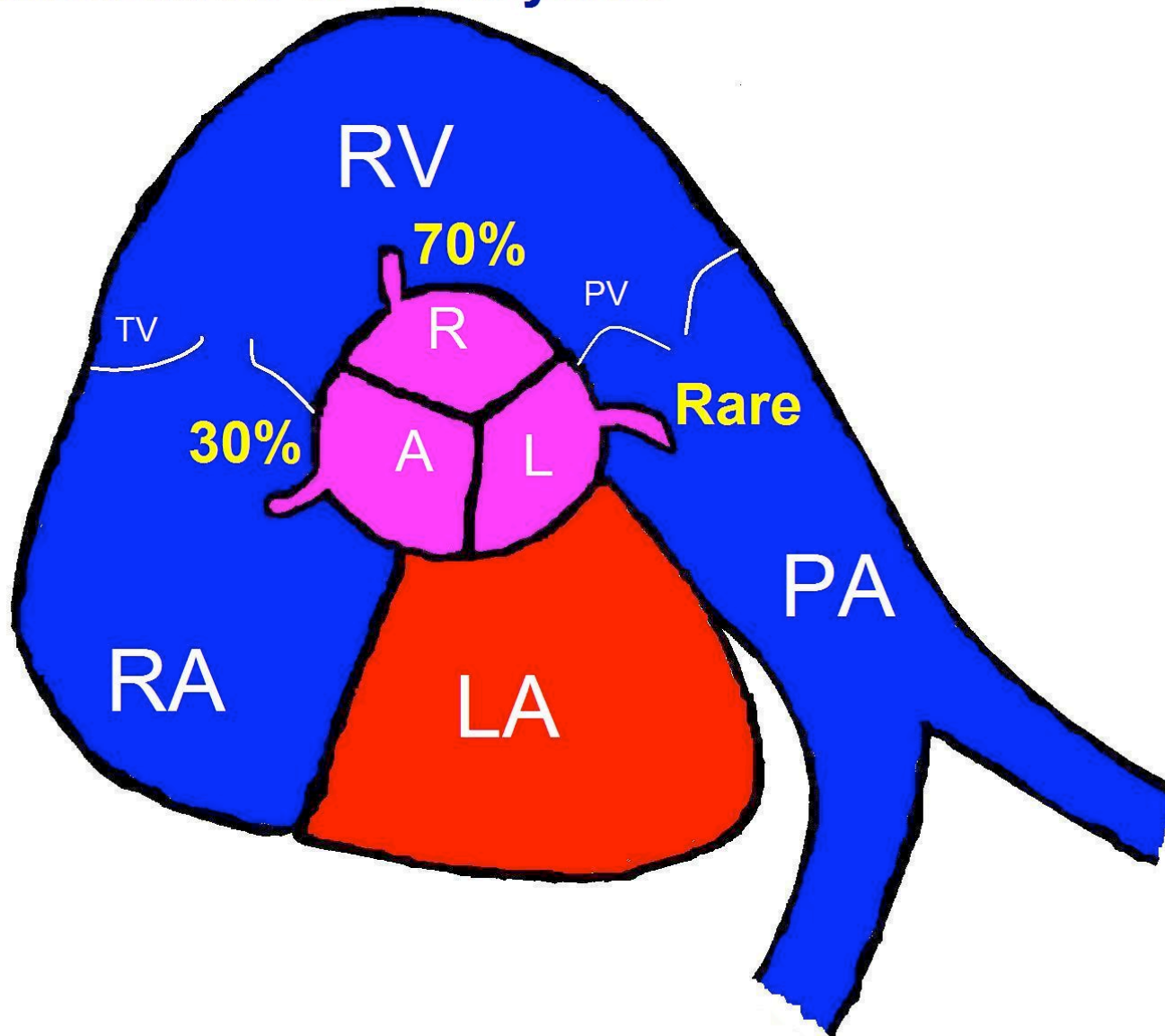
It is only congenital:

- A localized area in the aortic wall lacking a media layer (Absence of elastic fibers), causing a localized wall weakness.
- The weak area gives way to aortic pressure to form an aneurysm which extends like a „windsock“ into an adjacent heart chamber.
- The Aneurysm may rupture either spontaneously or in reponse to a heavy isometric effort, causing an *aorto-cardiac fistula*.
- 3 Times more common in men than in women.
- Occurs in second and third decade of life.

■ Aneurysm of the sinus of Valsalva



Origines and sites of perforation of aneurysms



Clinical presentations

- If unruptured:

- No symptoms.
- Or local compression: AV node
Coronary artery
- Aortic valve regurgitation.

- If small perforation:

Small shunt with no symptoms.

- If acute large rupture:

Large acute L-R shunt with CHF.

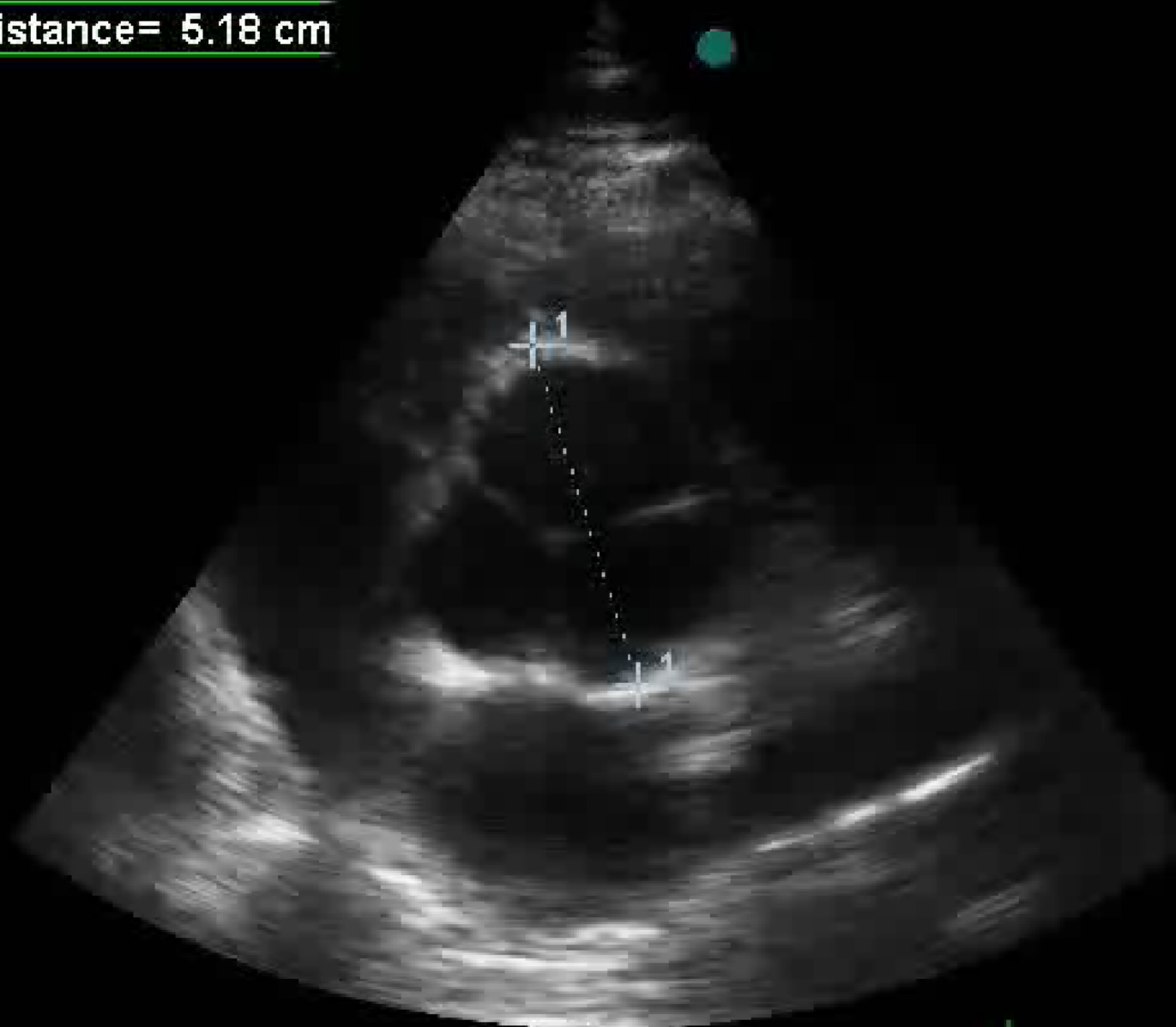
Differential Diagnosis

**Diffuse dilatation of all 3
sinuses of valsalva:**

- It is a separate, also congenital clinical entity.**
- Usually all 3 sinuses are diffusely dilated.**
- Is associated with connective tissue disorder, such as Marfan Syndrome.**

1 Distance= 5.18 cm

2D 15 cm
67 f/s
f: 1.7 MHz H
DR: 65 dB
R: 2.0 G: 51



P: 0 dB
Tis: 0.9
MI: 1.0





2D 15 cm
67 f/s
f: 1.7 MHz H
DR: 65 dB
R: 2.0 G: 51

P: 0 dB
Tis: 0.9
MI: 1.0



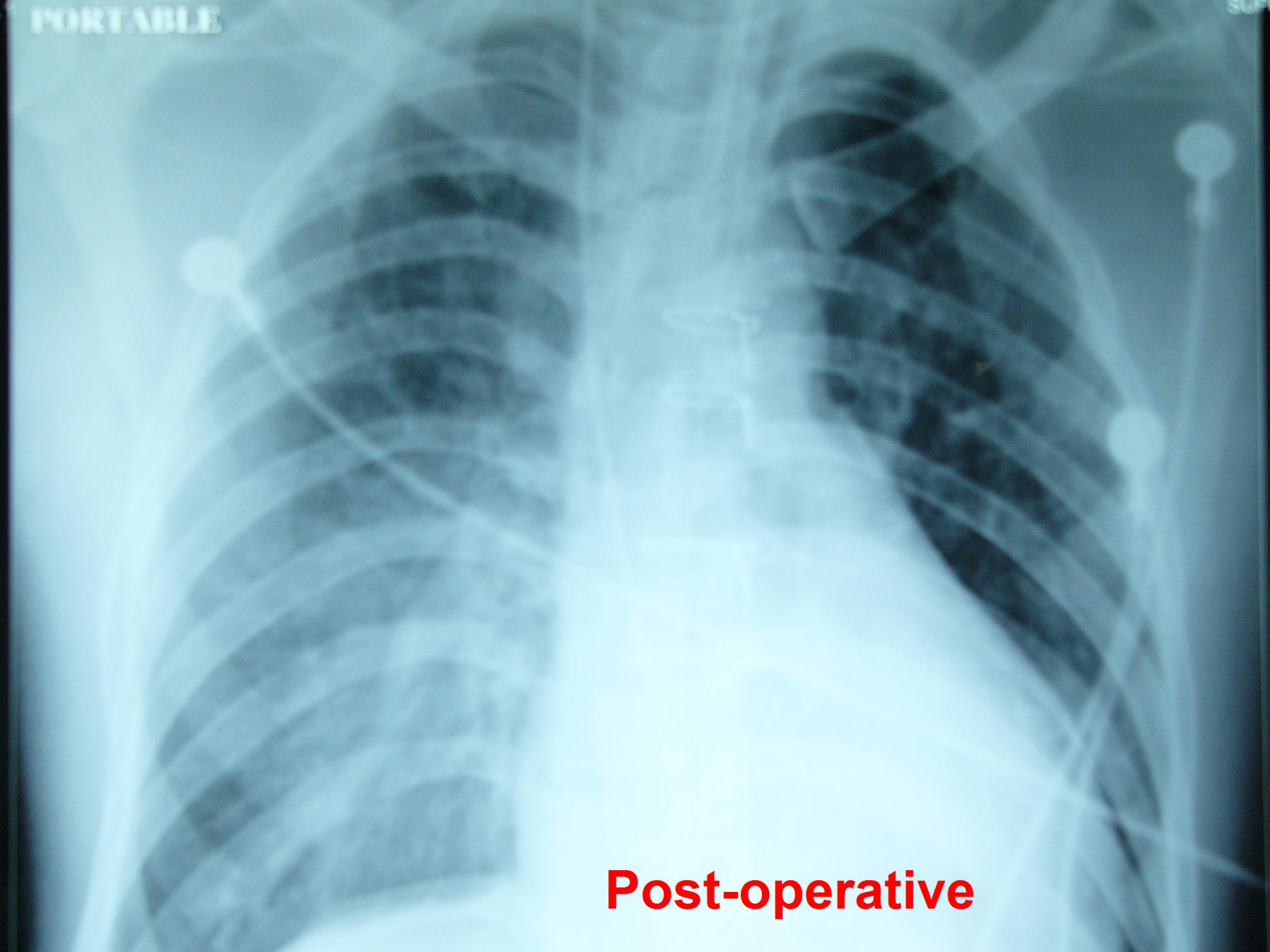
Surgical treatment

The patient was subjected to urgent surgical correction.

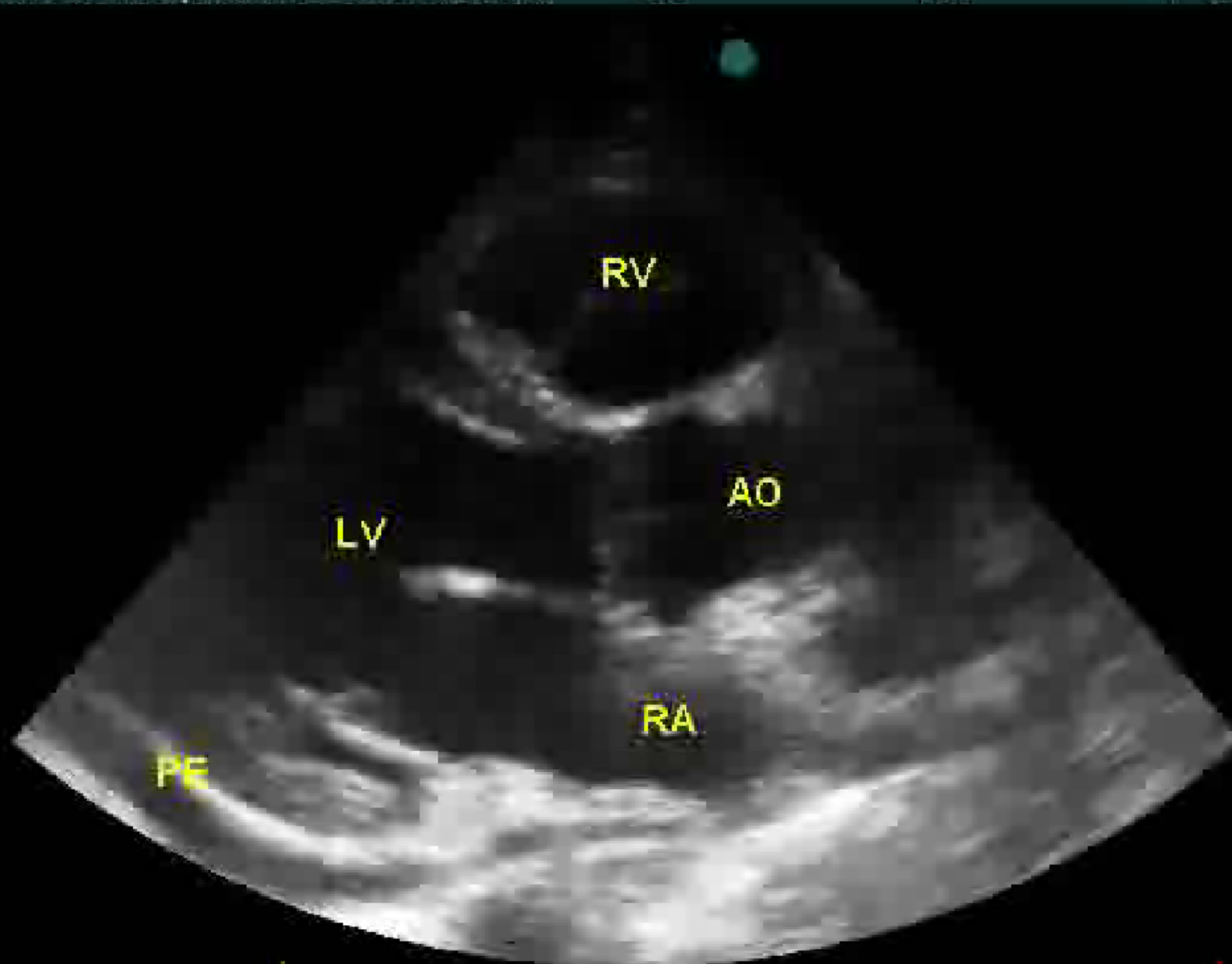
- **Trimming of the aneurysm.**
- **Direct closure by stitching.**
- **Goretex patch suture.**

PORTABLE

Post-operative



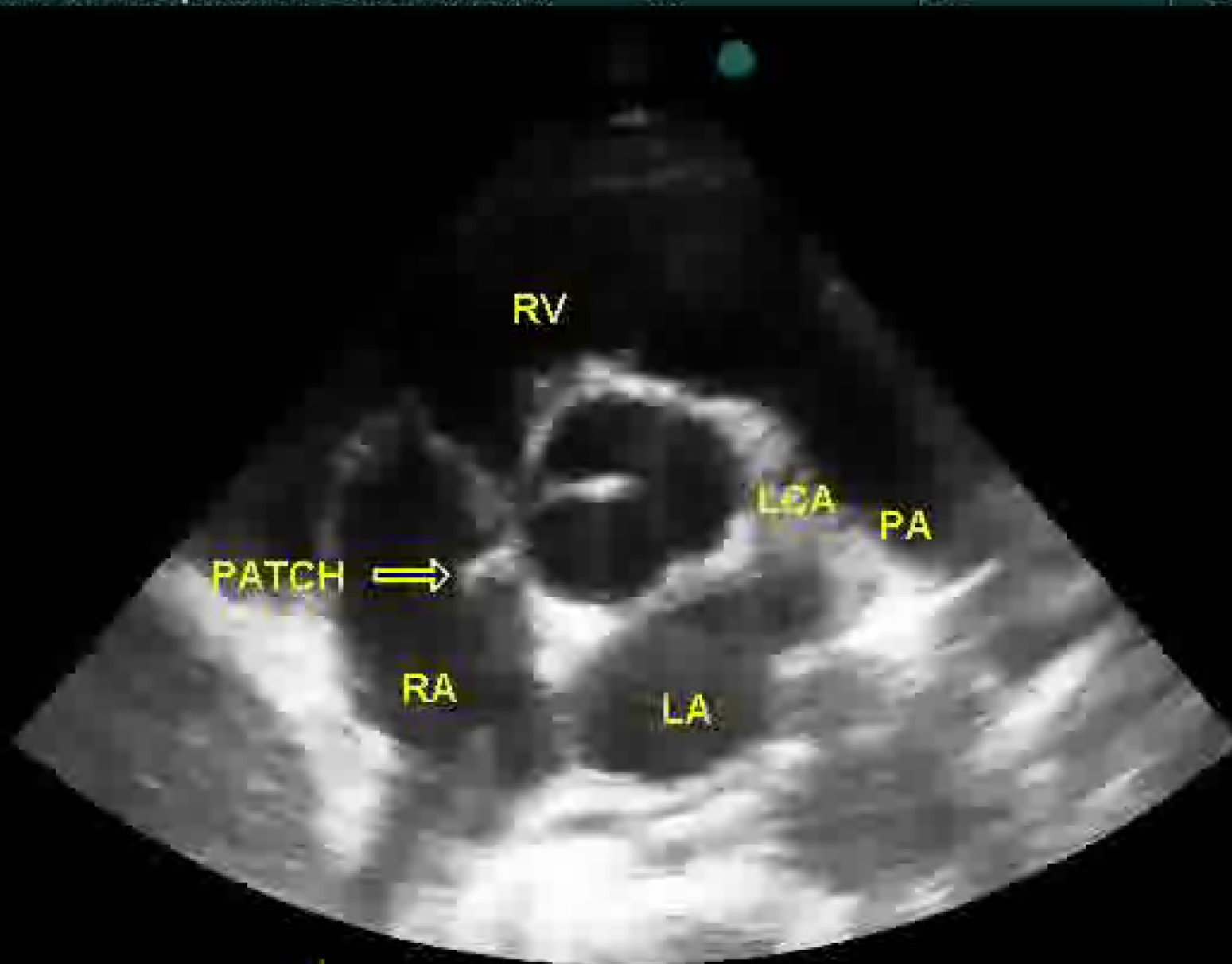
2D 13 cm
59 f/s
f: 1.7 MHz H
DR: 66 dB
R: 2.0 G: 47



P: 0 dB
Tis: 0.9
MI: 1.1



2D 13 cm
59 f/s
f: 1.7 MHz H
DR: 65 dB
R: 2.0 G: 47

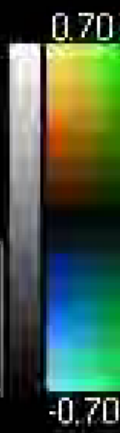
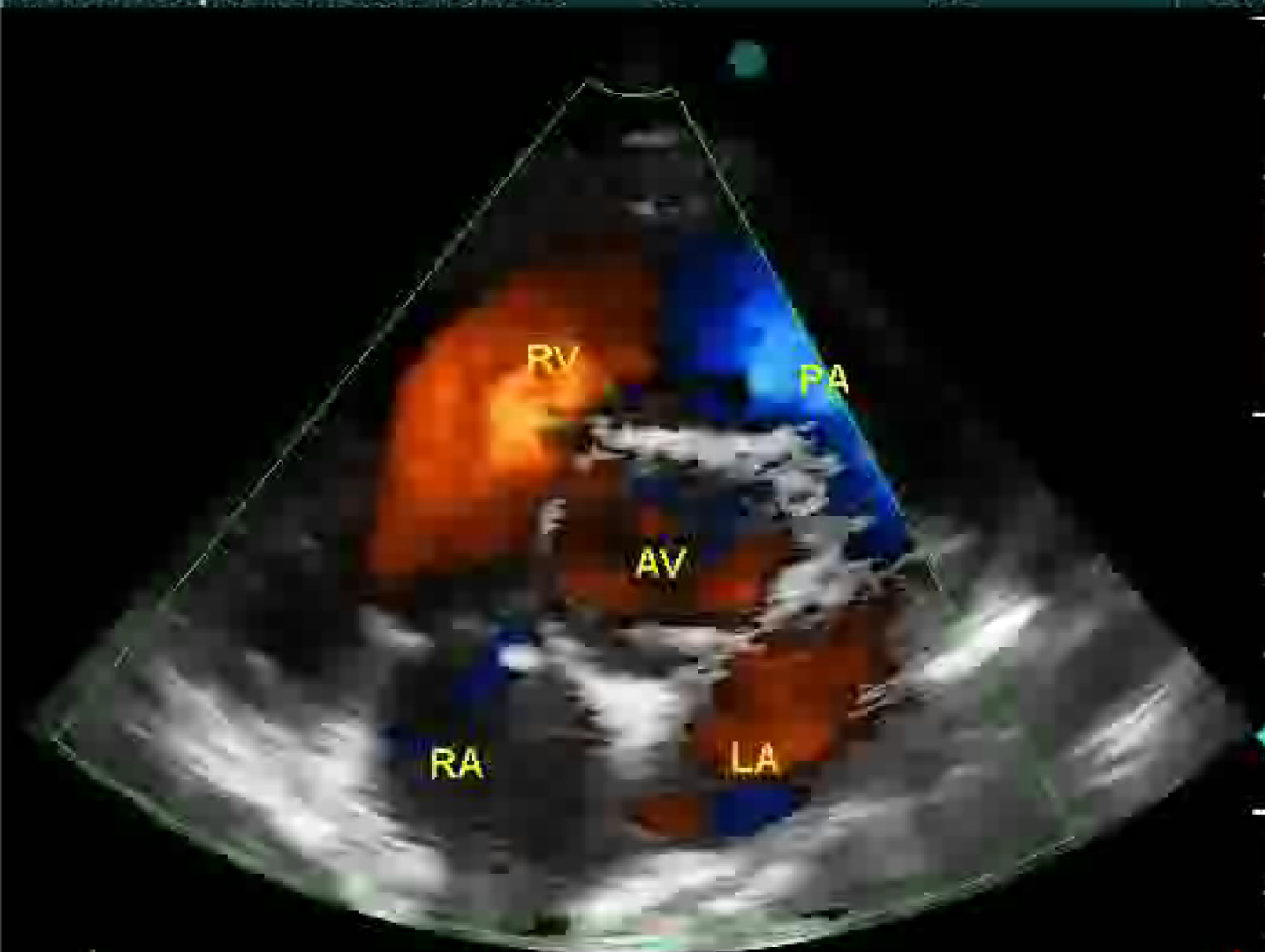


P:0dB
TIs:0.9
MI:1.1



2D 12 cm
9 f/s
f: 1.7 MHz H
DR: 65 dB
R: 4.0 G: 43

CFM G: 58
f: 2.2 MHz
PRF: 4000Hz
LVR: 2.9 cm/s



P: 0 dB
T: 1.1
M: 1.2



HR: 71 BPM

Case 2

**A 42-year-old man with
4th heart sound**



- A 42-year-old caucasian engineer presented for progressive effort induced **shortness of breath** since about 4 months.
- He had consulted several physicians for his complaint and he was repeatedly diagnosed with chest infection and treated as such, with no improvement.
- Upon presentation SOB had progressed to resting dyspnea and was associated with palpitation and perspiration.
- He denied chest pain, limb pain and peripheral edema.

Physical examination

- Patient appeared ill, short of breath.
- BP 110/70 mmHg, HR 120 BPM, Temp. 36.7 C, Transdermal O2 sat. 89-90%. Weight 120 kg at a height of 189 cm.
- Chest was clear.
- No recognizable jugular vein congestion.
- Cardiac auscultation revealed a loud 4th heart sound, over the left lower sternal border. No murmurs.
- No hepatomegaly, no peripheral edema.

ECG

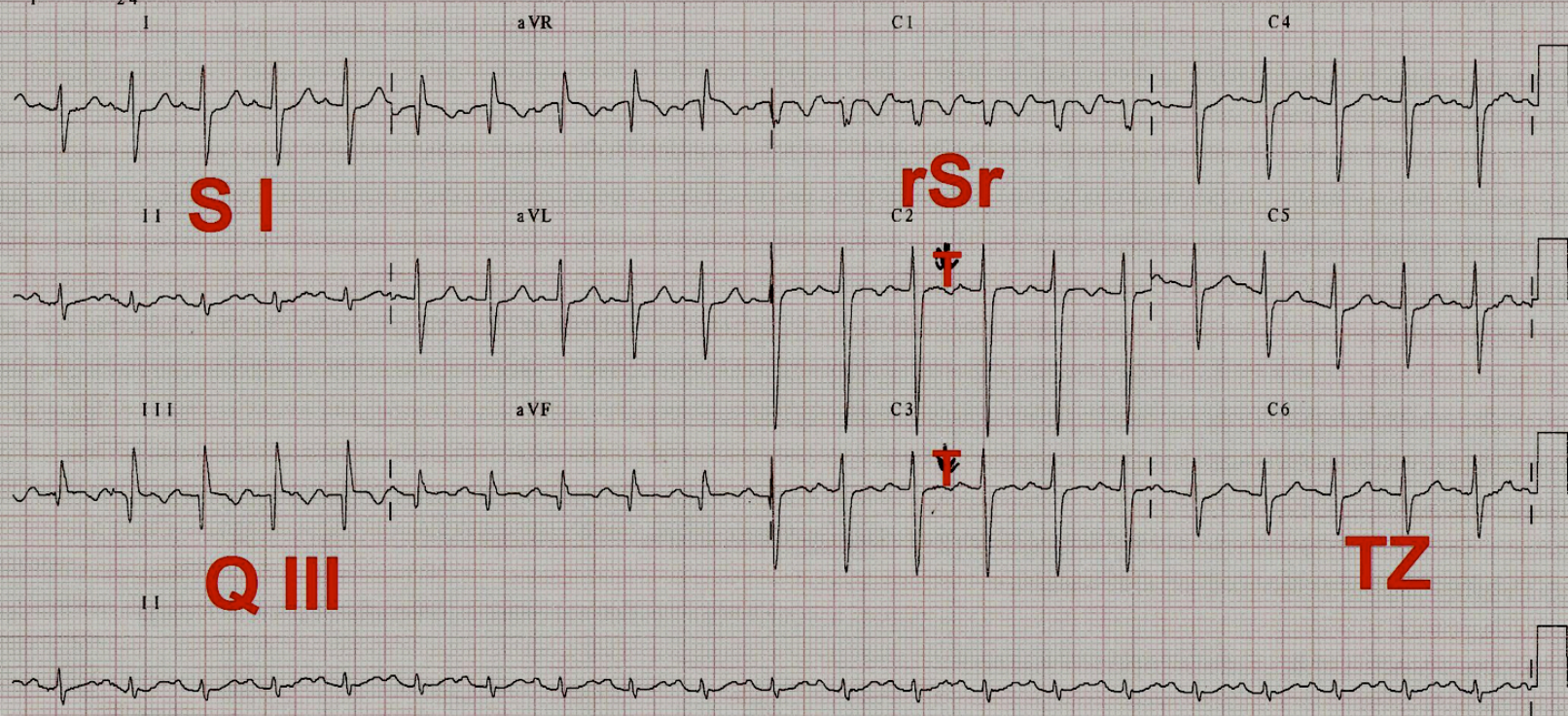
Rate	126
PR	124
QRSD	93
QT	303
QTc	439

-- Axis --	
P	- 64
QRS	144
T	- 24

(M) [REDACTED]
481583
South Africa Private
27/01/1963
Mar 1 2005

9/2/08
3:40pm

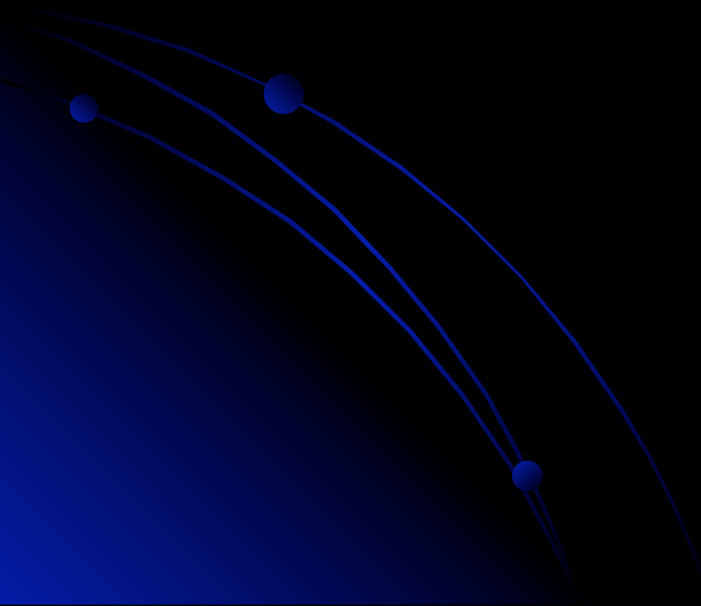
1002



25 mm/s 10 mm/mV γ 0.15 Hz - 40 Hz HP709 05004

ECG

- **Right ventricular hypertrophy and pressure overload.**
 - SI-QIII so-called McGuinn-White sign.
 - T inversion in right precordial leads.
 - Transitional zone (TZ) shift to V6.

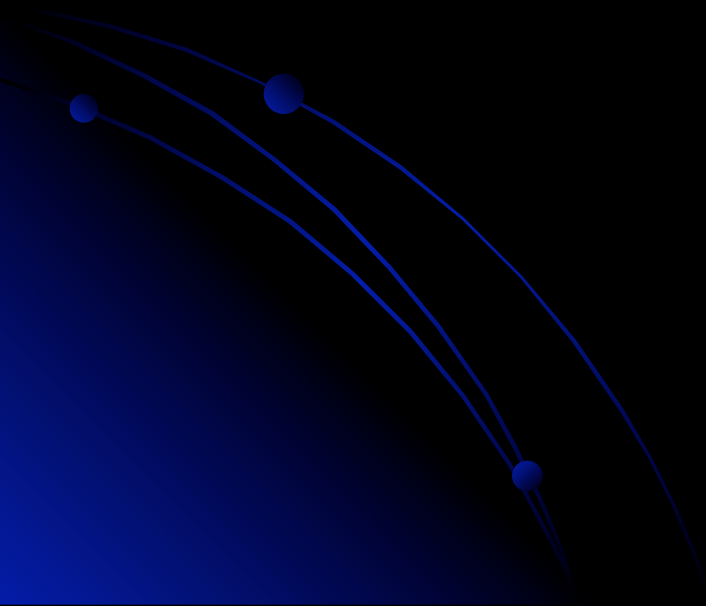


Chest X-Ray



Chest X-Ray

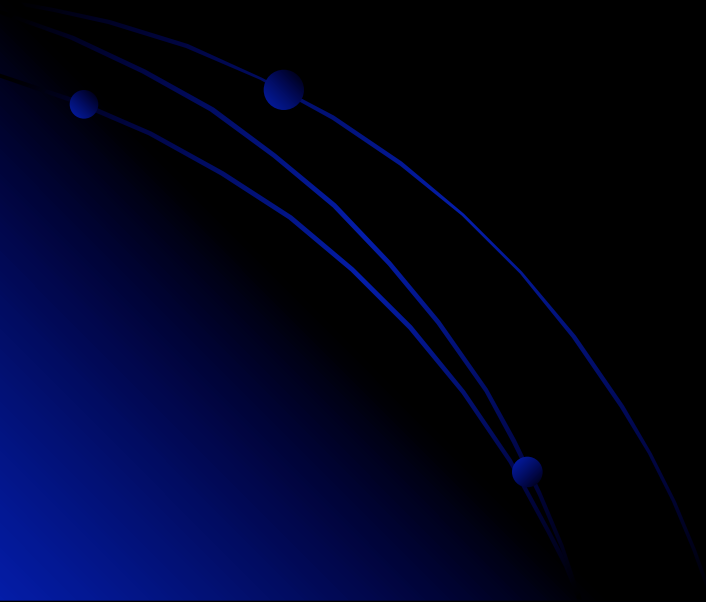
- **Bulging of the right heart edge indicating possibility of right atrial dilation.**



Lab tests

- CBC, E'lytes, blood sugar, creat., urea, PTT and PT-INR: All normal.
- Uric acid: ↑10.2 mg/dl (7.0).
- LDH: ↑ 627 U/L (240).
- CPK: ↑ 227 U/L (170), with normal CKmB.
- Troponin: ↑ 0.4 (0.01).
- D-Dimer: ↑↑ 7 '543 ng/ml (< 500).

Echocardiogram



2D 17 cm
57 f/s
f: 1.7 MHz H
DR: 65 dB
R: 2.0 G: 47

RV

LV

RA

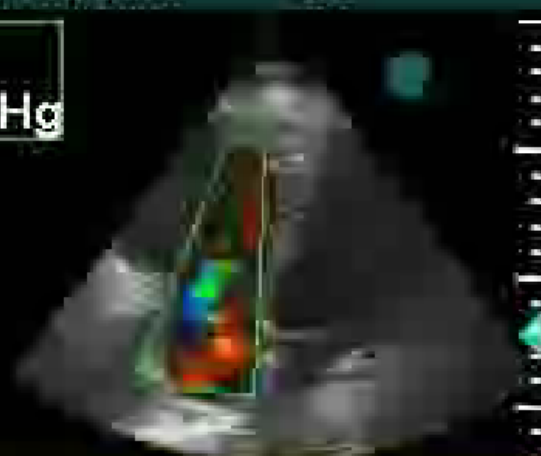
LA

P: 0 dB
Tis: 1.0
MI: 1.0



HR: 119 BPM

1 TRpeakV= 3.59 m/s
PeakPG = 51.66 mmHg



2D 17 cm
28 f/s
f: 1.7 MHz H
DR: 65 dB
R: 4.0 G: 47

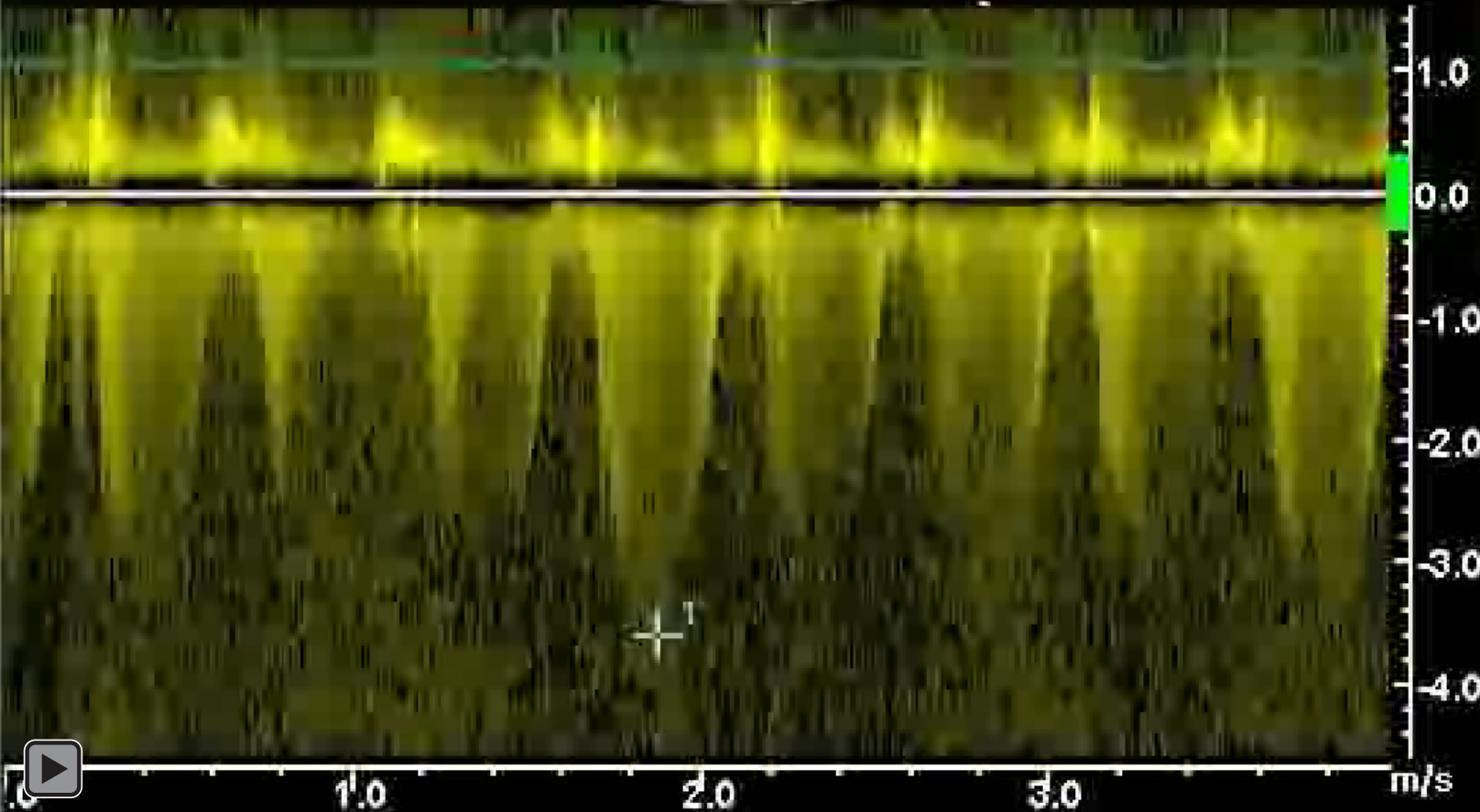
CFM G: 58
f: 2.2 MHz
PRF: 4000 Hz
LVR: 2.9 cm/s

CW G: 78.5
f: 1.82 MHz
VEL: 6.1 m/s
LVR: 32 cm/s
0: 0°

P: 0 dB
Tis: 0.8
MI: 0.1

0.70

-0.70

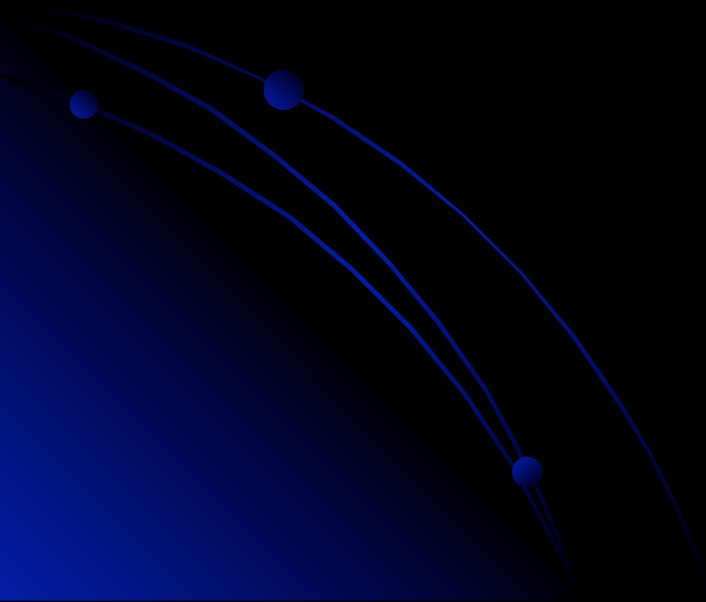


Echocardiogram

- Dilation of right ventricle and right atrium with compression of left chambers.
- Reduced contractility of right ventricle.
- Tricuspid regurgitation.
- Elevation of pulmonary pressure to 55-60 mmHg.

Right ventricular failure due to pressure overload

Doppler Ultrasound of venous system of both legs

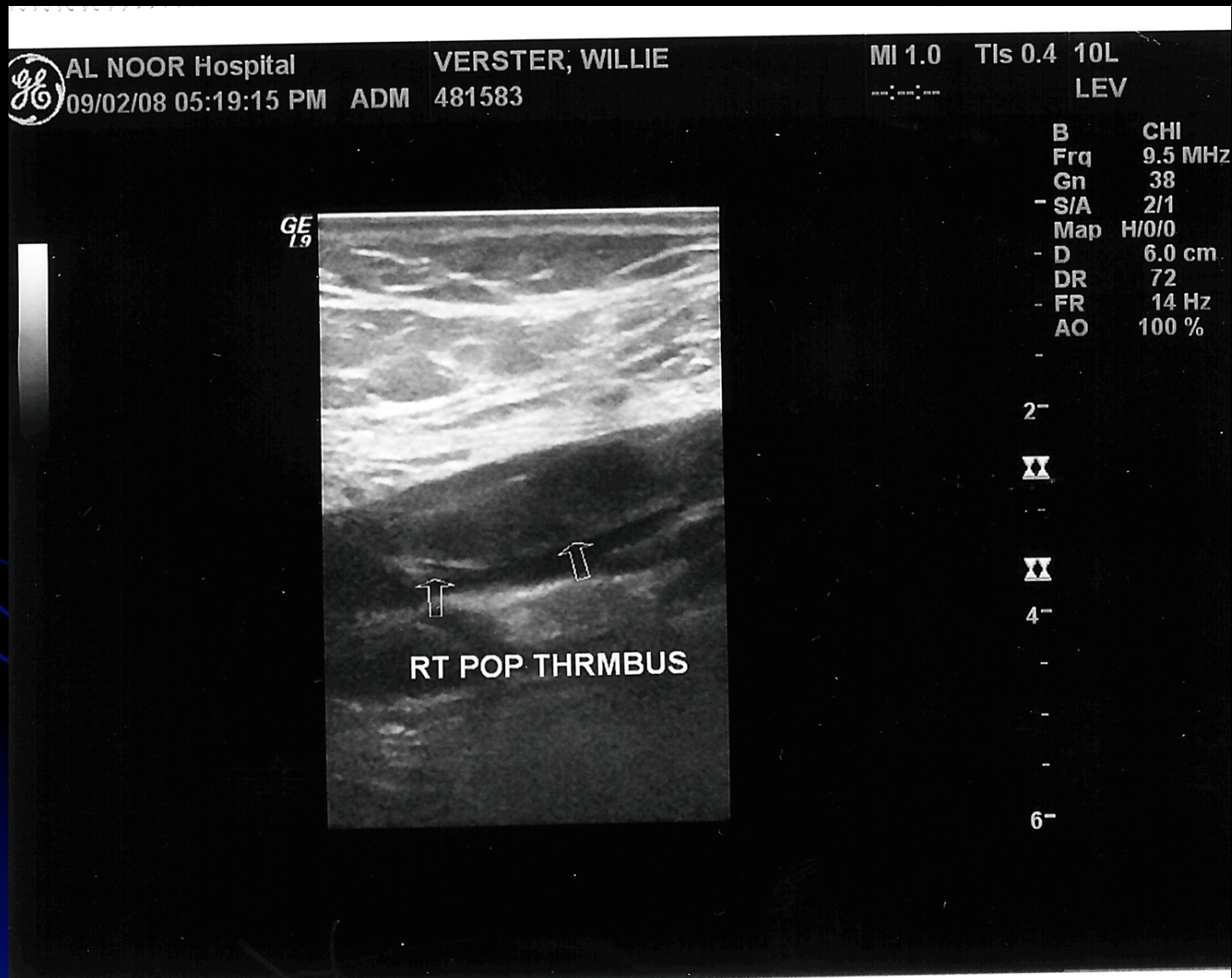


Doppler Ultrasound of venous system of both legs


- **Venous system of left leg: Normal.**
- **Venous system of right leg:**
 - **Large thrombus of right popliteal vein extending caudally to the deep calf veins, with residual eccentric lumen and increased closure pressure.**

Findings corresponding to chronic partially recanalized DVT.

Doppler study of venous system of right leg

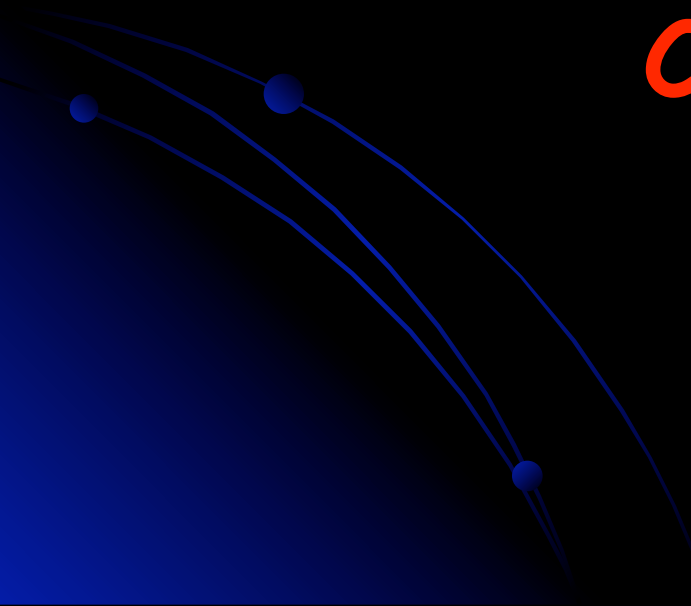


Chest CT-scan

- Ruled out large significant thrombi in the large and segmental pulmonary arteries.
 - Assessment of small pulmonary artery branches is only possible with a high resolution CT scan.
- 

Diagnosis:

Chronic (micro)thrombo-
embolic pulmonary
hypertension
CTEPH



Treatment:

Admission to ICU.

- O2 4L/min, bedrest.
- Overlapping anticoagulation with Enoxaparin (Clexane®) 1mg/kg s.c. BD. and Warfarin.
- Spironolactone 50 mg p.o. OD
- Allopurinol 300 mg p.o. OD.

Clinical course

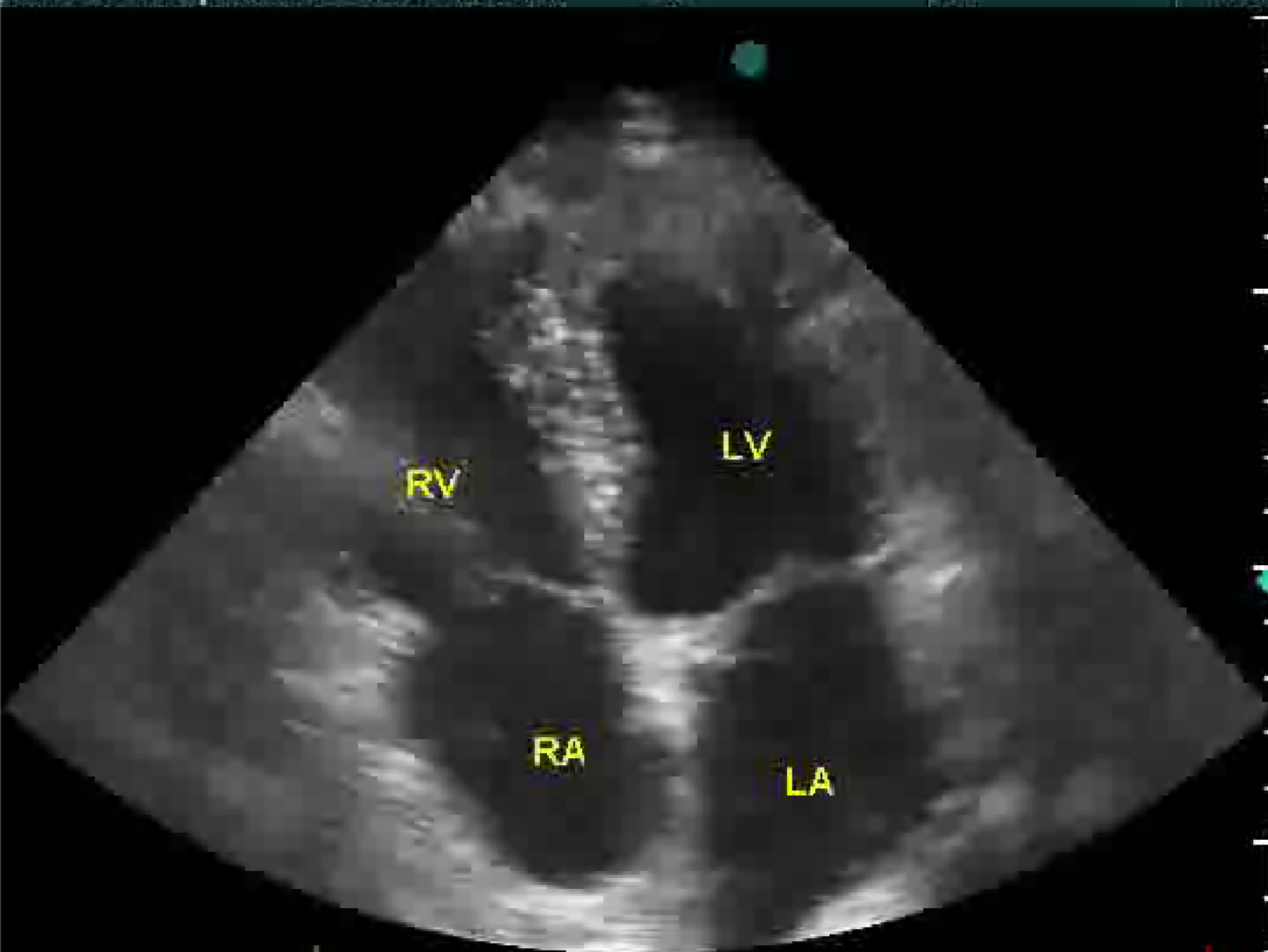
After 8 days of hospitalization:

- Impressive subjective clinical improvement with gradual normalization of O₂ saturation.
- Disappearance of rightventricular 4th heart sound.
- Normalization of pulmonary pressure in echocardiogram.
- Normalization of chest X-Ray.

5 days after discharge:

- Repeat Ultrasound of venous system of left leg: Complete elimination of DVT.

2D 17 cm
50 f/s
f: 1.7 MHz H
DR: 65 dB
R: 2.0 G: 48



P: 0 dB
Tis: 1.0
Mi: 1.0



HR: 119 BPM

1 TRpeakV= 2.11 m/s
PeakPG = 17.76 mmHg

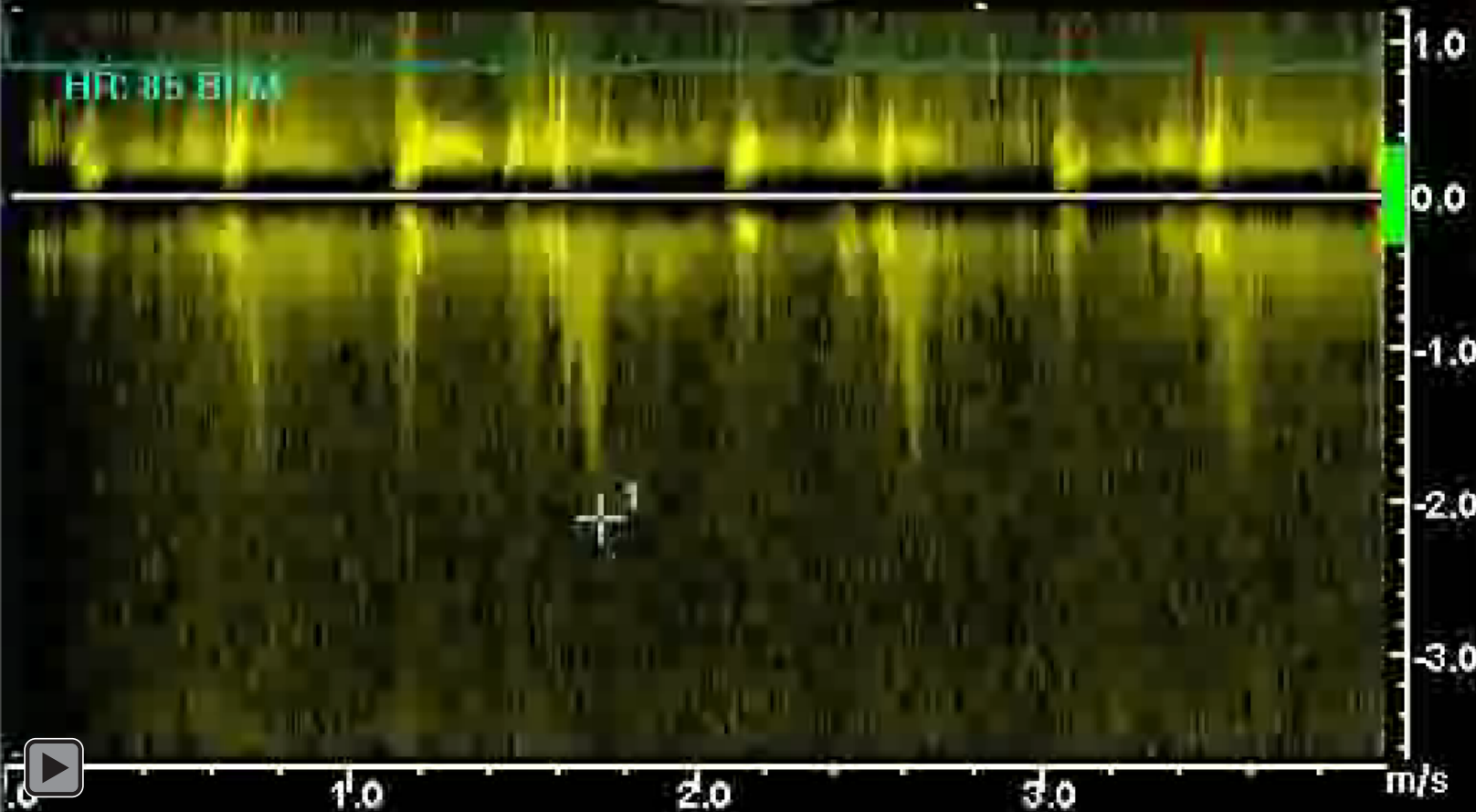


2D 17 cm
18 1/s
f: 1.7 MHz H
DR: 65 dB
R: 4.0 G: 48

CFM G: 58
f: 2.2 MHz
PRF: 4000 Hz
LVR: 2.9 cm/s

CW G: 75.6
f: 1.82 MHz
VEL: 4.9 m/s
LVR: 32 cm/s
0.0°

HFC 85 dB



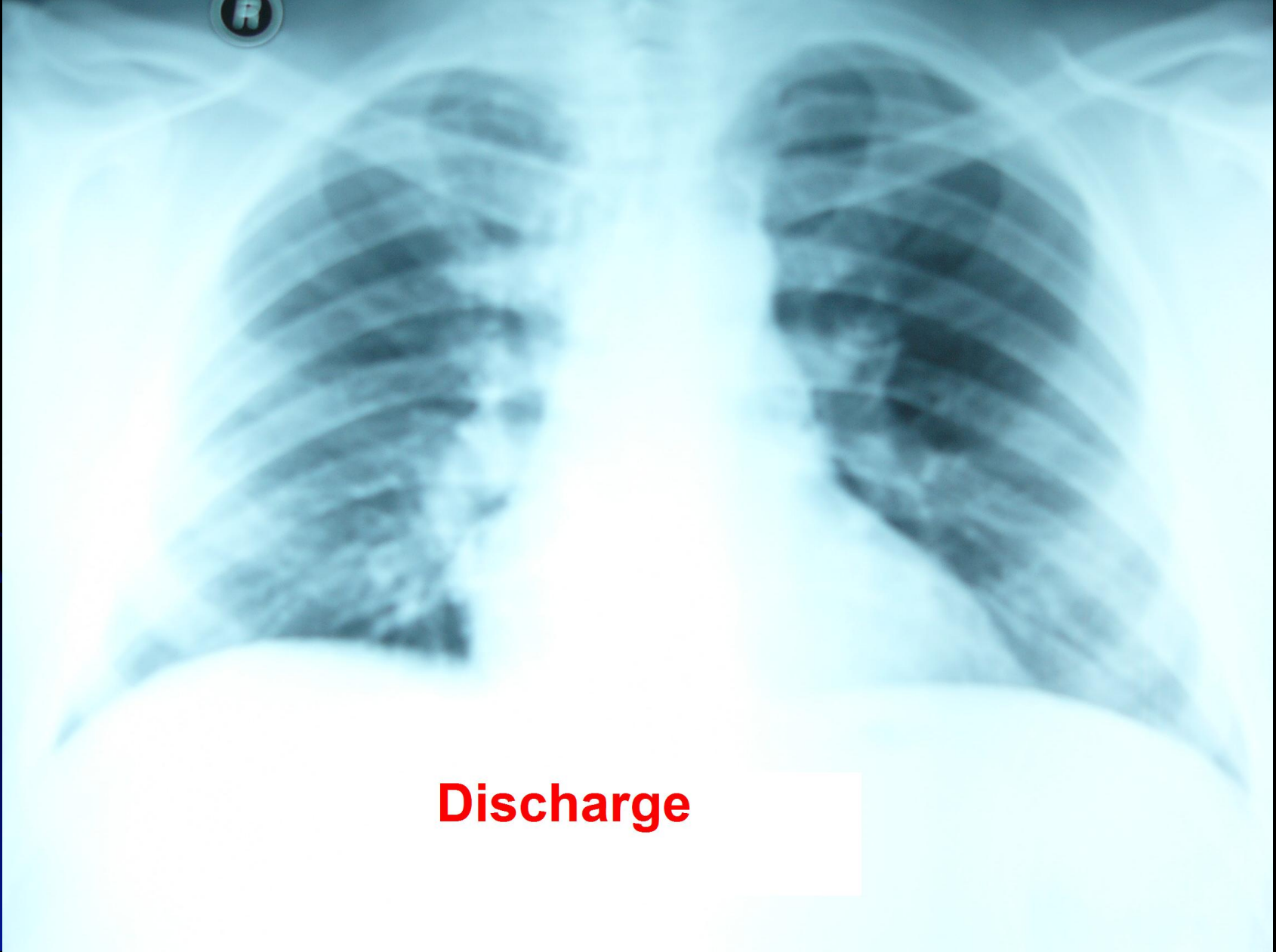
P: 0 dB
Tis: 0.8
MI: 0.1

0.70

-0.70




Chest film



Discharge

Additional lab investigations performed for underlying thrombophilic conditions

- **Protein C deficiency.**
 - **Protein S deficiency.**
 - **Anti thrombin III deficiency.**
 - **Factor V Leiden mutation.**
 - **Antiphospholipid antibodies.**
- 

Additional lab investigations for underlying thrombophilic conditions

- **AT III, Protein C, Protein S, Factor V mutation: All normal.**
- **Antiphospholipid antibodies
(Anticardiolipin antibodies)**
 - **IgG: ↑ 39.9 (N:< 10)**
 - **IgM: ↑ 12.4 (N:< 7.0)**

**Both significantly elevated
corresponding to:**

Hughes Syndrome

