

# **PAH and severe impaired RV function presented with severe MR with cardiogenic shock.**

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# Case presentation

- A 49-y-old woman presented with progressive dyspnea for 2 weeks.

## Past medical Hx:

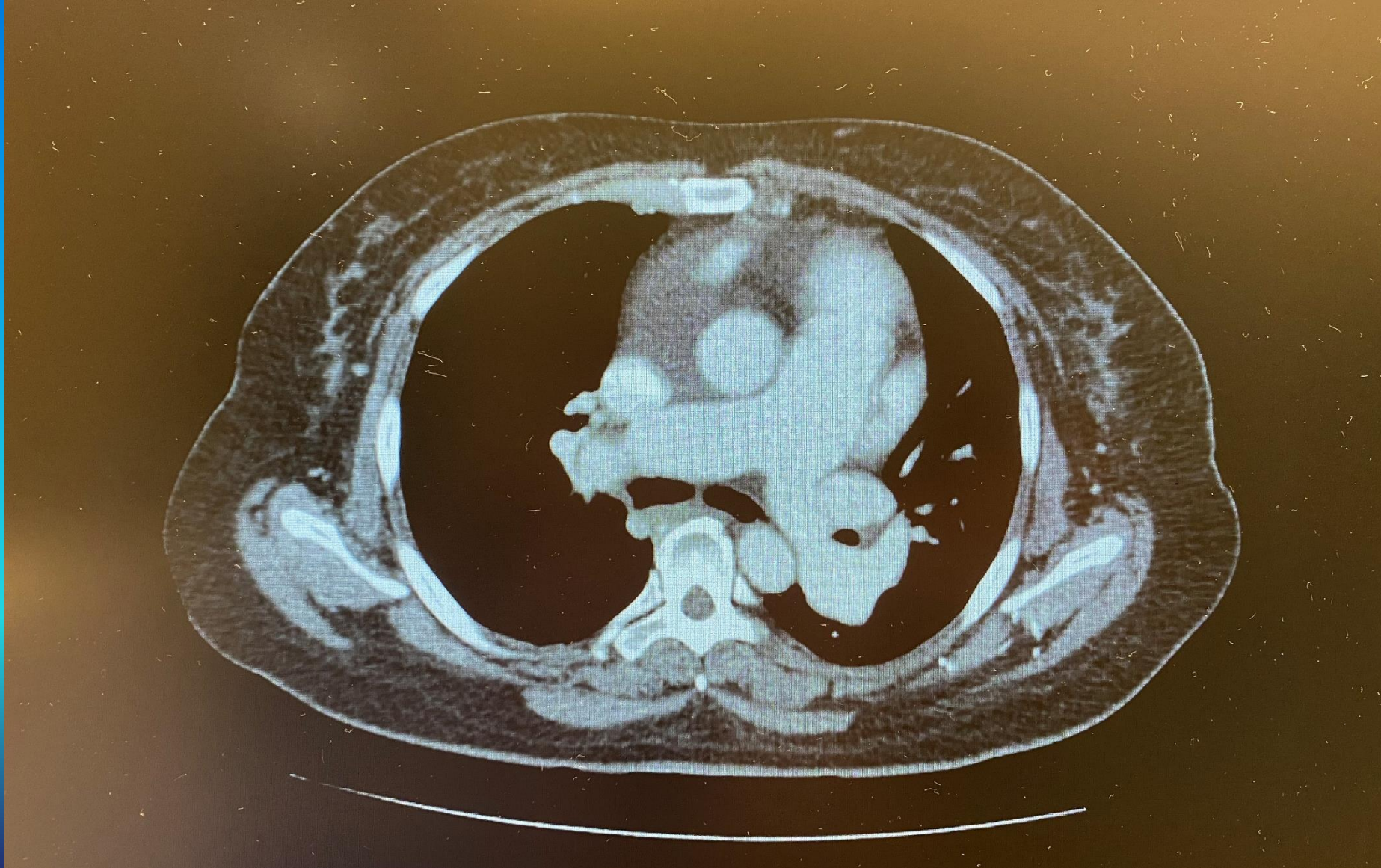
- Aug 2017, she had progressive dyspnea without chest pain or orthopnea or edema. She went to nearby hospital. EKG showed RVOT – PVC and impaired LVEF (EF - 40%). She was referred to another hospital.
- Nov 2017, she was diagnosed of PVC induced cardiomyopathy and s/p RF ablation x 2 times (from 2<sup>nd</sup> hospital). She was told that no anymore PVC and EF was improved.
- Jan 2018, Echo show normal LV systolic function with EF 61%. Normal RV, no evidence of ARVC and normal RA. Moderate TR with presence of pulmonary HT (TR Vmax = 3.8 m/s) and mild-mod MR.

# Case presentation

- During 2018, she still had progressive dyspnea and EKG demonstrated of AF. Echo showed dilated RV and D-shaped septum, TAPSE 0.85 cm, TR Vmax 2.1 m/s. Suspected pulmonary HT (pre-capillary PH) was diagnosed.
- CTPA was done and showed dilated MPA, filling defect at both lower lobe. She was diagnosed of Peripheral CTEPH and warfarin was initiated.
- During 2019-2020, she had multiple admission due to CHF and RV dysfunction.
- Last Echo: Jan 2020; dilated RV and severely impaired RV systolic function, LVEF = 40%. Severe TR and mild to moderate MR. Dilated IVC.
- CTPA follow-up, no anymore pulmonary embolism.



# СТРА



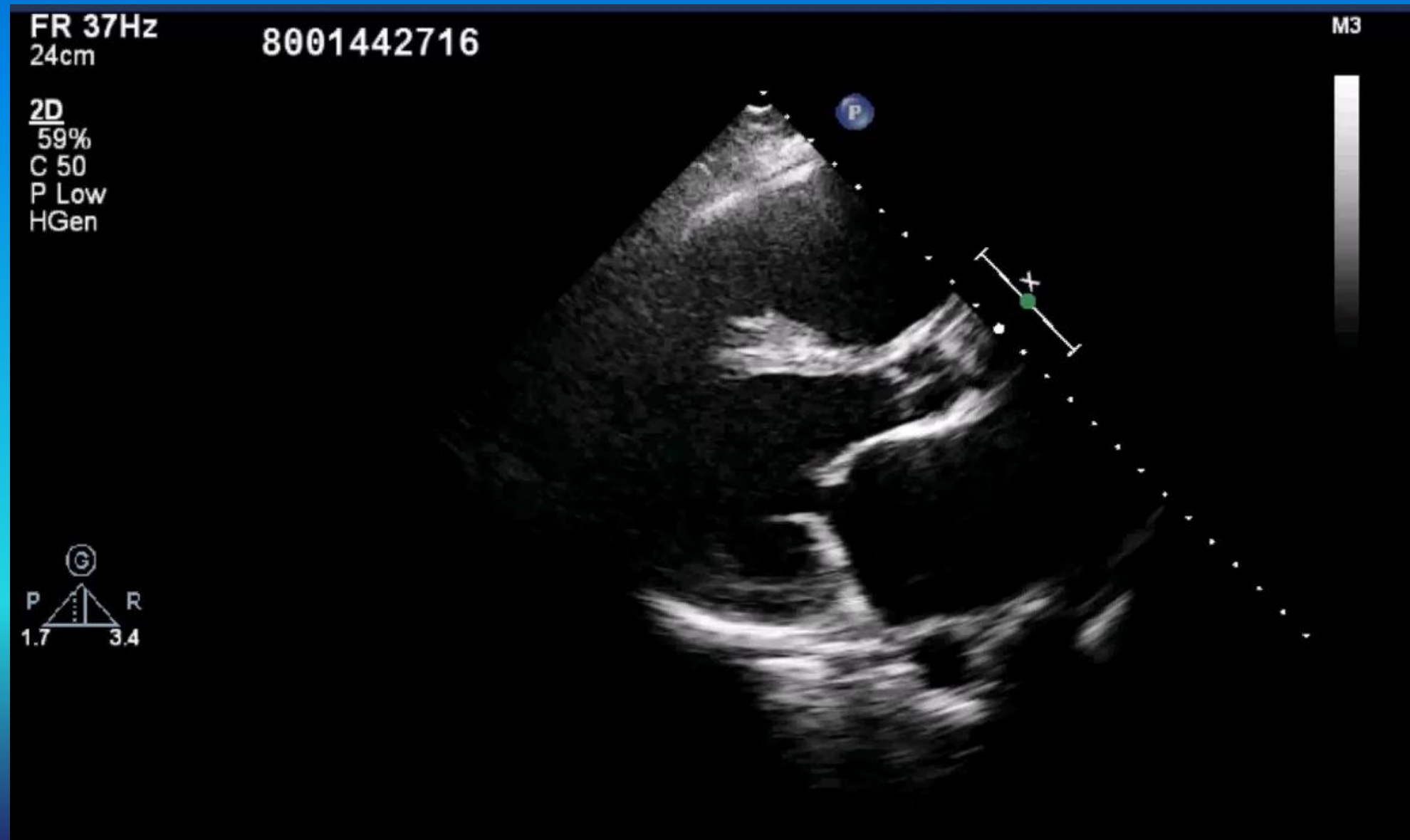




# Case presentation

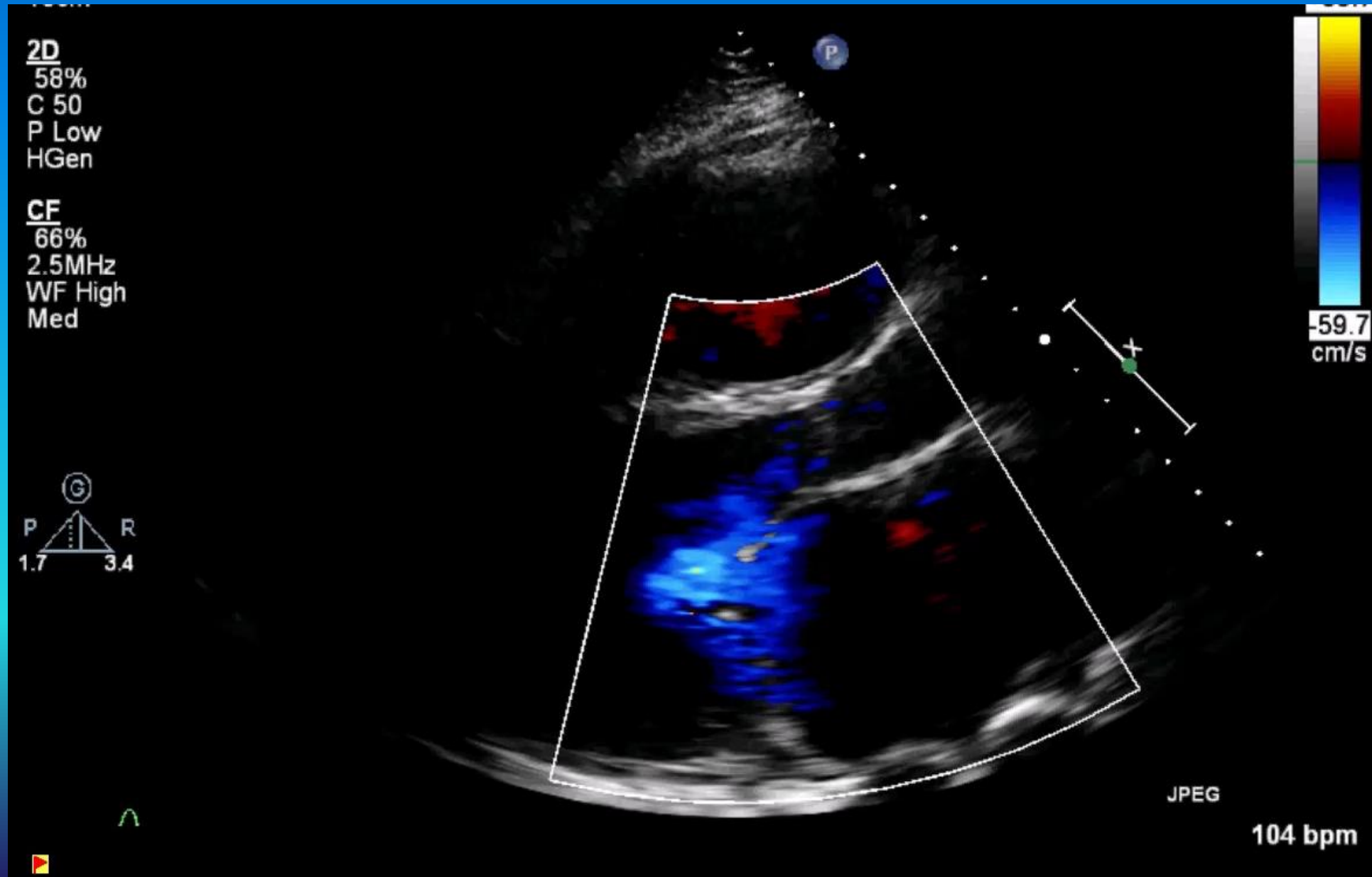
- Medications before transfer:
  - : Sildenafil (20) 2 x 3
  - : Spironolactone (25) 2 x 1
- She was referred to KCMH for proper management on Feb 2020.
- Echo (21/2/2020):
  - : Severe dilated LV, EF = 48%
  - : Severe dilated RV and impaired RV function (TAPSE 1.1 cm, RV FAC 26%)
  - : Severe biatrial enlargement
  - : Sever TR and severe MR.
  - : Pulmonary HT, estimated mean PAP = 38 mmHg

# Echocardiography: 21/2/2020



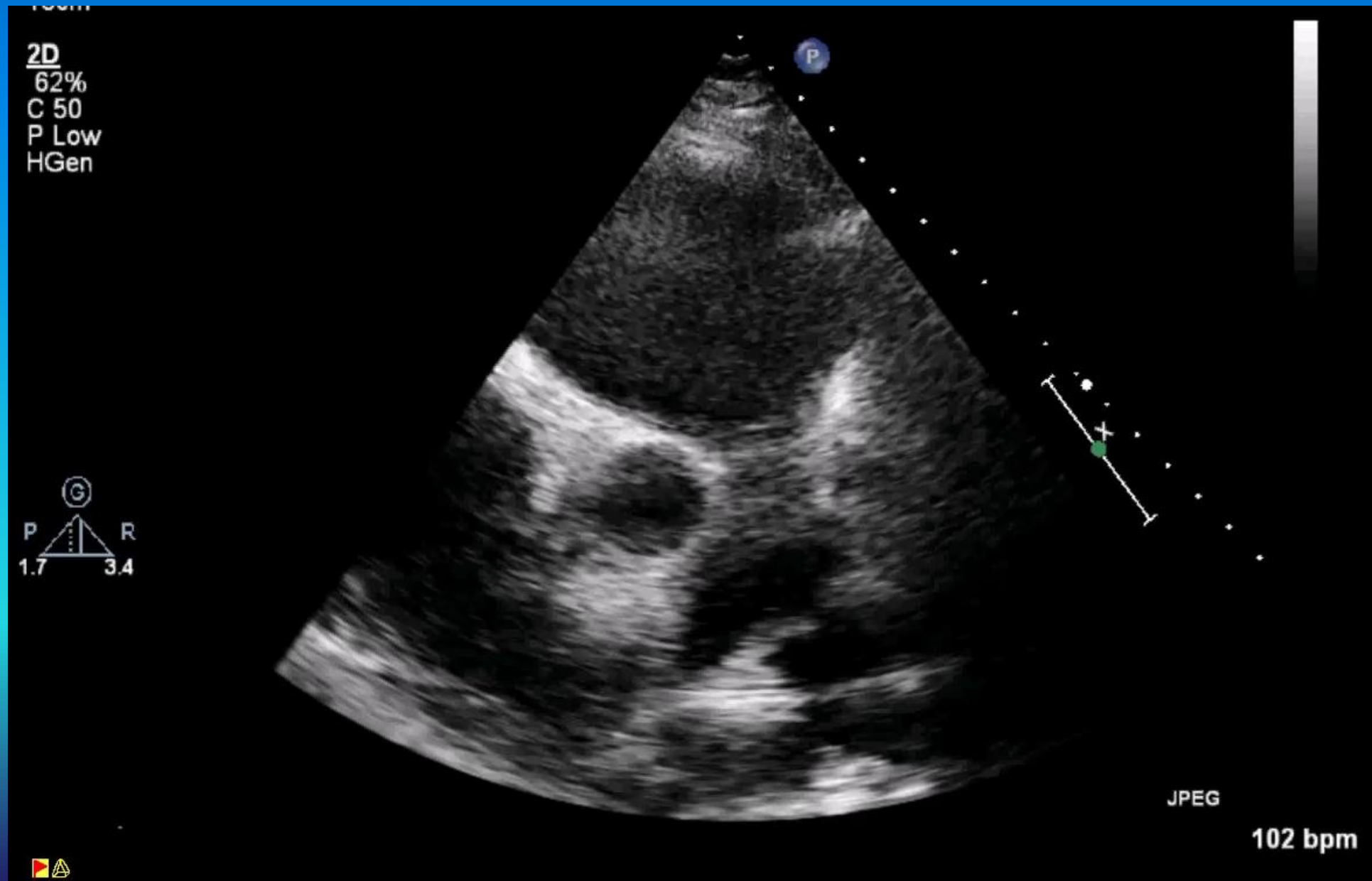


# Echocardiography: 21/2/2020

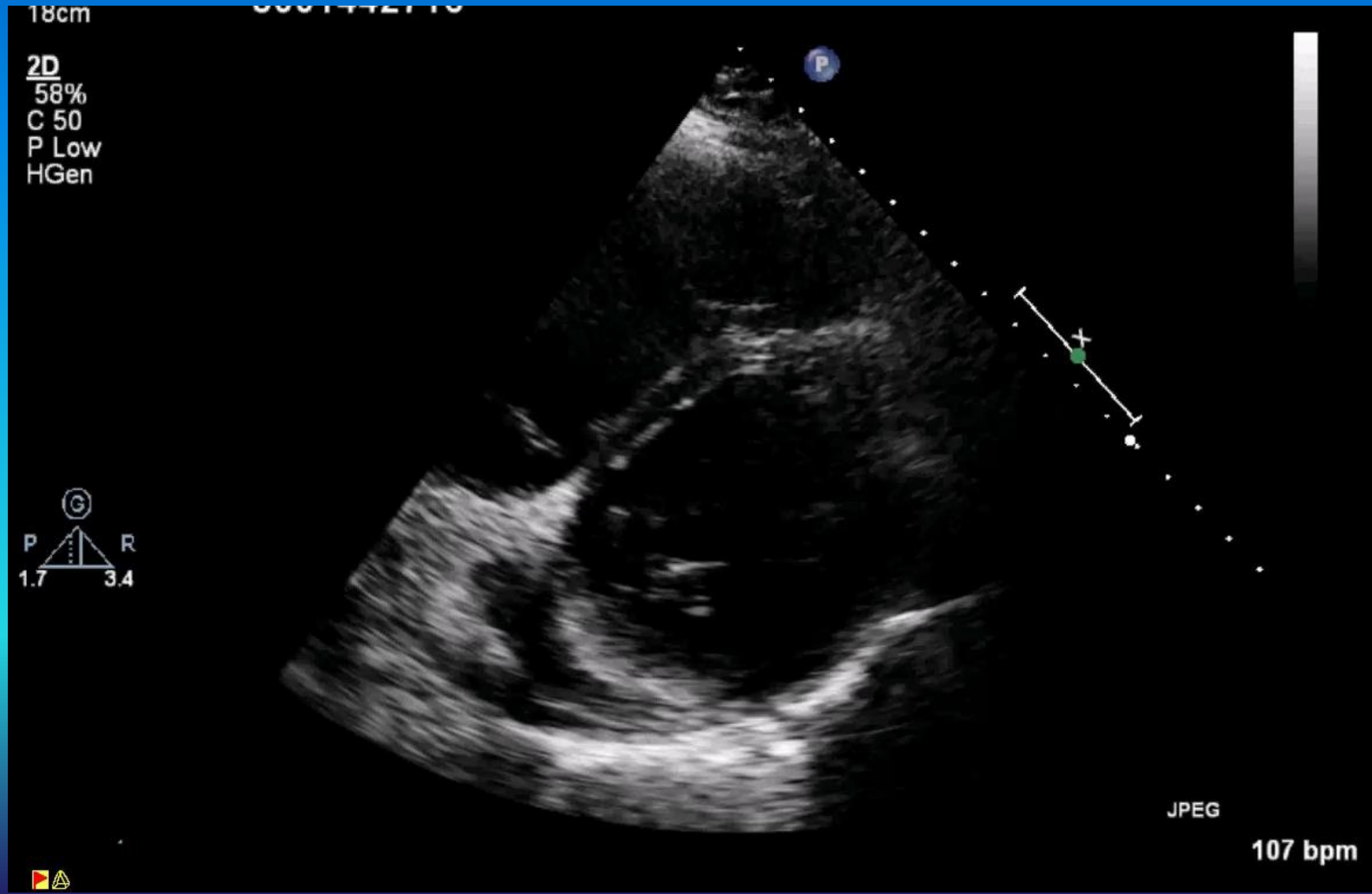




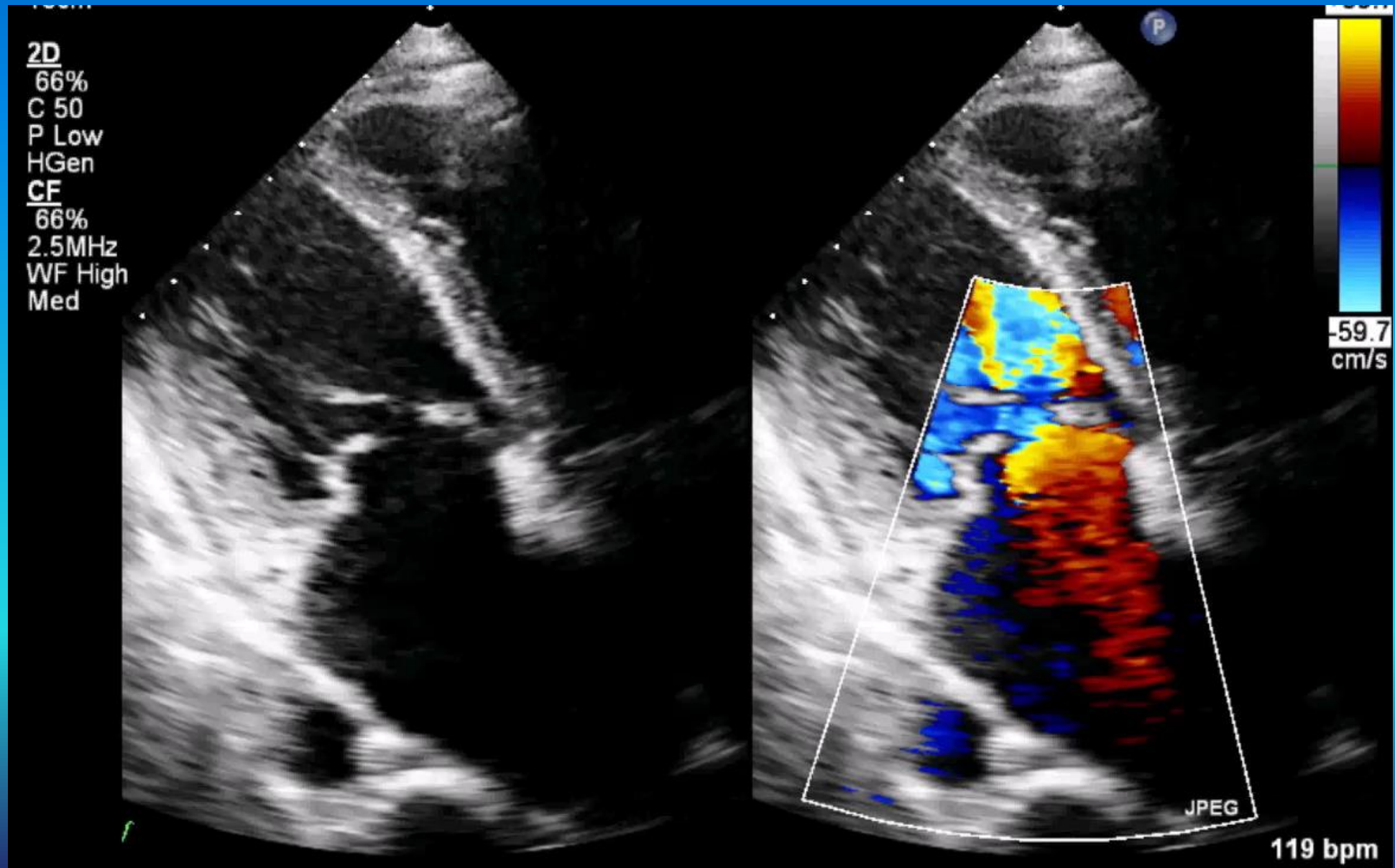
# Echocardiography: 21/2/2020



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FR 45Hz

18cm

2D

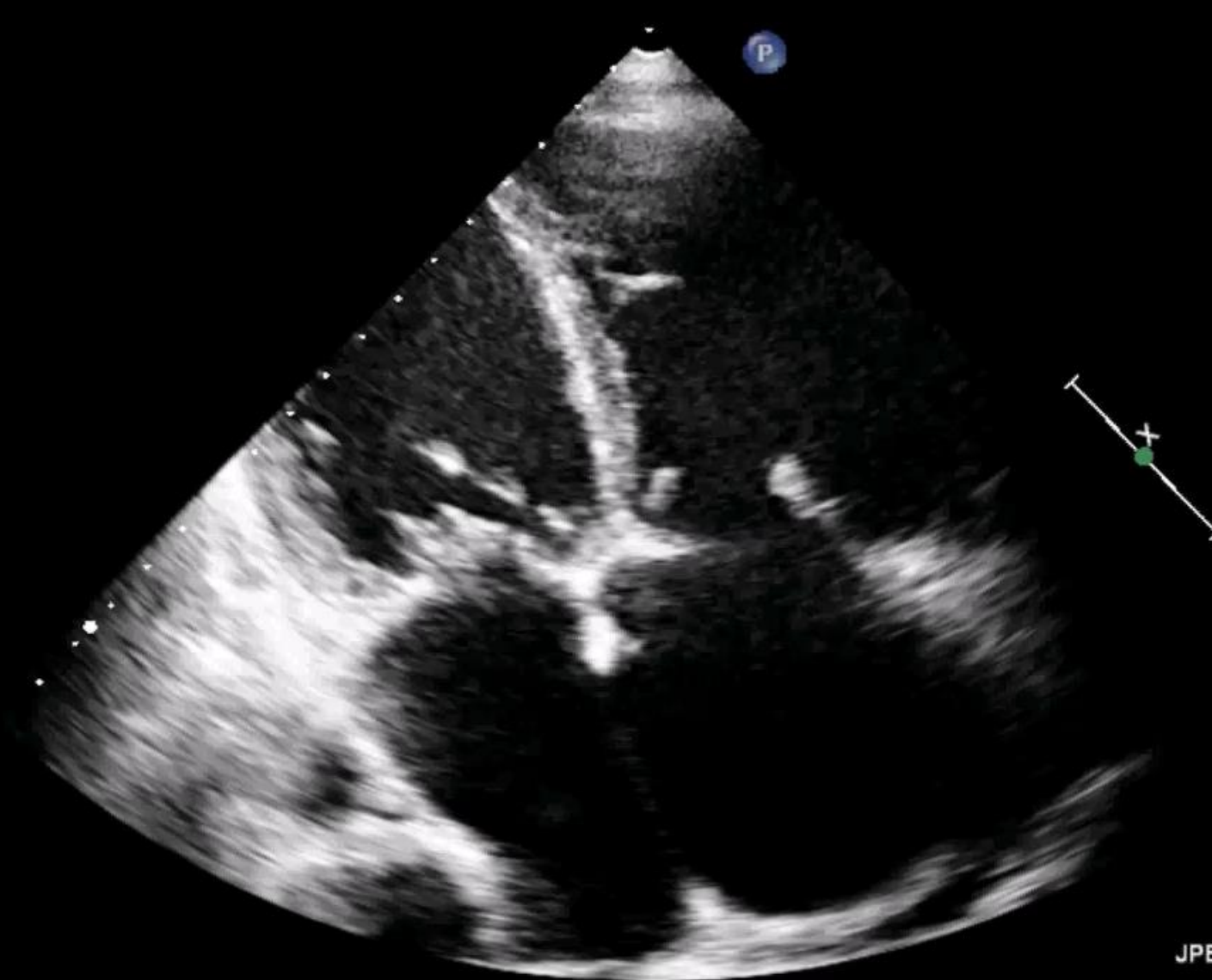
67%

C 50

P Low

HGen

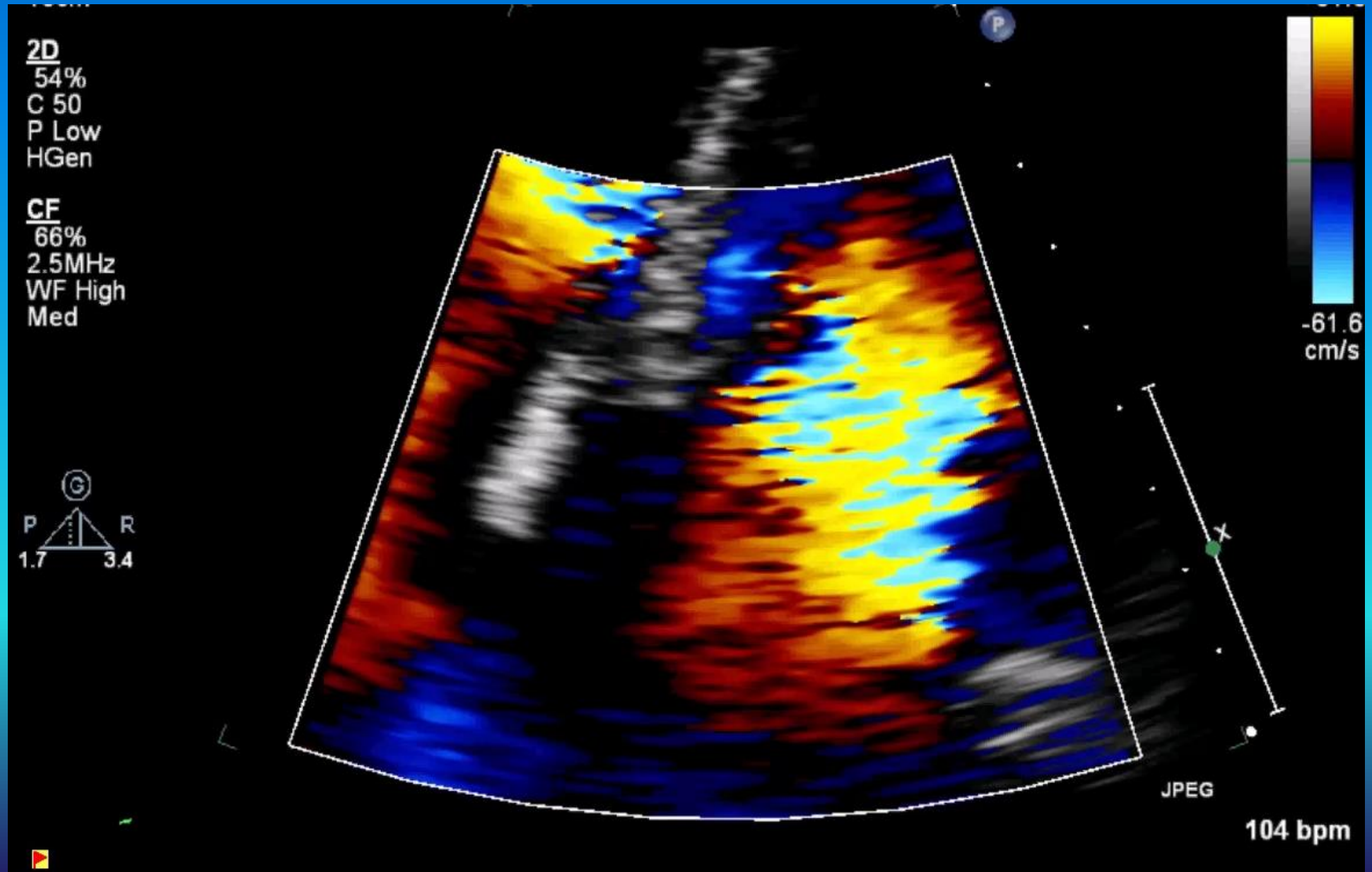
M3



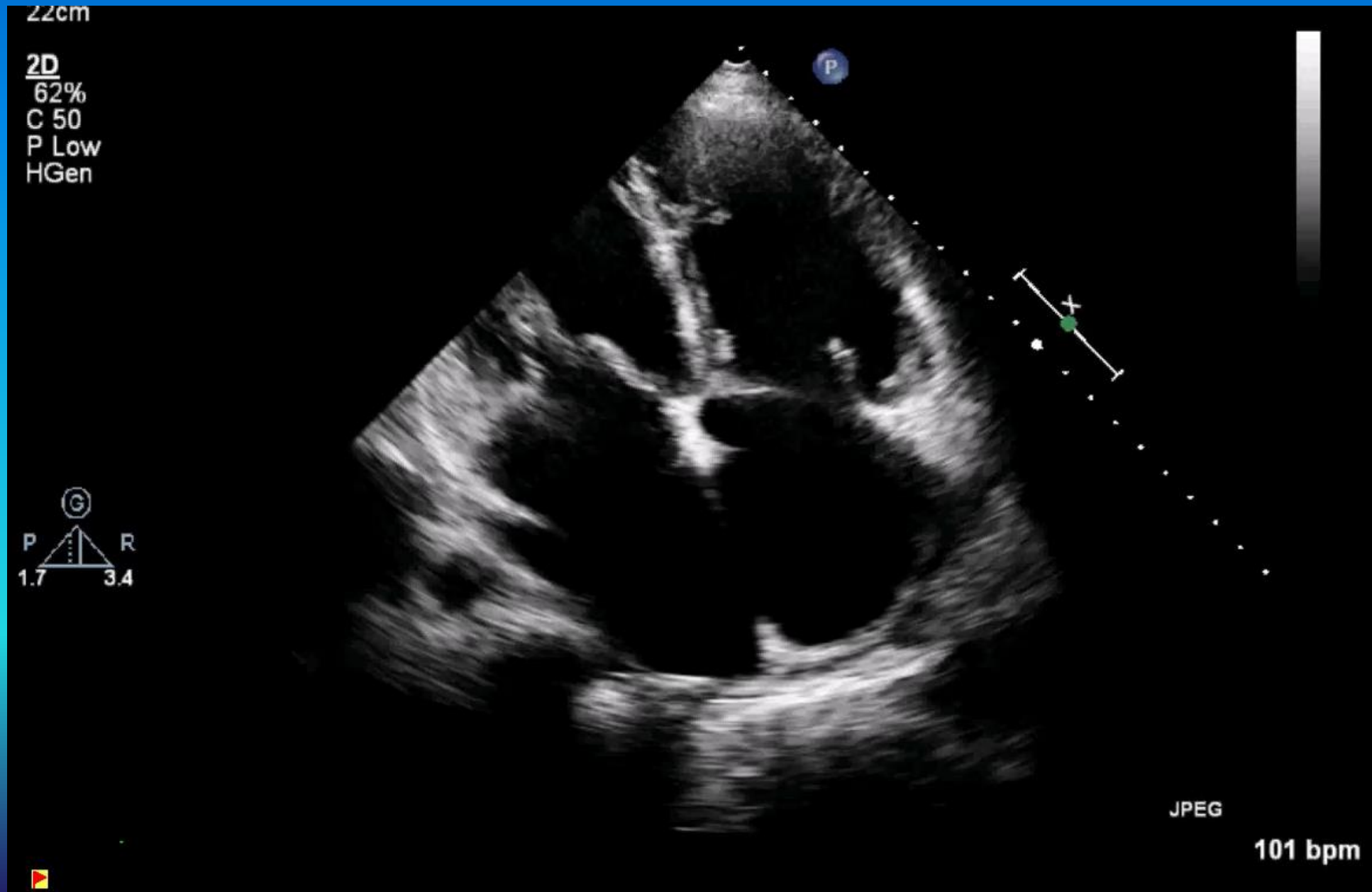
JPEG

108 bpm

# Echocardiography: 21/2/2020



# Echocardiography: 21/2/2020





# Case presentation

- During admission on Feb 2020 at KCMH, she was diagnosed of pulmonary HT due to left sided heart disease. She was discharged on 26/2/2020 with
  - : Furosemide (40 mg) 1-1-0
  - : Metoprolol (100 mg) 0.5 x 2
  - : Warfarin according to INR



# Case presentation

After D/C, her symptom of dyspnea was not improved and returned back on 11/3/2020.

## On Physical examination:

- Good consciousness, mild dyspnea
- BP – 178/101 mmHg, HR – 80/m irreg, RR – 22 /m
- Not pale, anicteric sclera, mild puffy eyelid
- JVP up to mandible
- Heart: LV heave with systolic ejection murmur and diastolic murmur at LSB. Loud P2
- Lung: crepitation both lower lung field
- Liver 3 FB below right costal margin
- Edema 3+



# Case presentation

## Lab Investigation:

- Cr – 1.49 mg/dl                      BUN – 21 mg/dl
- Alb – 2.8 g/dl
- Total bilirubin 5.0 mg/dl (direct 3.37 mg/dl)
- Na – 135 mmol/l, K – 2.4 , Cl – 95, Co2 – 30
- Hb – 10.8 g/dl
- INR – 2.54

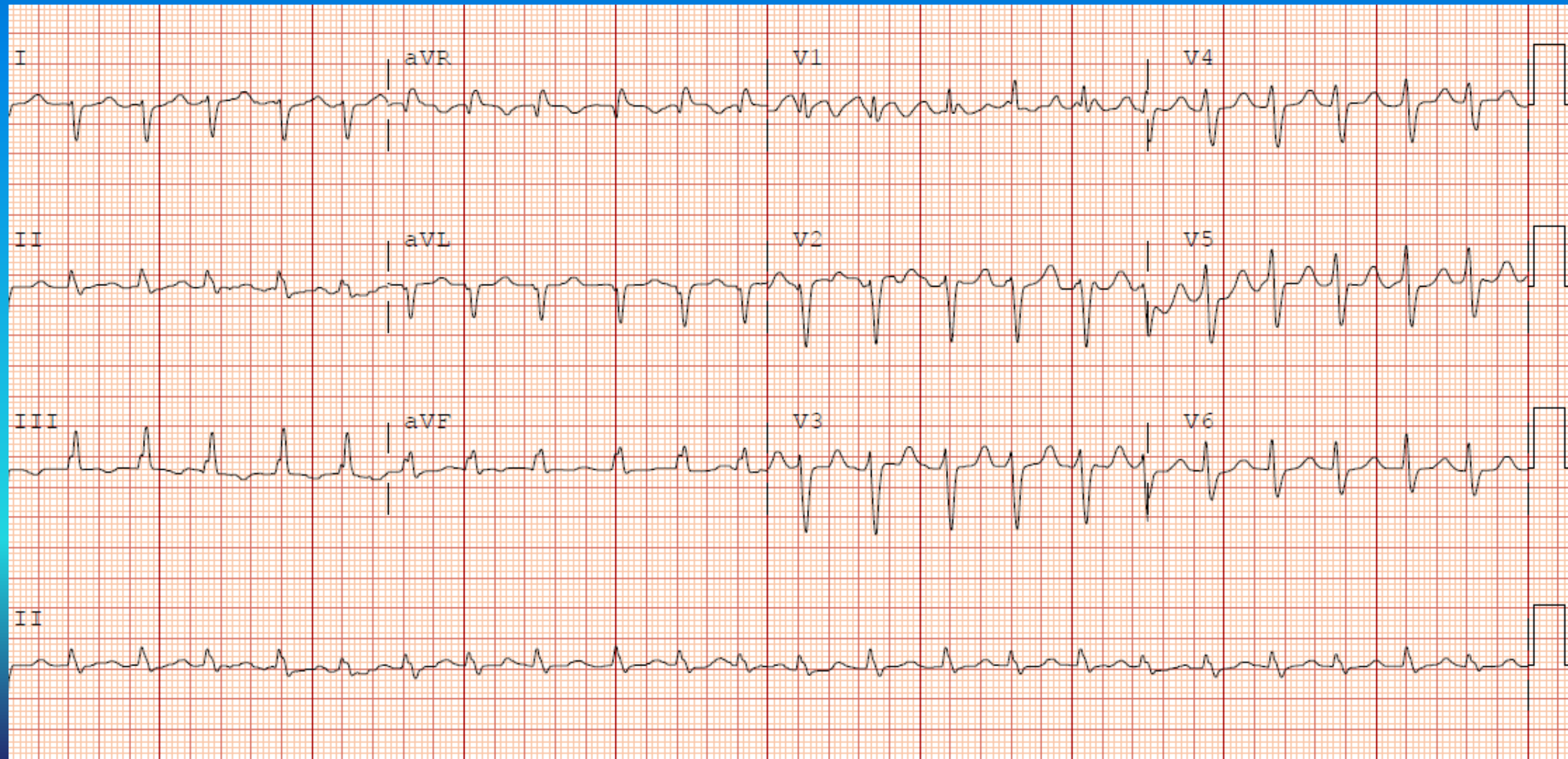
## Initial management:

- Furosemide 80 mg IV
- After diuretic Rx – urine output 100 ml / 8 h

# Case presentation

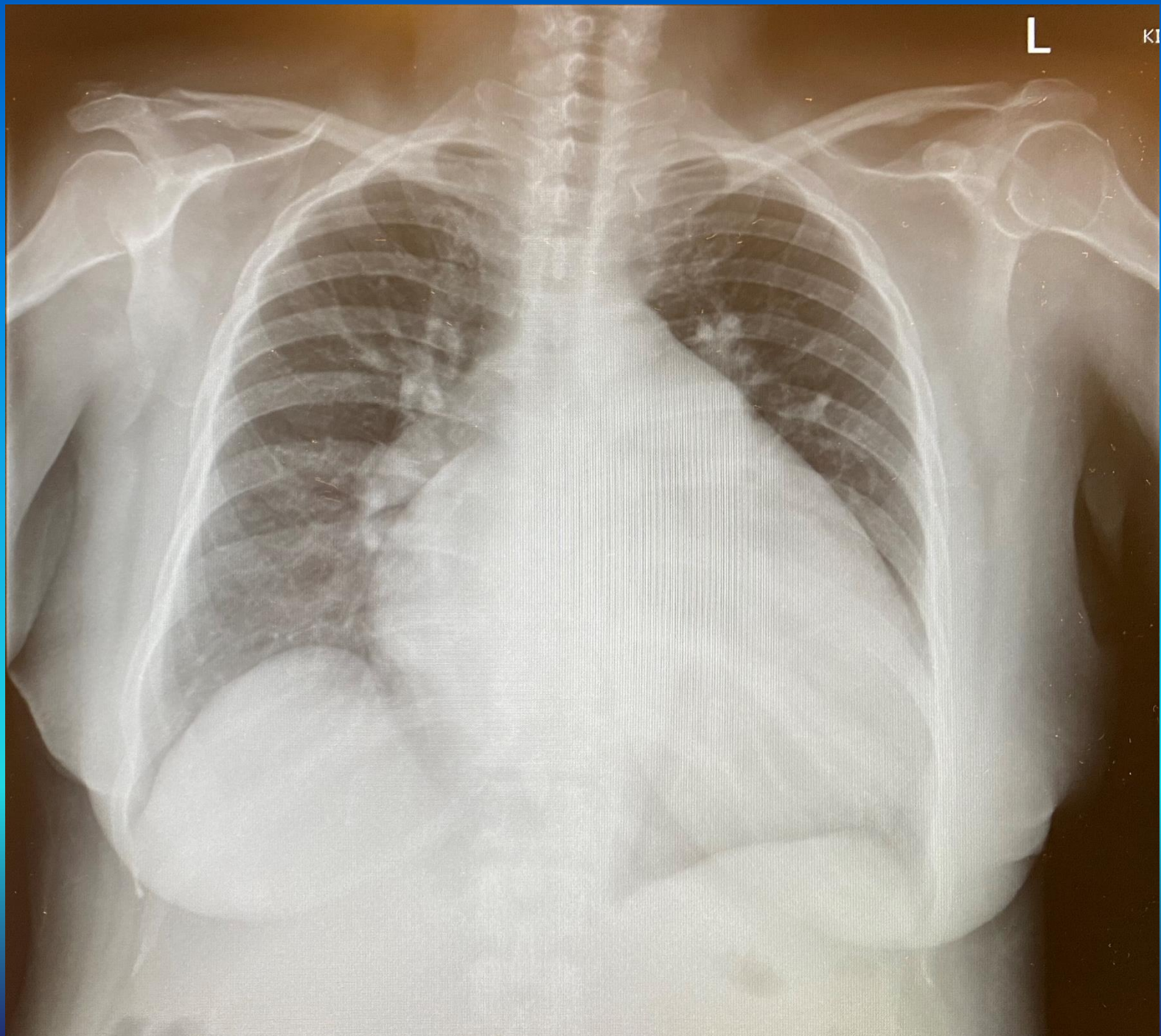
- She was notified because of markedly dyspnea.
- Her BP – cannot measure, pulse – low volume
- ABG – pH – 7.507, PaO<sub>2</sub> – 73.7, PaCo<sub>2</sub> – 25.2, lactate – 5.9
- She was transferred to CCU for close monitor.

# Case presentation

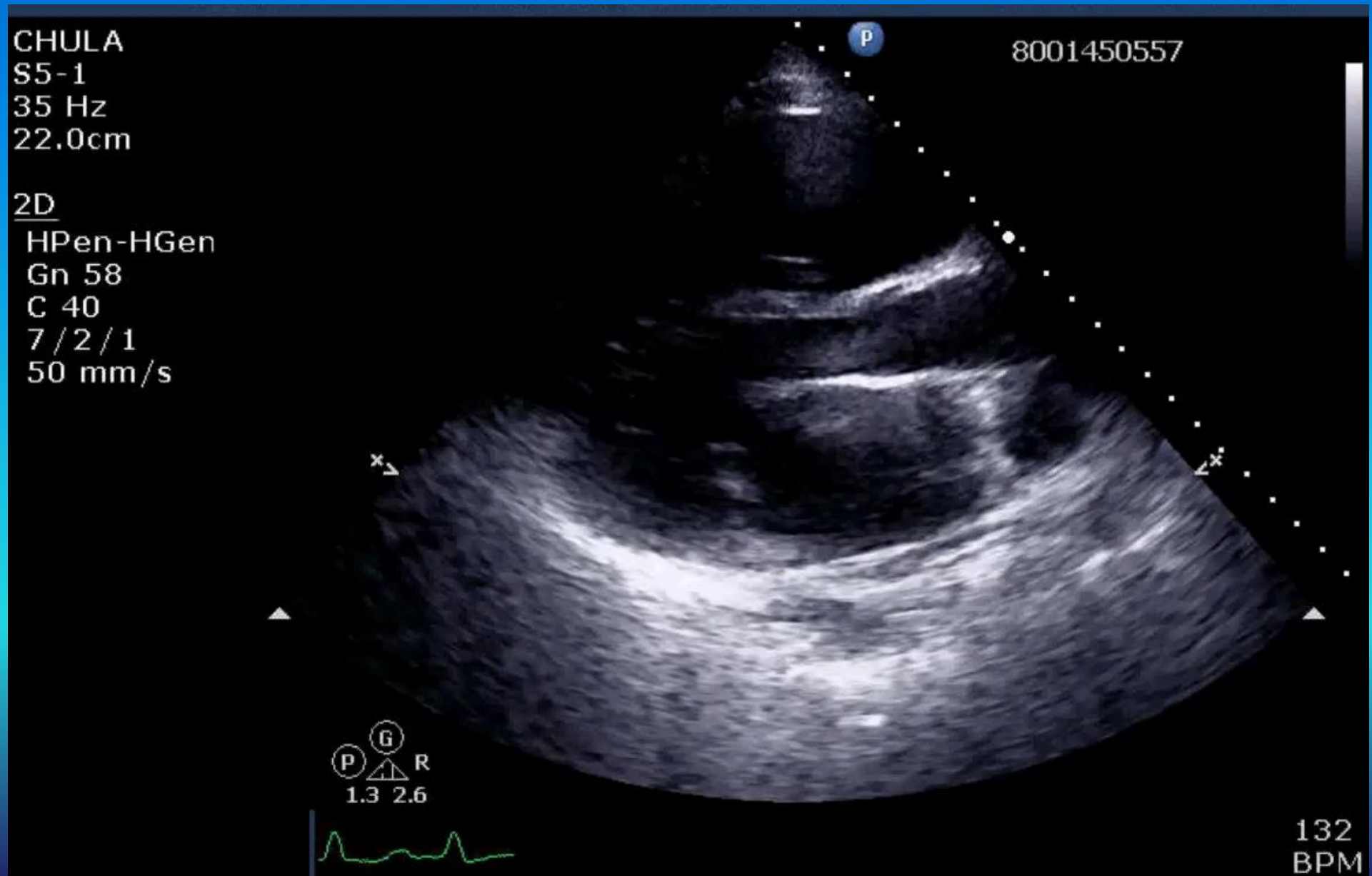




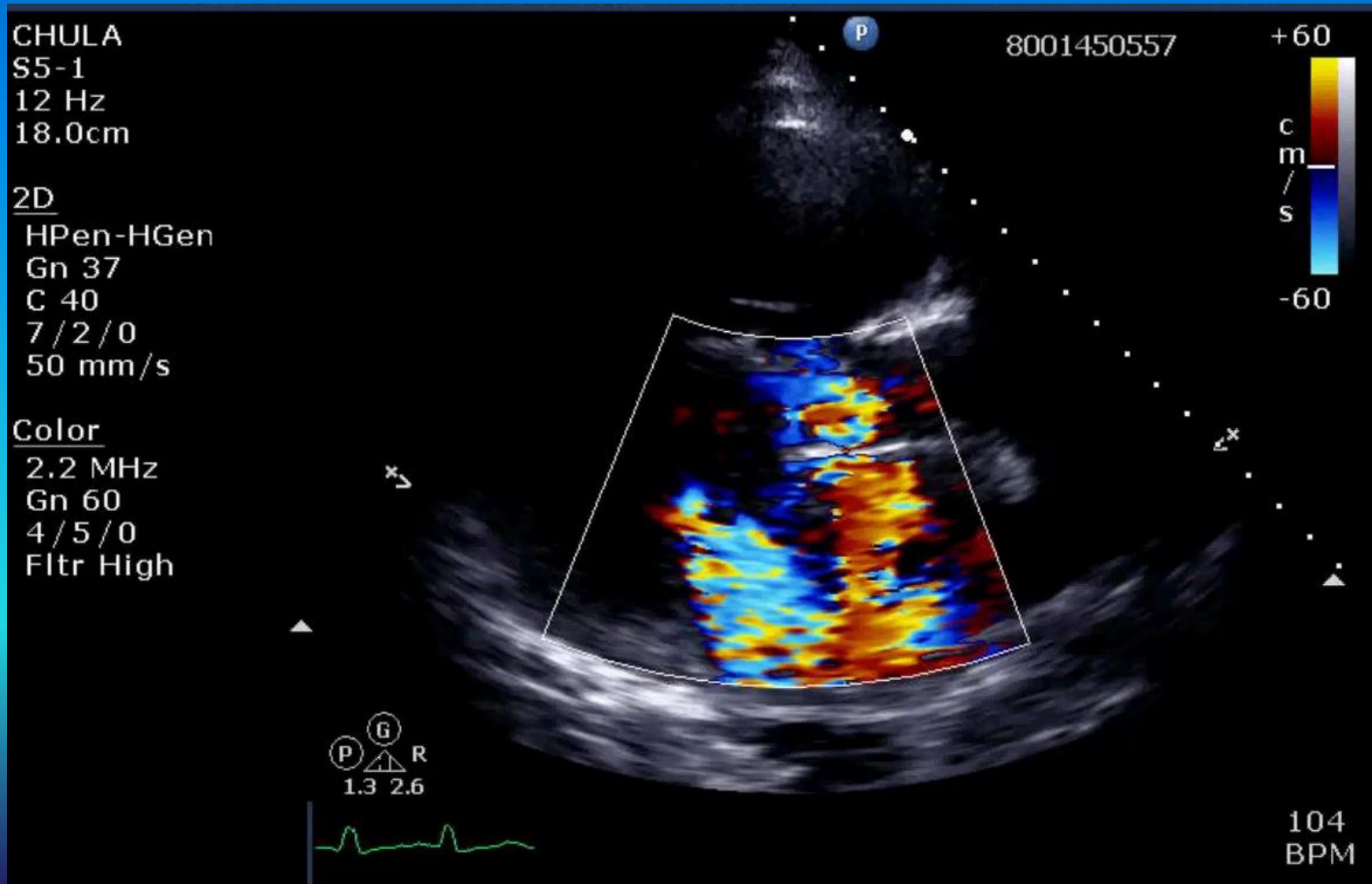
**CXR**



# Echocardiography: D1 – 11<sup>th</sup> Mar 2020

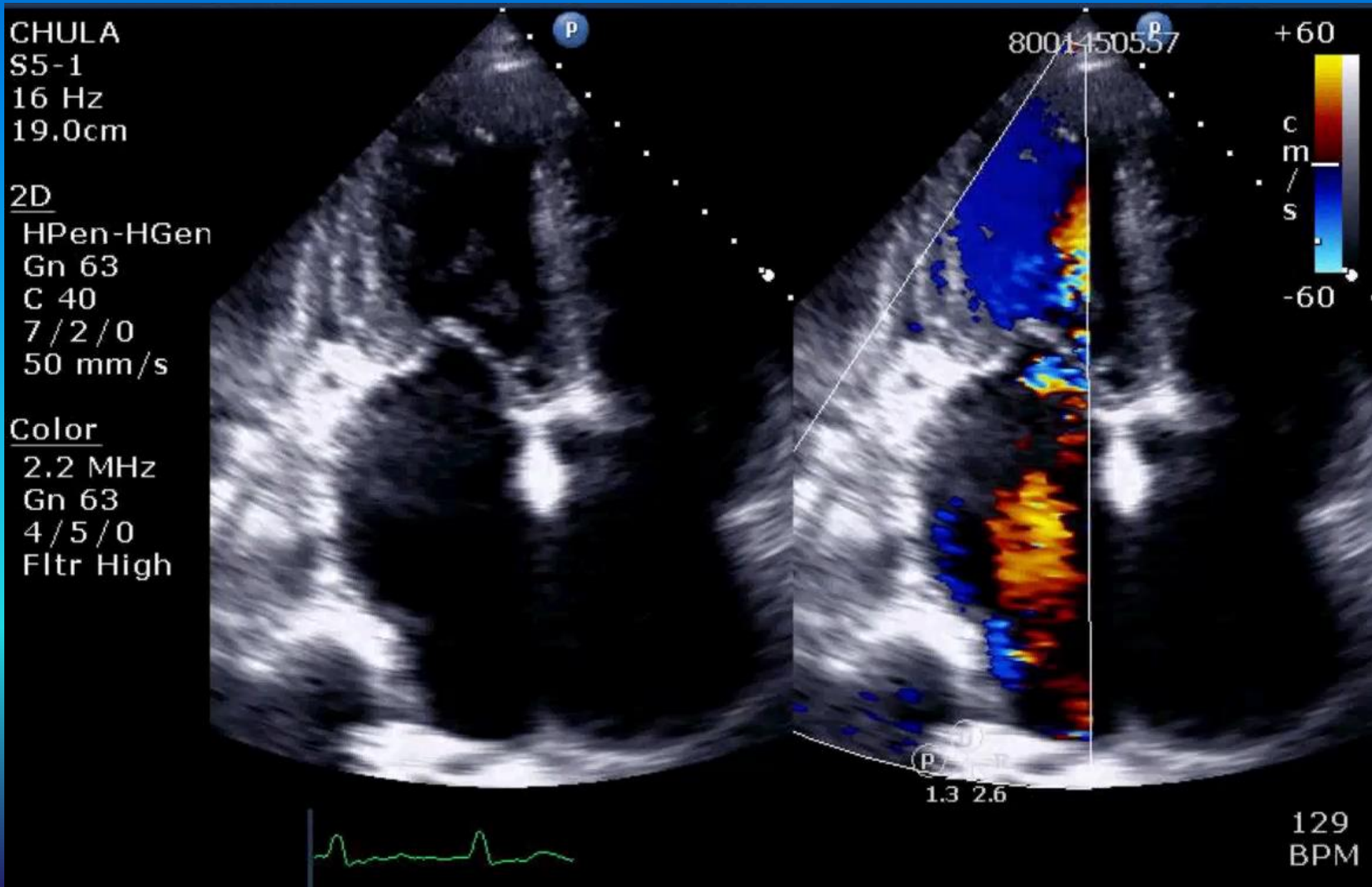


# Echocardiography: D1 – 11<sup>th</sup> Mar 2020

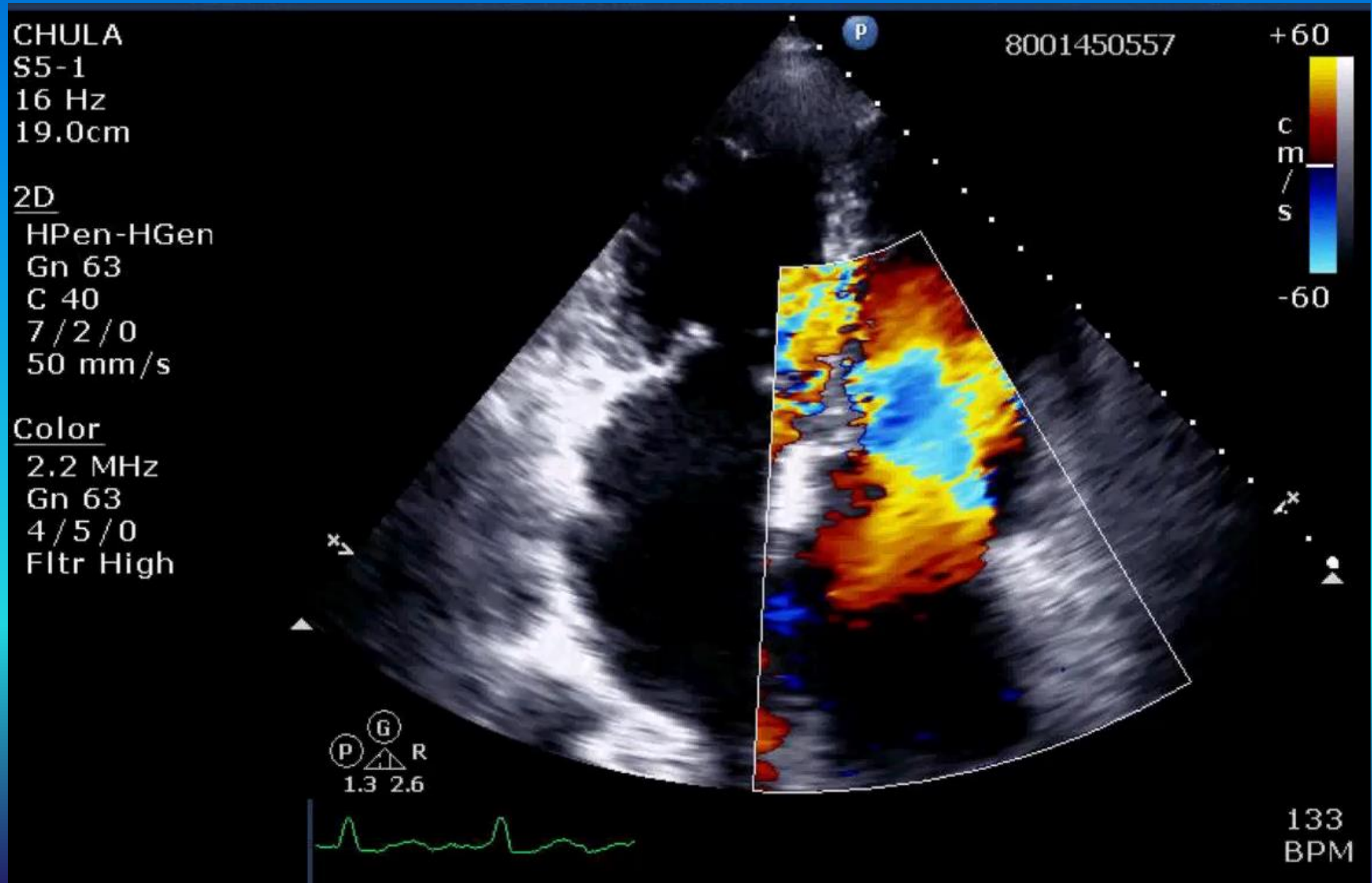




# Echocardiography: D1 – 11<sup>th</sup> Mar 2020



# Echocardiography: D1 – 11<sup>th</sup> Mar 2020



# Case presentation

- How to management?
  - a. More aggressive diuresis
  - b. Start Inotropic – Dobutamin / Millinone
  - c. Amiodarone to control AF
  - d. Withdraw Metoprolol
  - e. IABP
  - f. ECMO
  - g. Emergency MV repair + TV repair

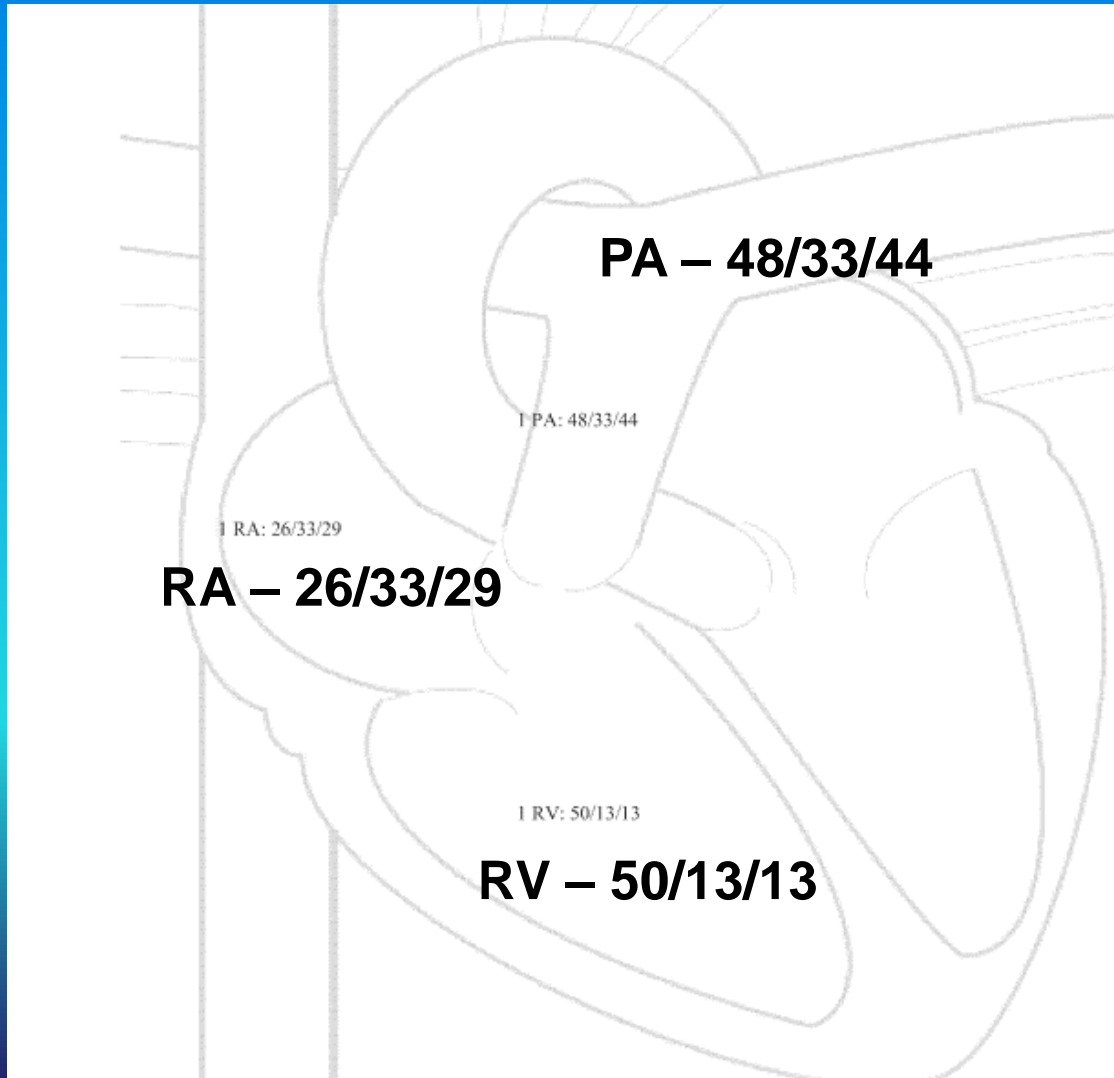


# Case presentation

- At CCU: BP - 82/52/61 mmHg
- Furosemide 120 mg IV then 20 mg /h
- Amiodarone 150 mg iv in 30 min
- She was sent to Cath lab for Swan Ganz catheter and IABP insertion.

# Case presentation

- Cath data:



Fick CO – 7.58

SVR – 369 dyne.sec/cm-5

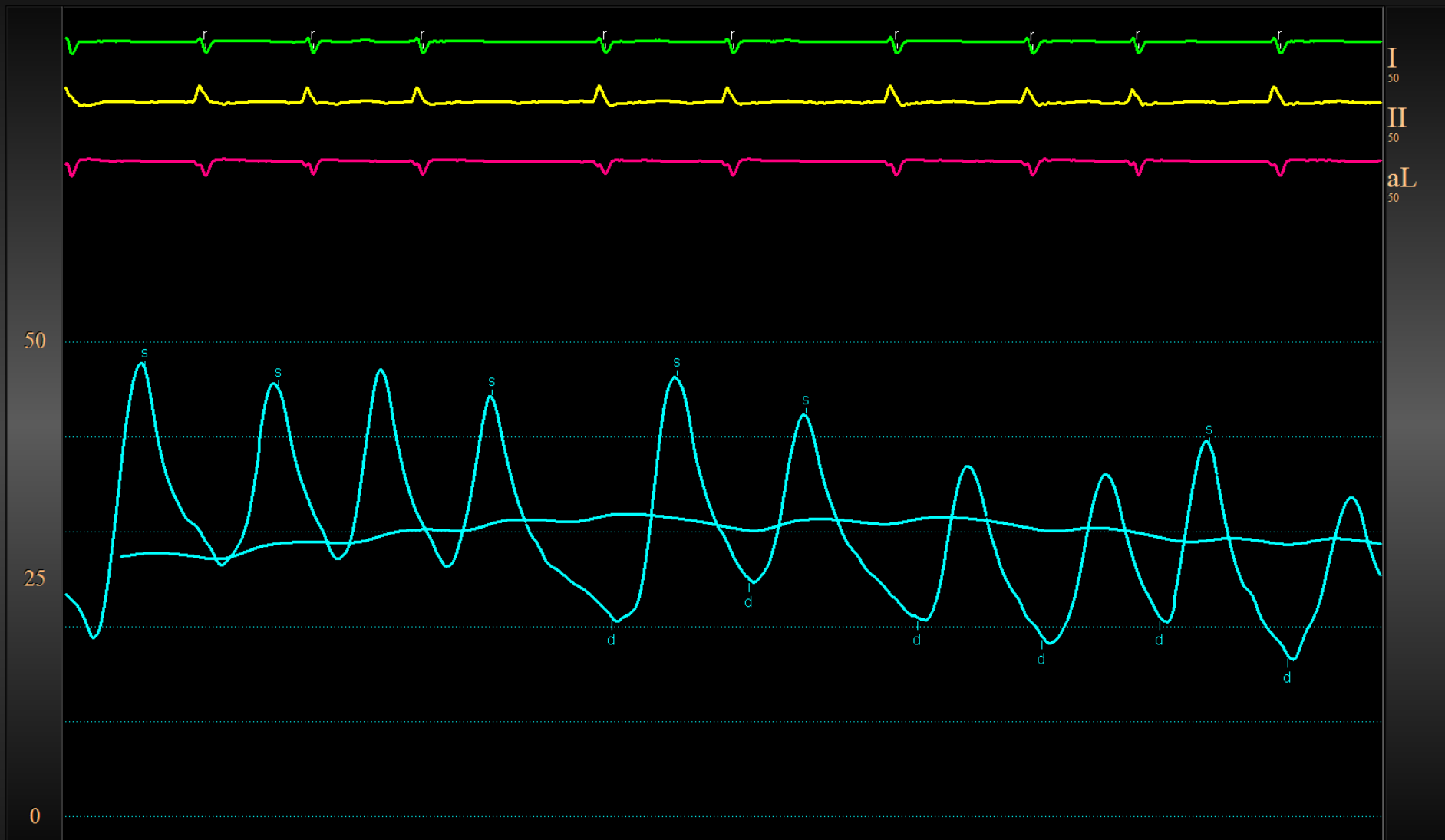
PVR – 1.85 wood unit

TPG/DPG – 14/3

RVAWi – 5.45 g/m2/beat

PA 43 /19 (29 )<sup>1</sup>

103<sup>M</sup>

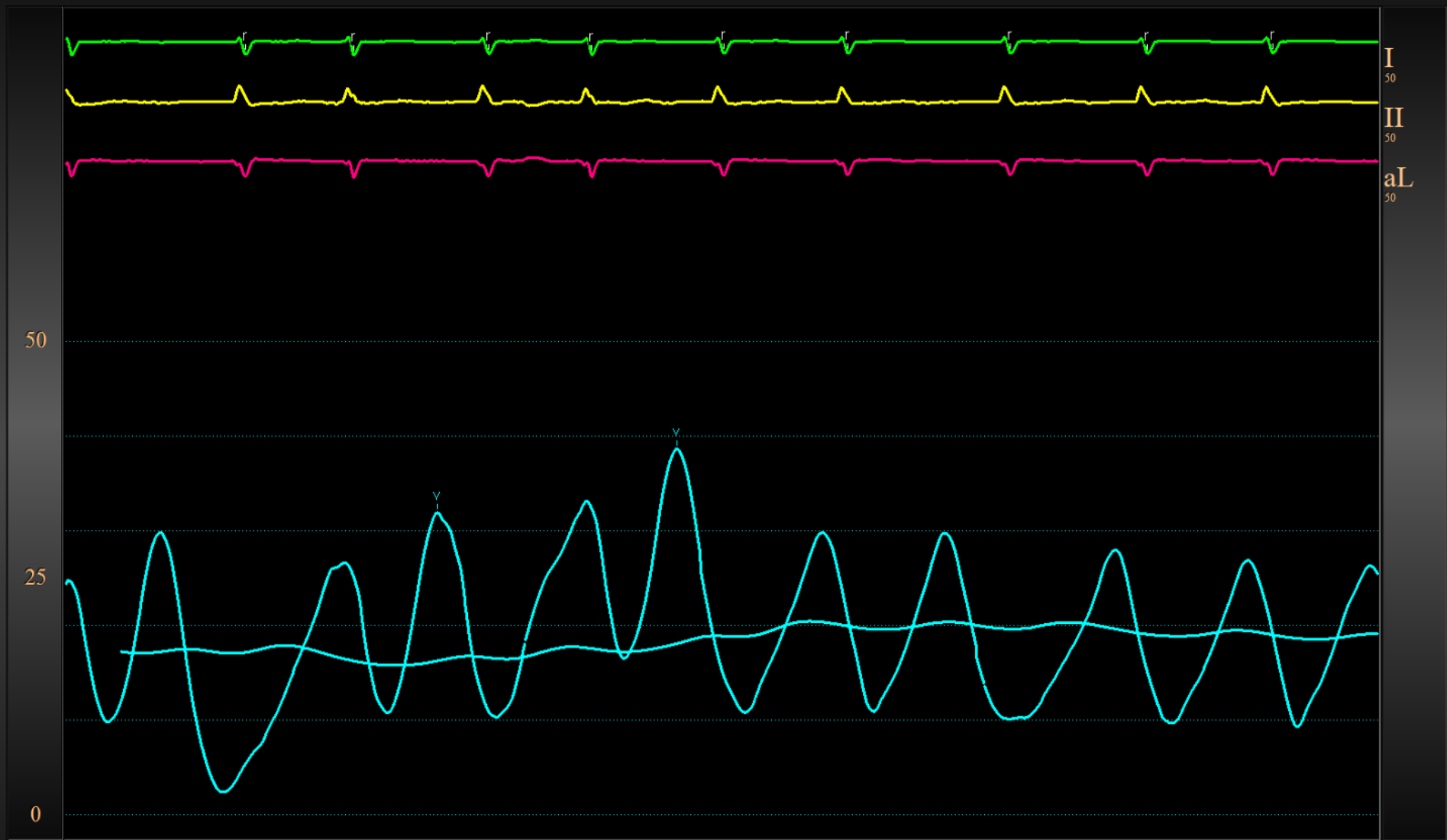


98 %<sup>103</sup>



PW -99/34 (22 )<sup>1</sup>

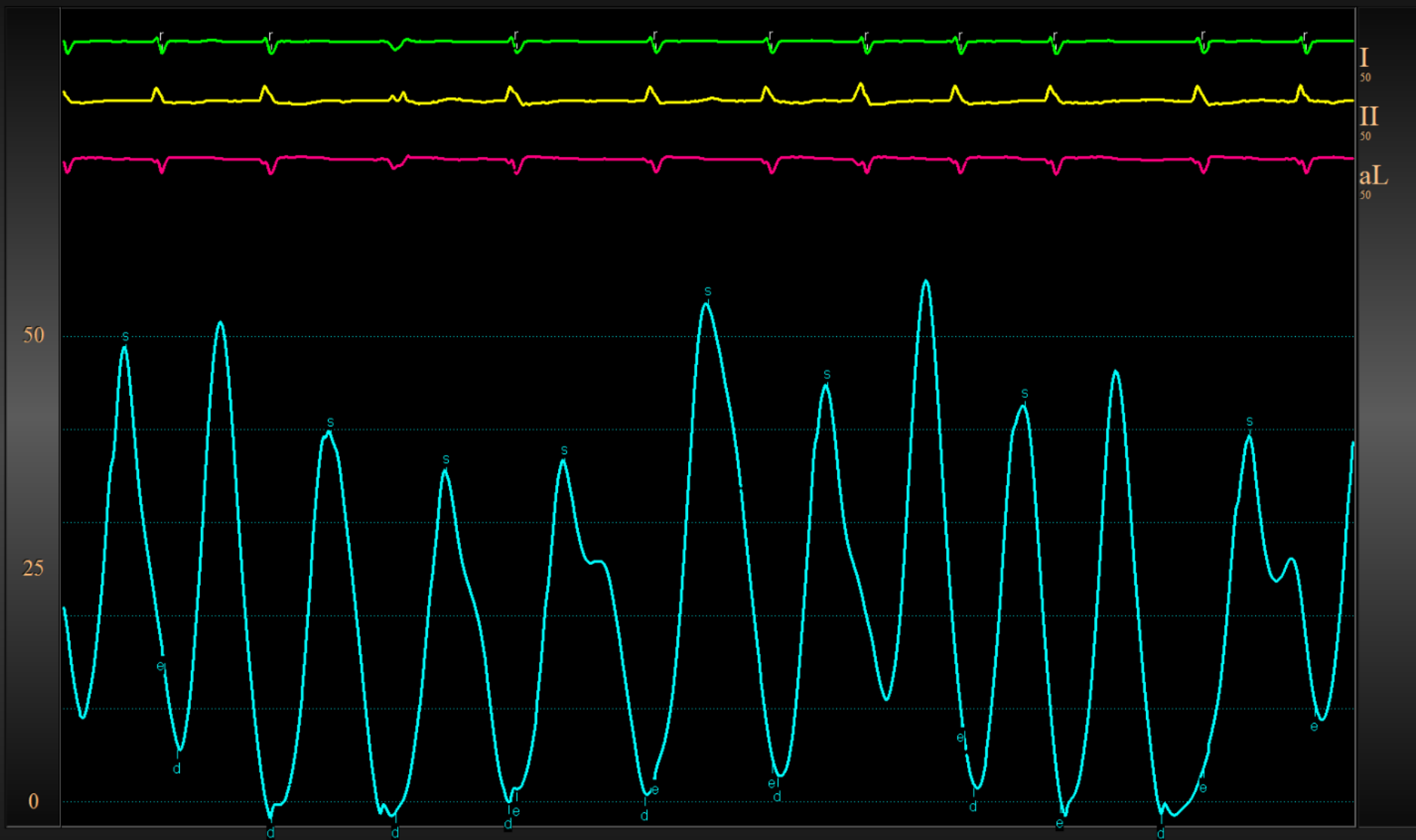
106<sup>M</sup>



0 %<sup>0</sup>

RV 43 / 0 6 1

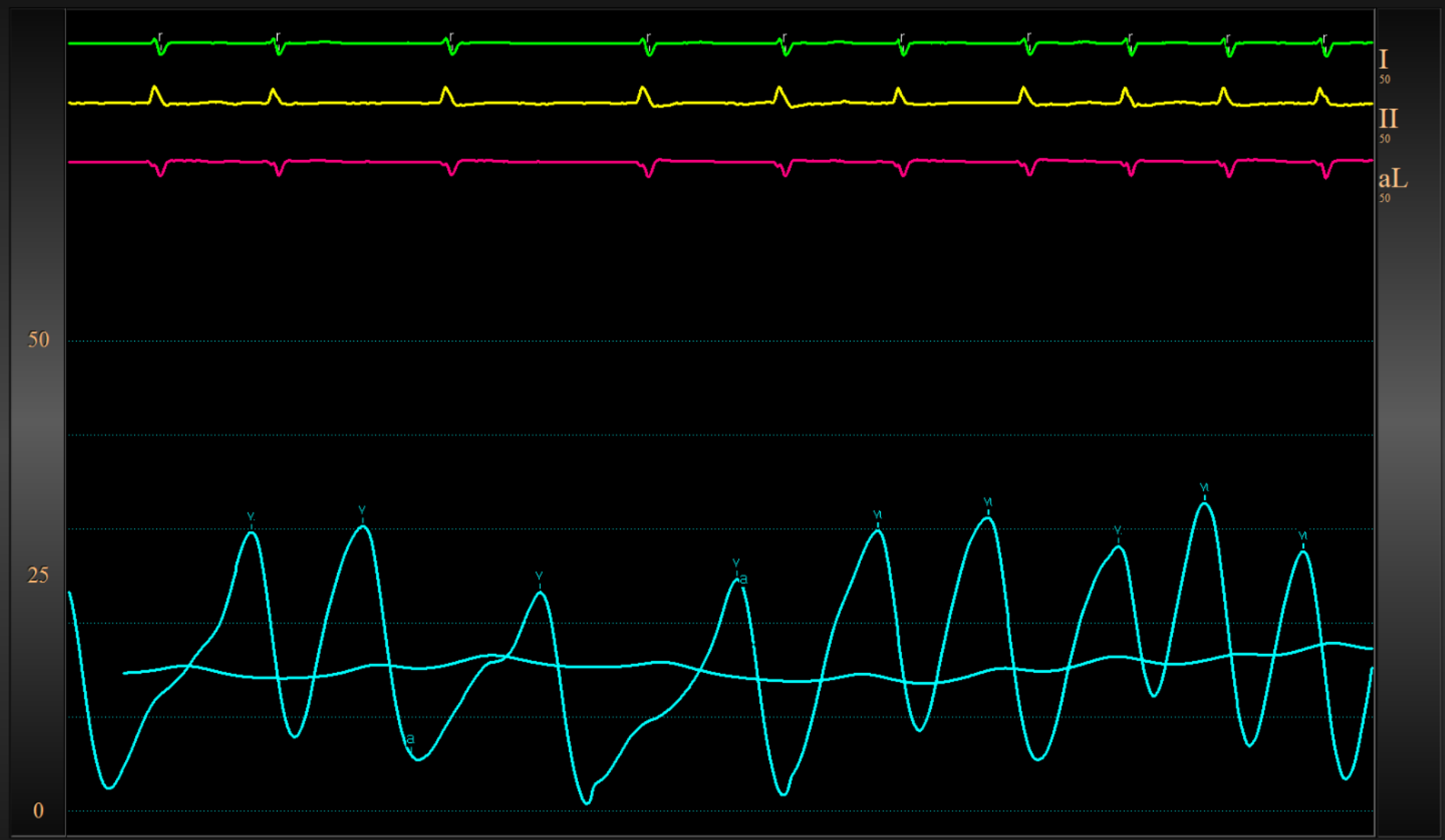
101<sup>M</sup>



0 %<sup>0</sup>

RA 22 / 27 (15)

102<sup>M</sup>

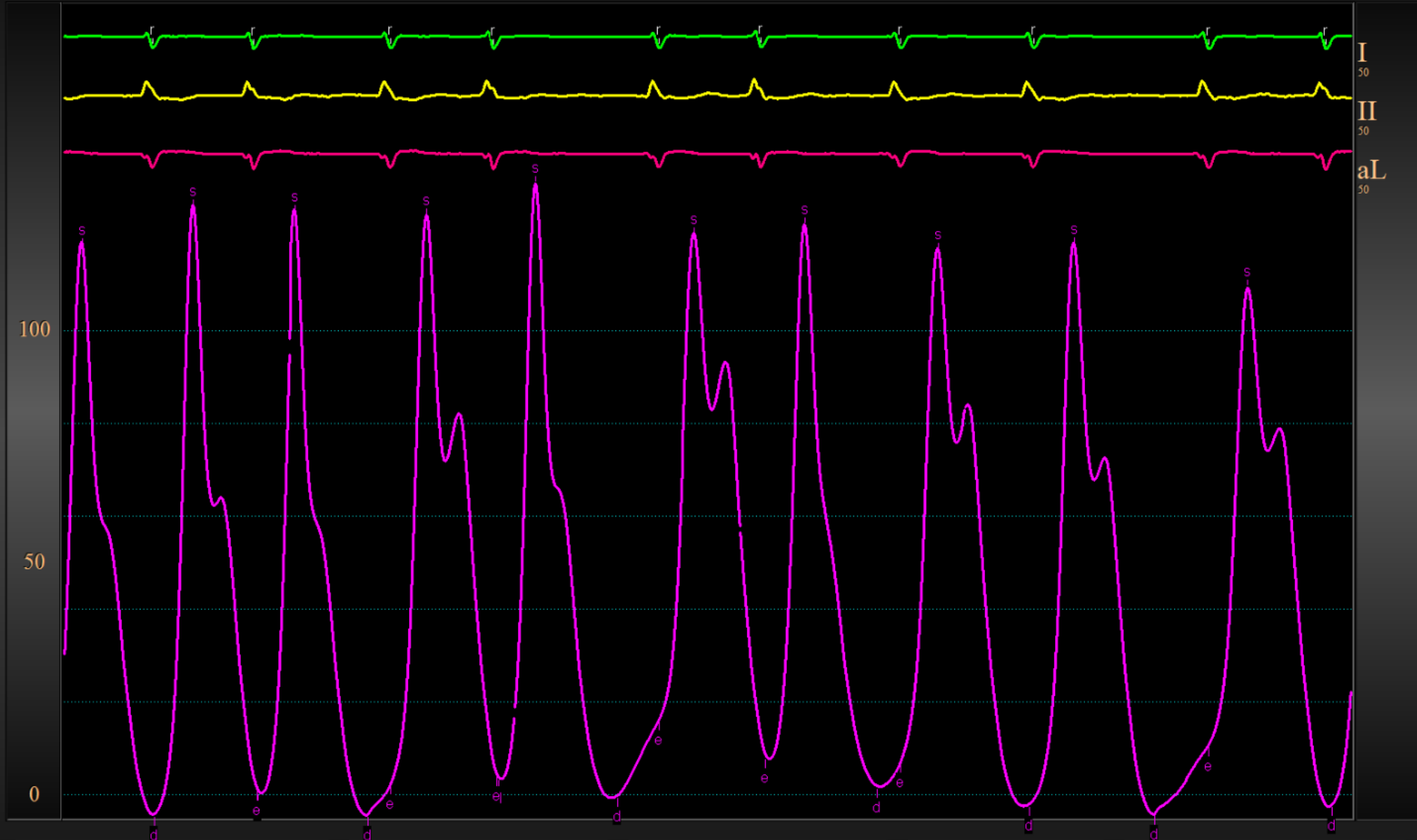


97 %<sup>91</sup>



LV 120/0 4 1

103<sup>M</sup>



97 %<sup>99</sup>

# Case presentation

- After return back from Cath lab., no urine output and patient was getting worse with increase of lactate level.

# Case presentation

- How to management?
  - a. More aggressive diuresis
  - b. Start Inotropic – Dobutamin / Millinone
  - c. IABP
  - d. ECMO
  - e. Emergency MV repair + TV repair
  - f. Emergency percutaneous MV repair
  - g. Suggest end-of-life care

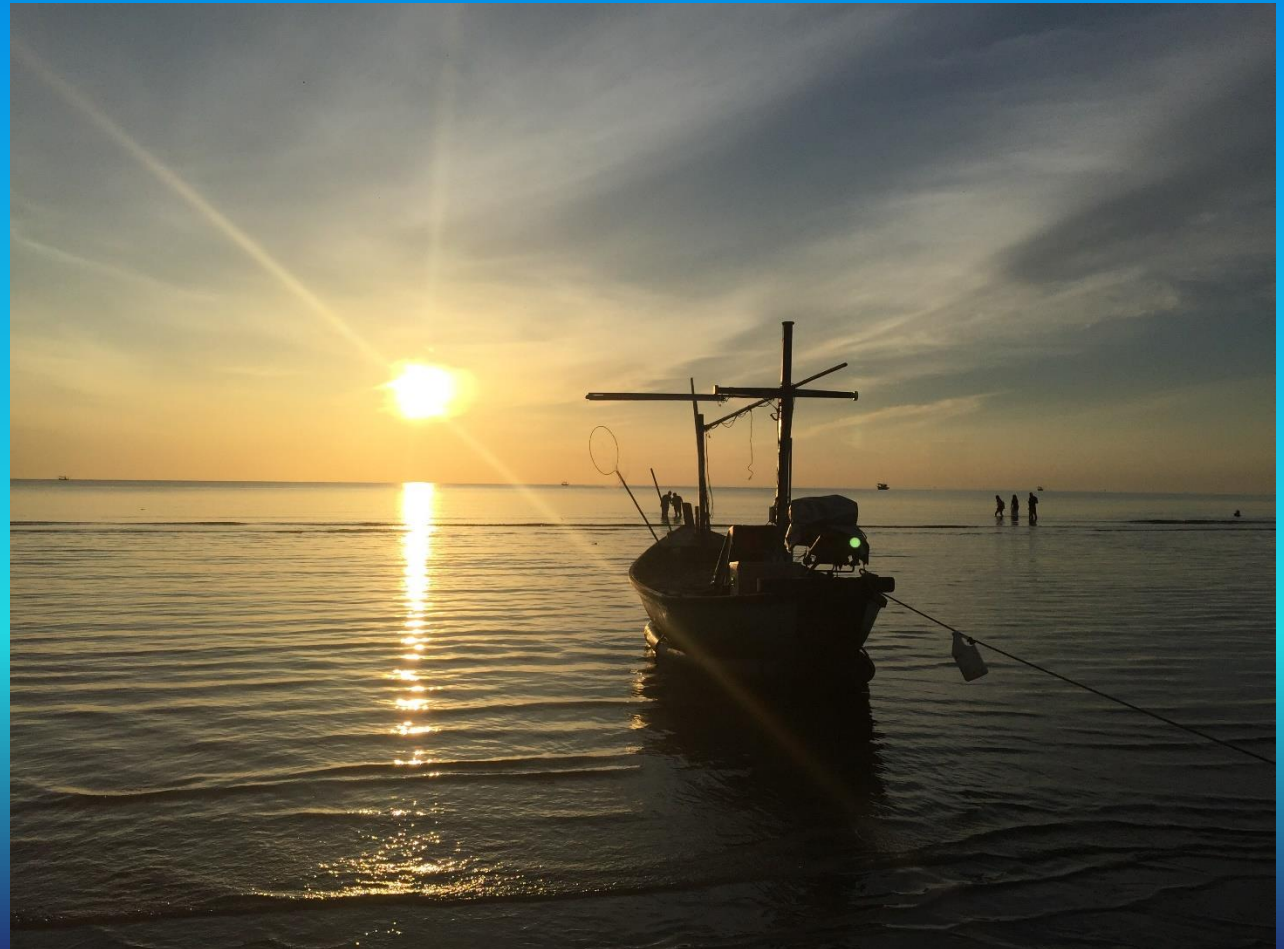
# Case presentation

- VA - ECMO was started.
- After ECMO, she was stable and urine output was 200-300 ml/h (no need for CRRT) and lactate level slightly improved.



# Case presentation

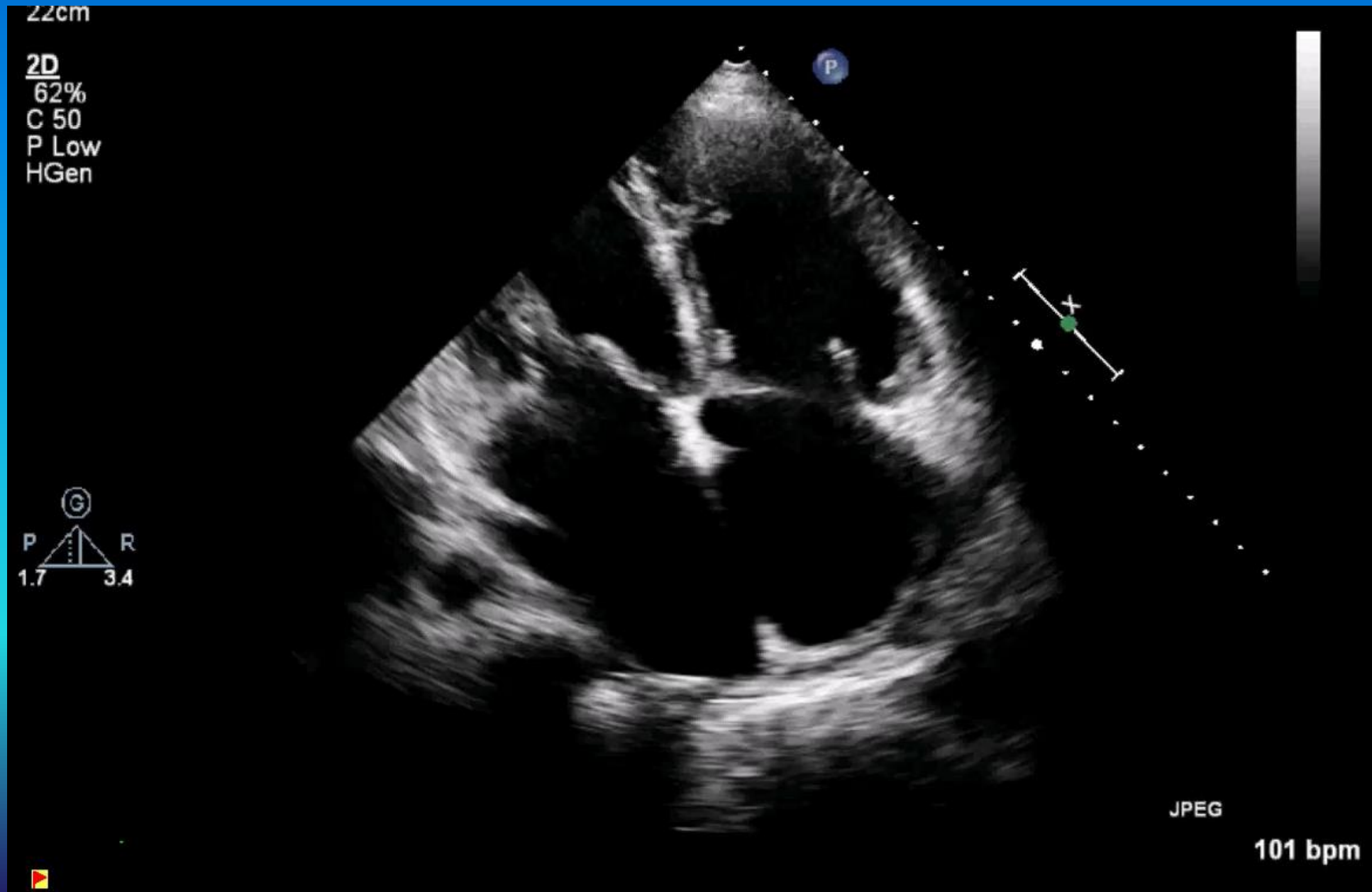
- What's next?



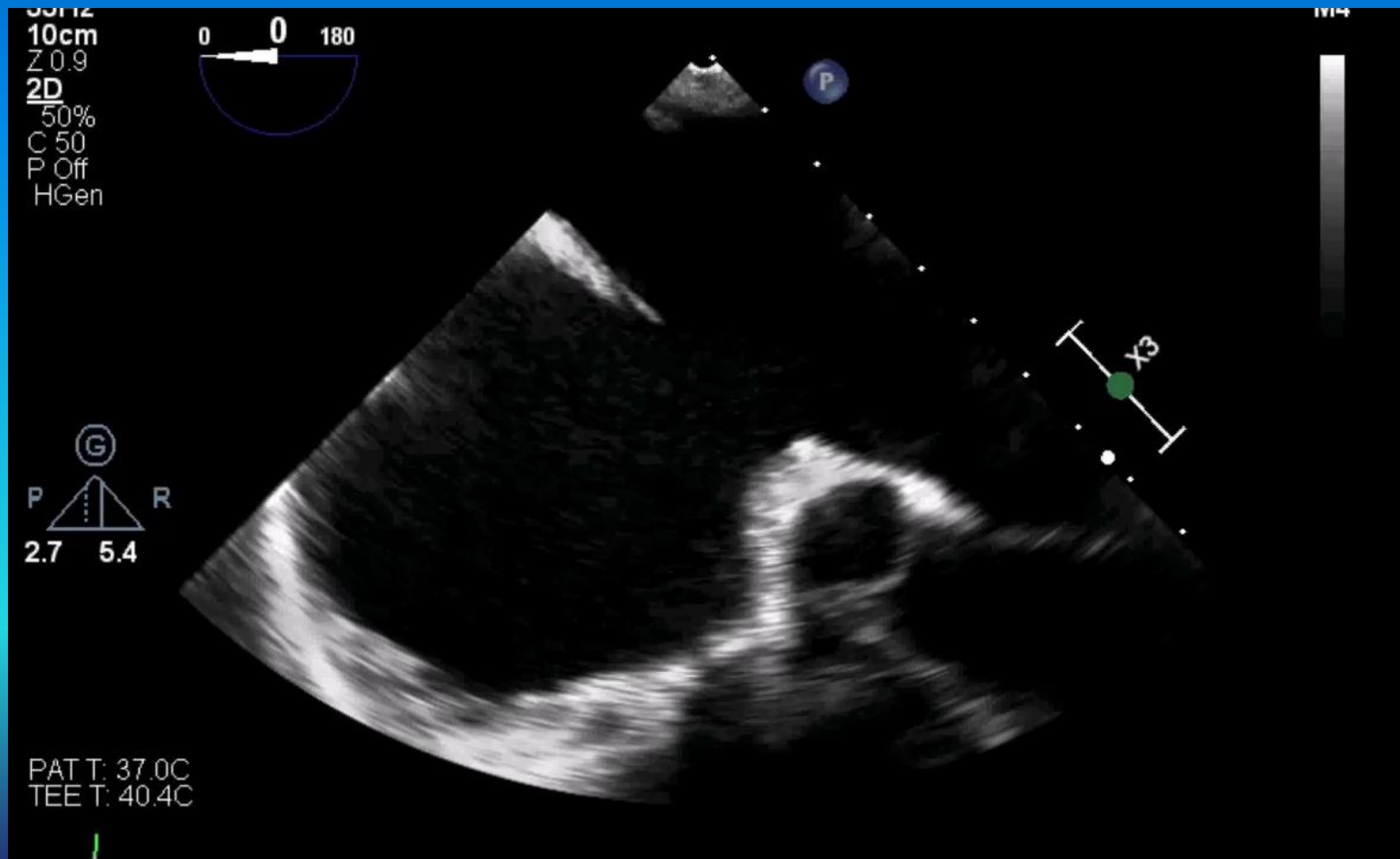
# Case presentation

- CVT consultation for emergency MV repair and TV repair and questionable of ASD.
- CVT requested for good quality imaging for ASD.

# Echocardiography: 21/2/2020

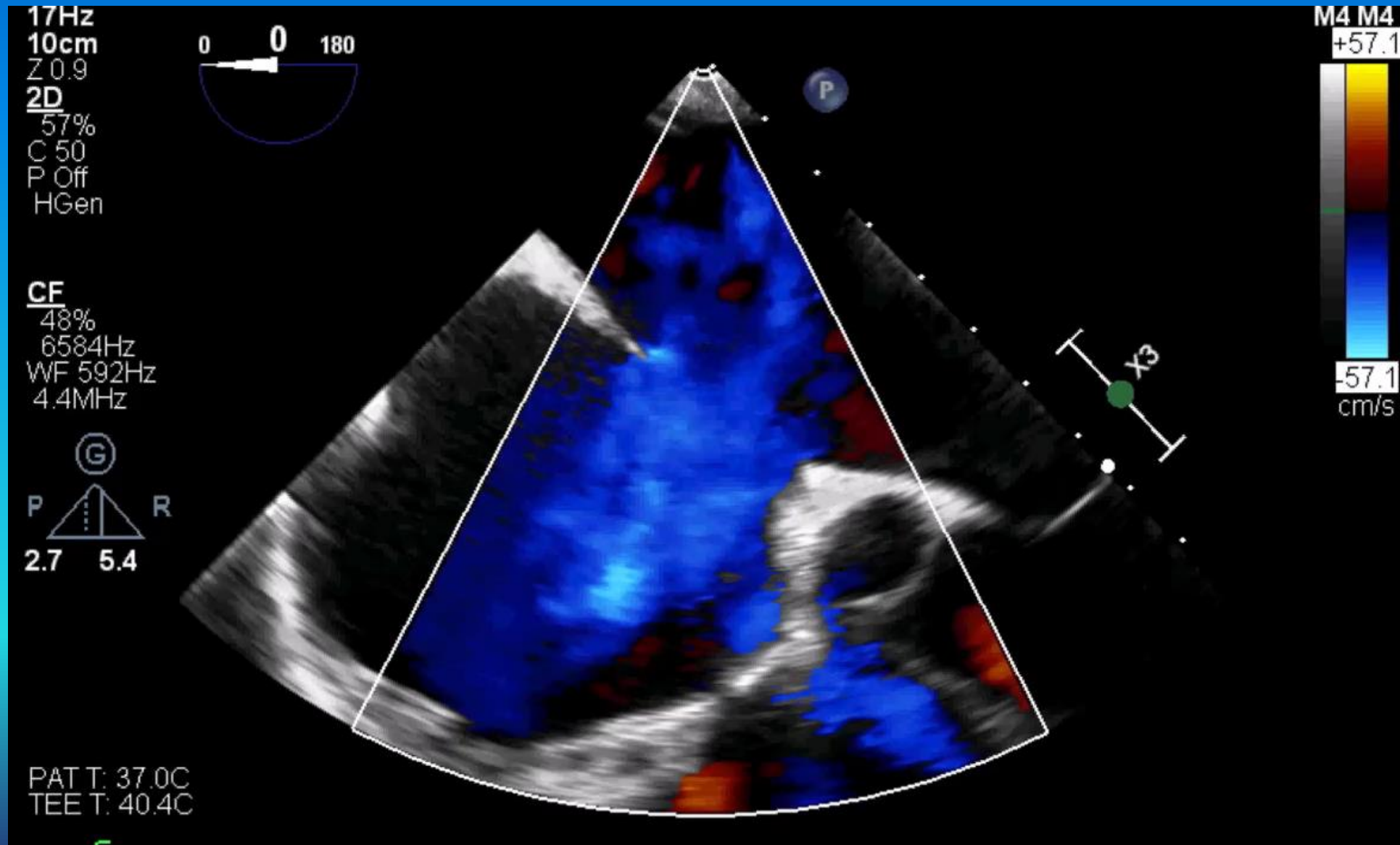


# TEE: D4 – 14<sup>th</sup> Mar 2020

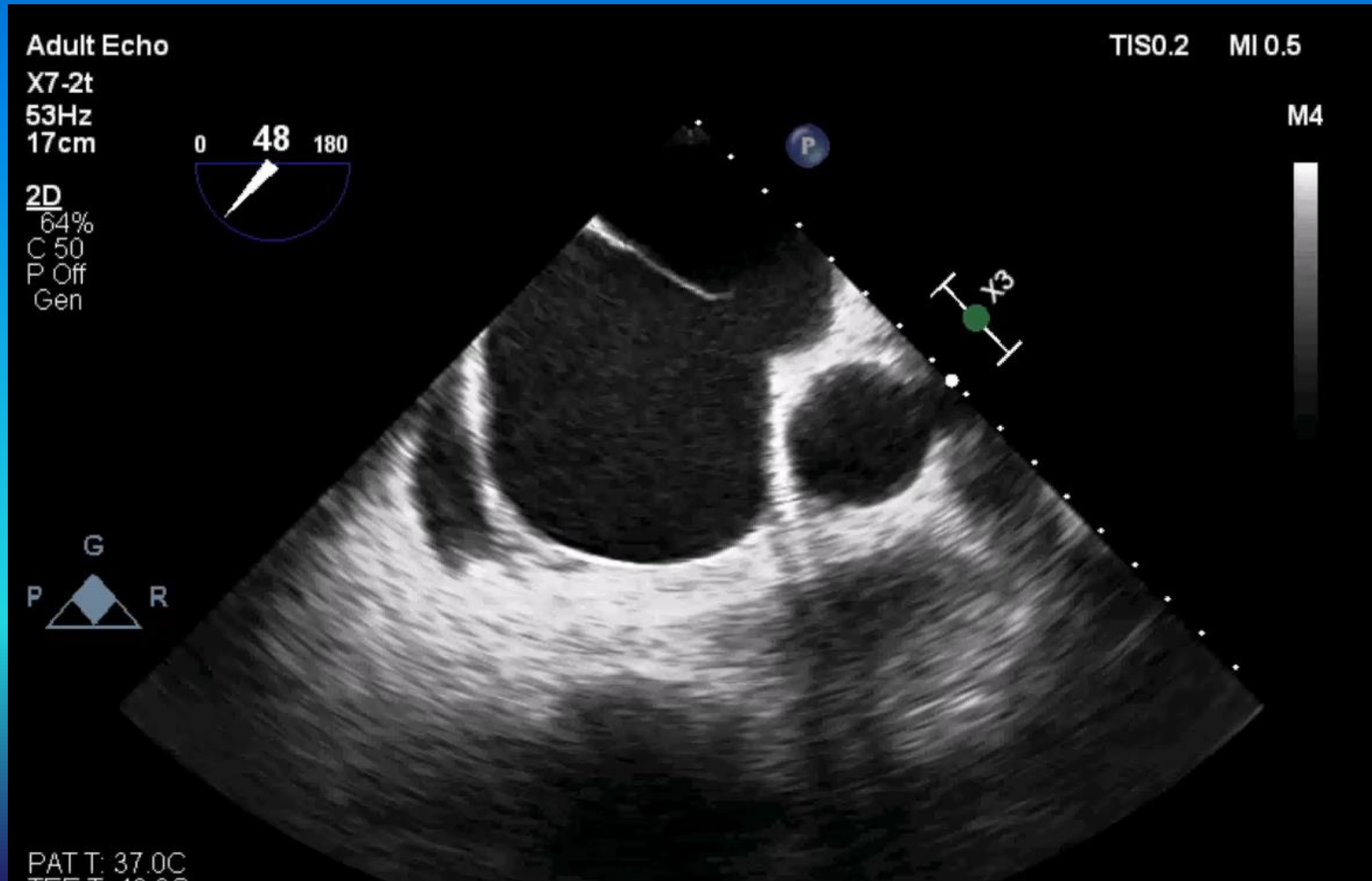




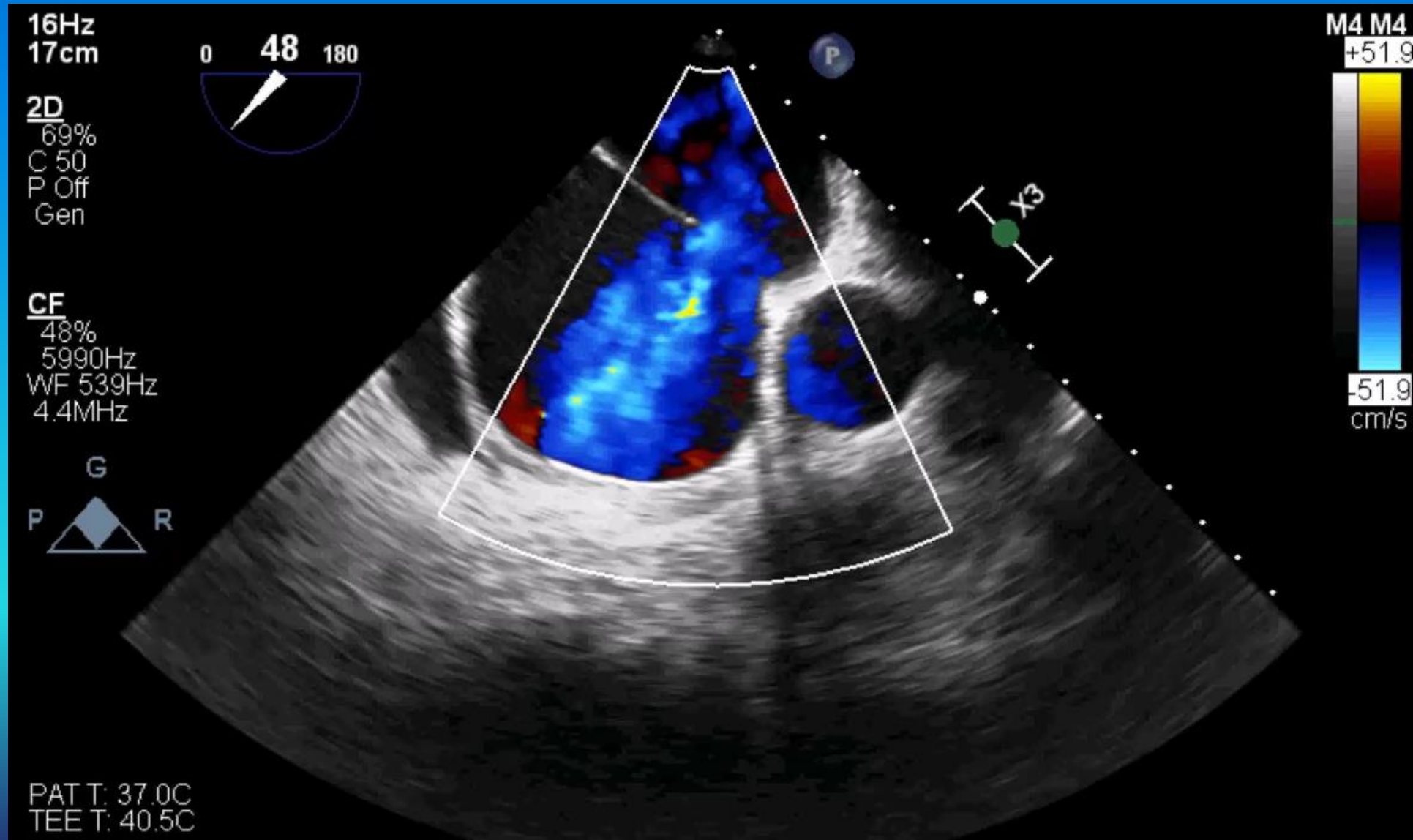
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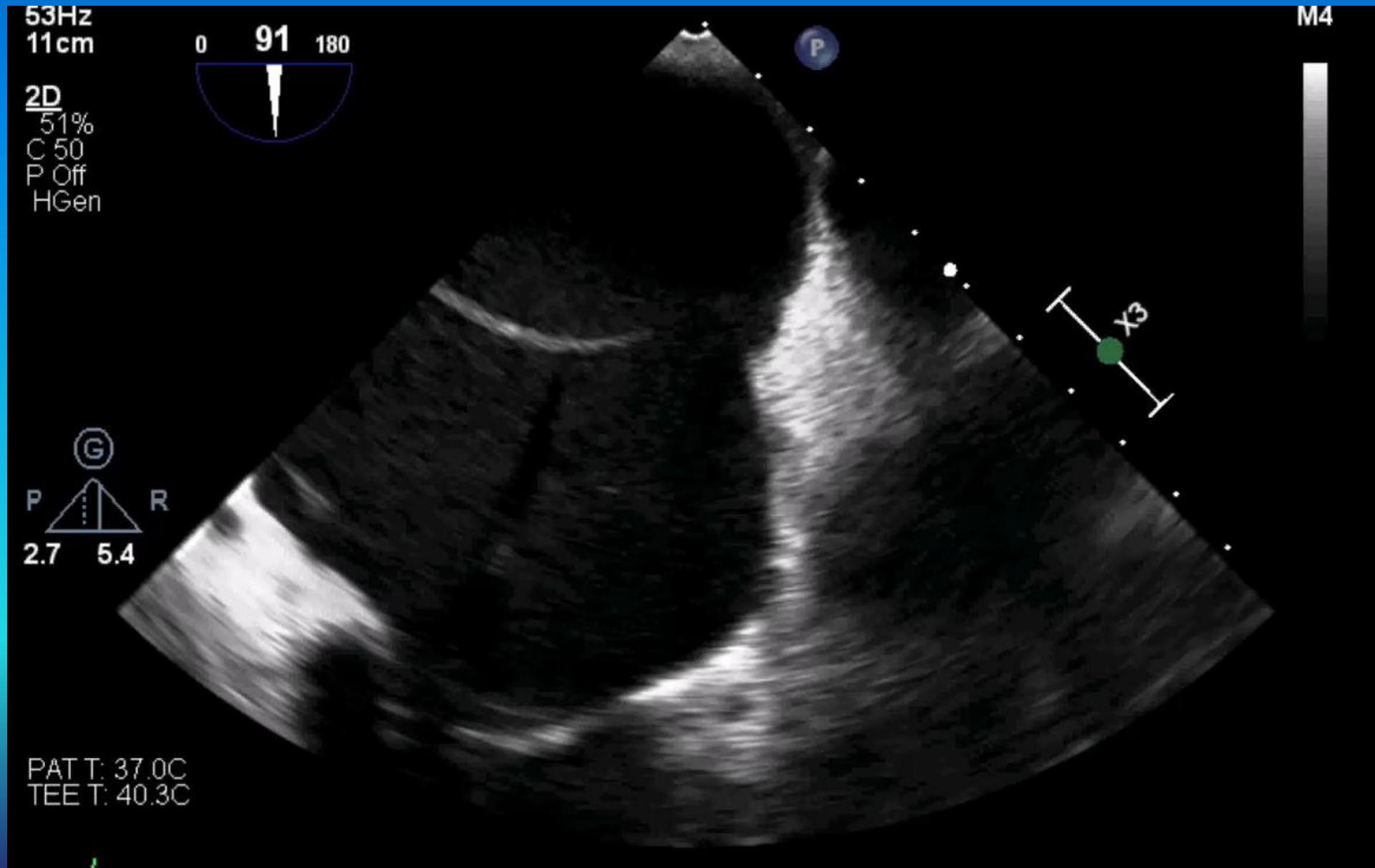
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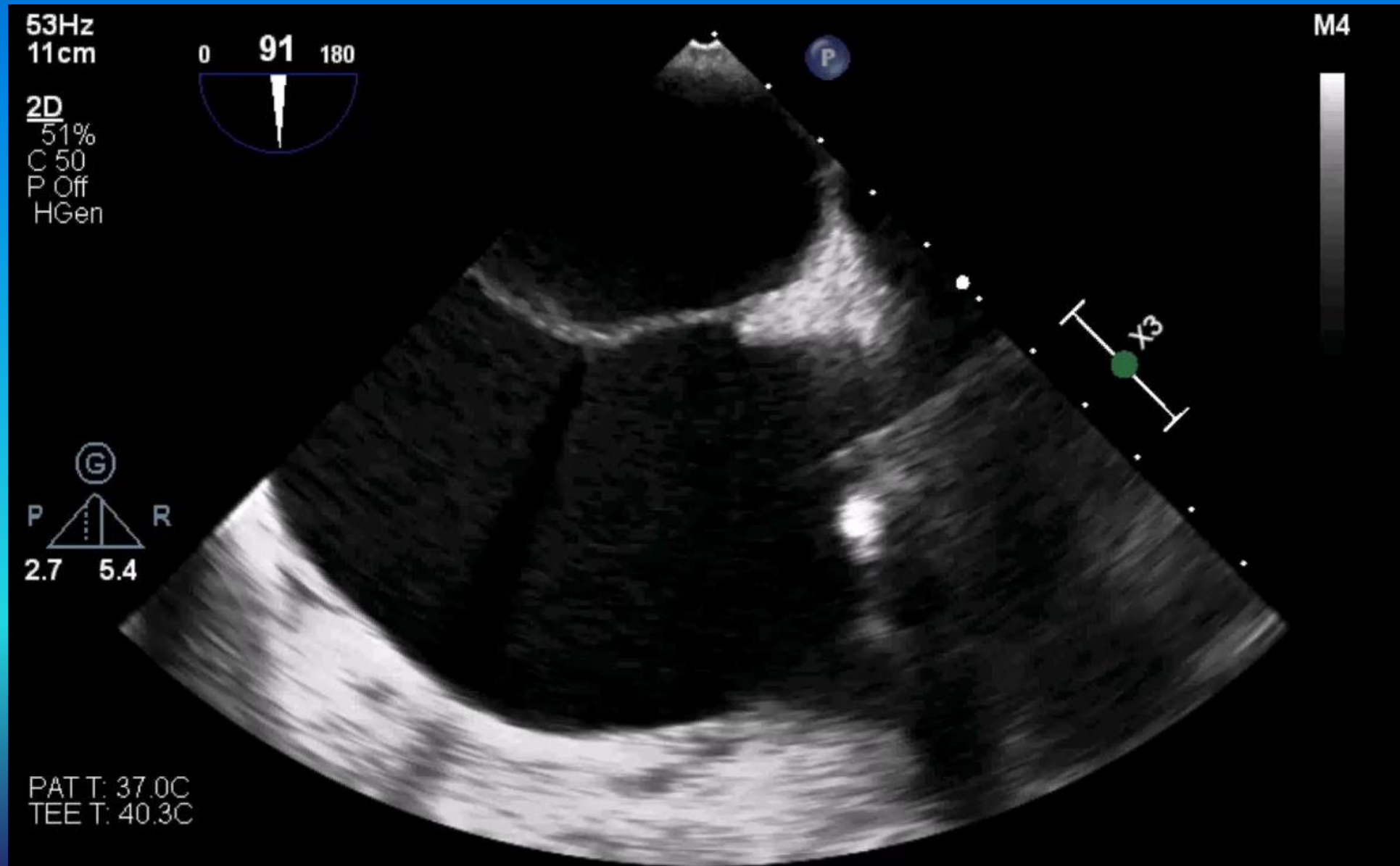


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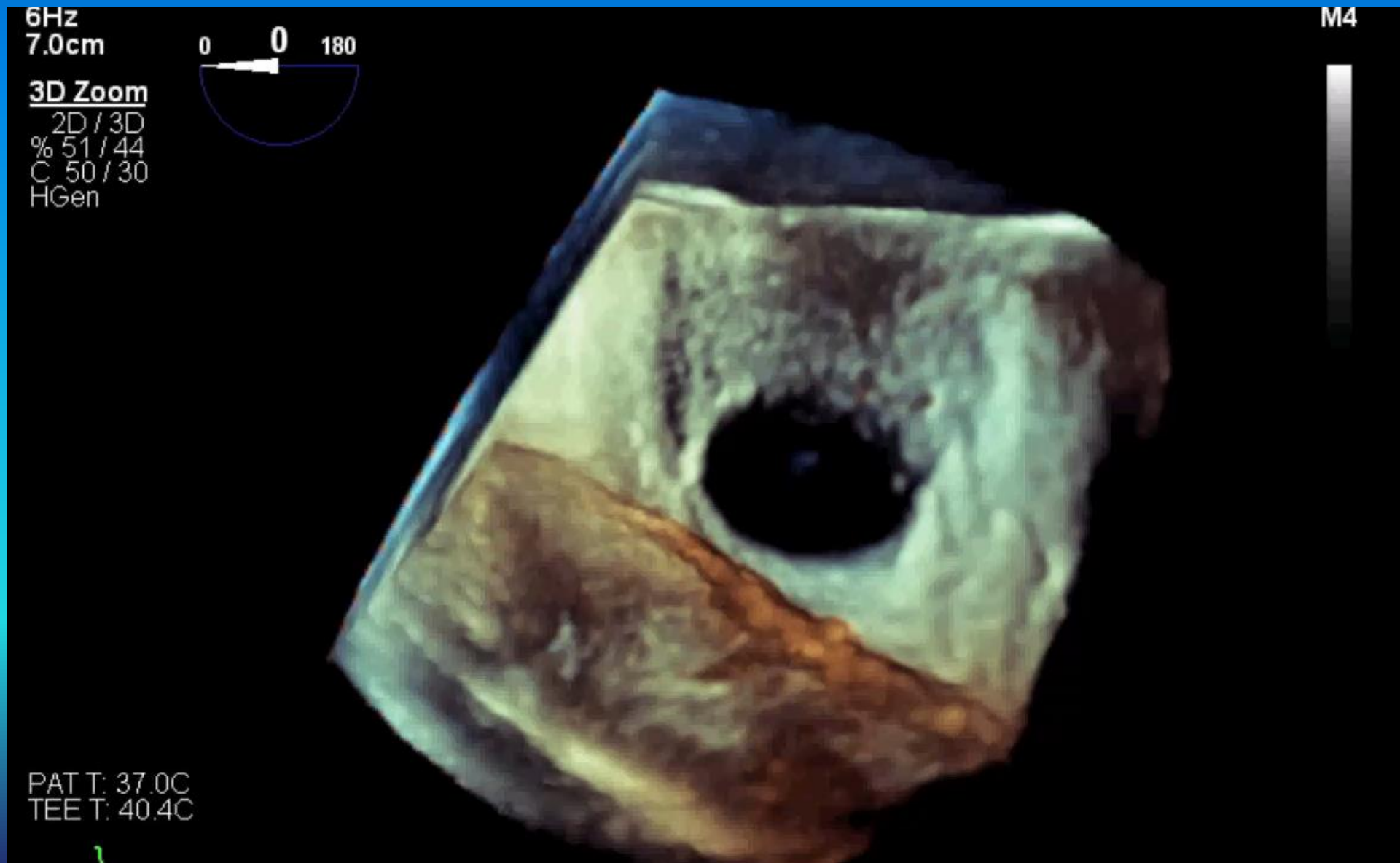




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# Case presentation

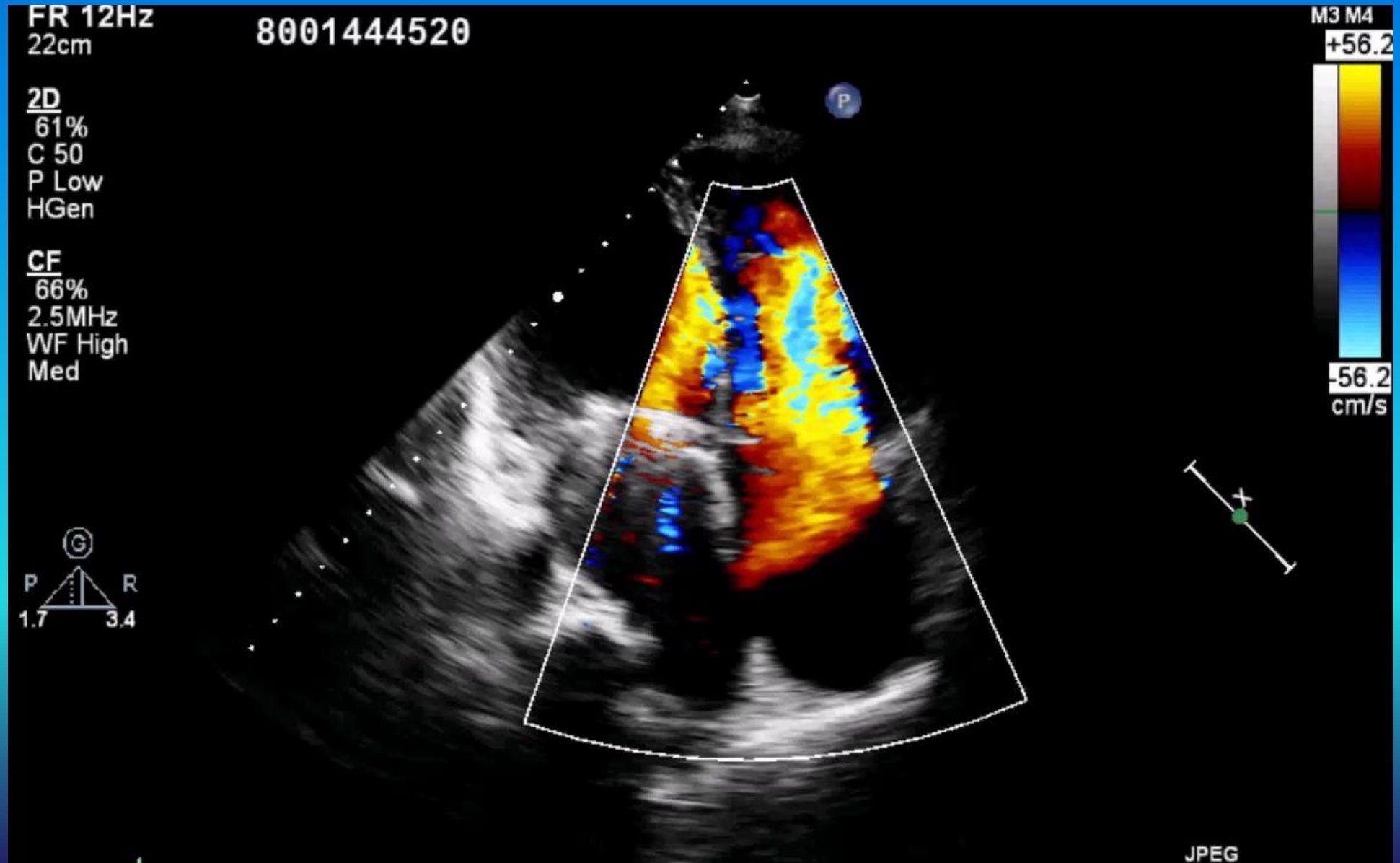
- How to management?
  - a. Emergency MV repair + TV repair + ASD closure
  - b. Percutaneous ASD closure and edge-to-edge for MV repair
  - c. Suggest end-of-life care

# Case presentation

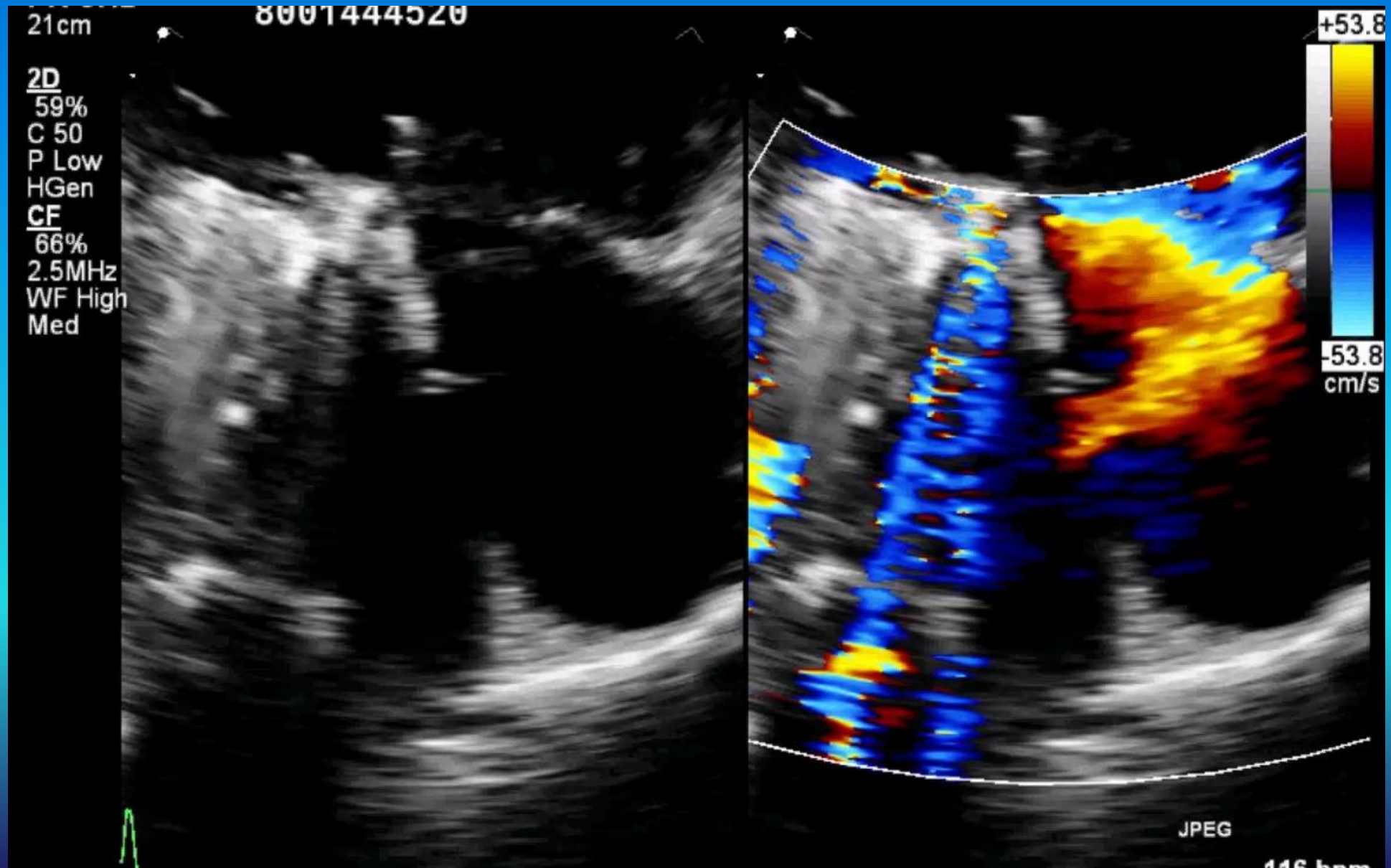
- Urgent ASD closure (using suture edge to edge without pericardial patch) with MVR + TV repair was done with good result.
- ECMO was removed in the operative theater and ET-tube was removed in the next morning.



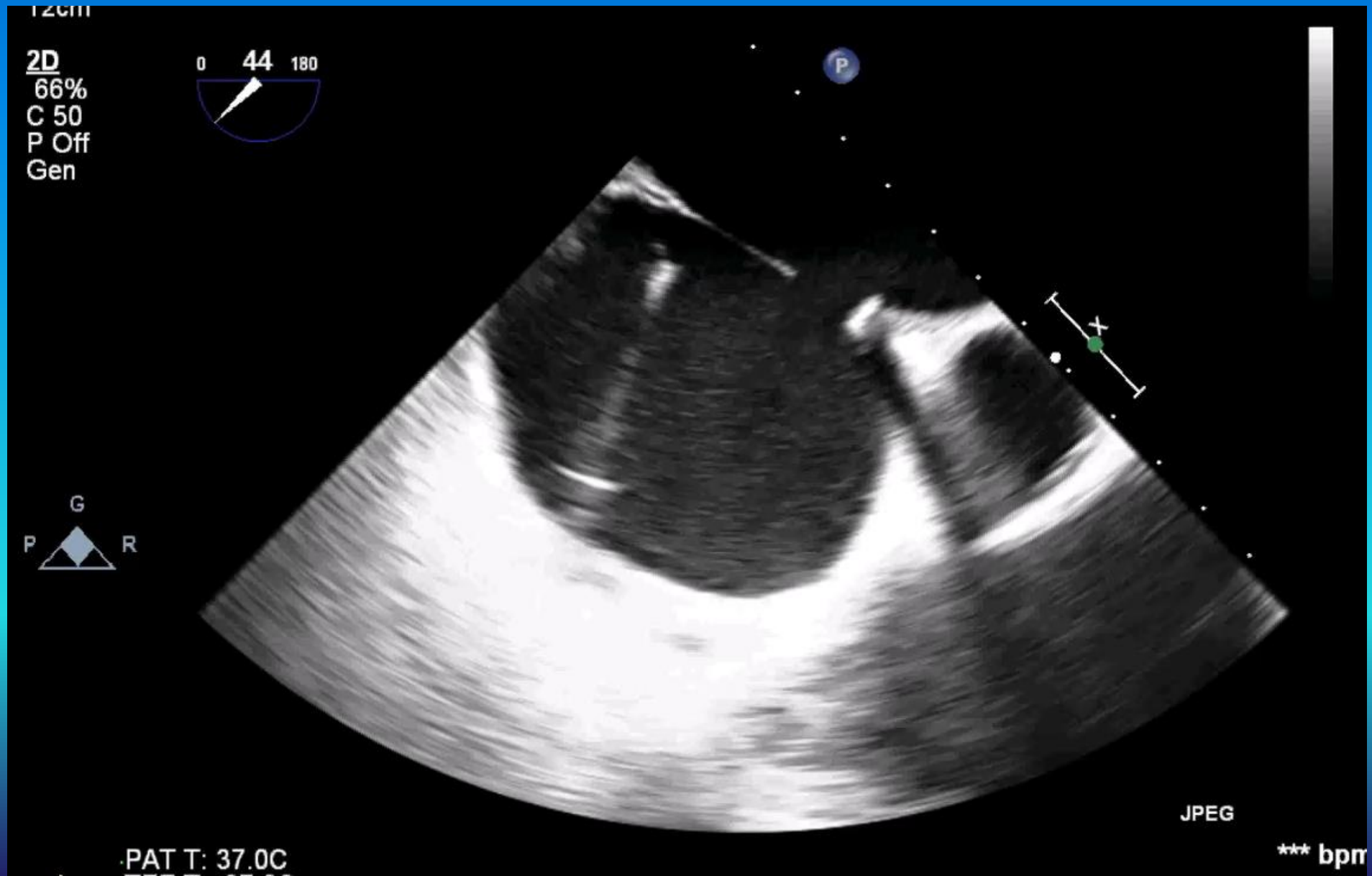
# Echocardiography: 18<sup>th</sup> Mar 2020 (3 days post Sx)



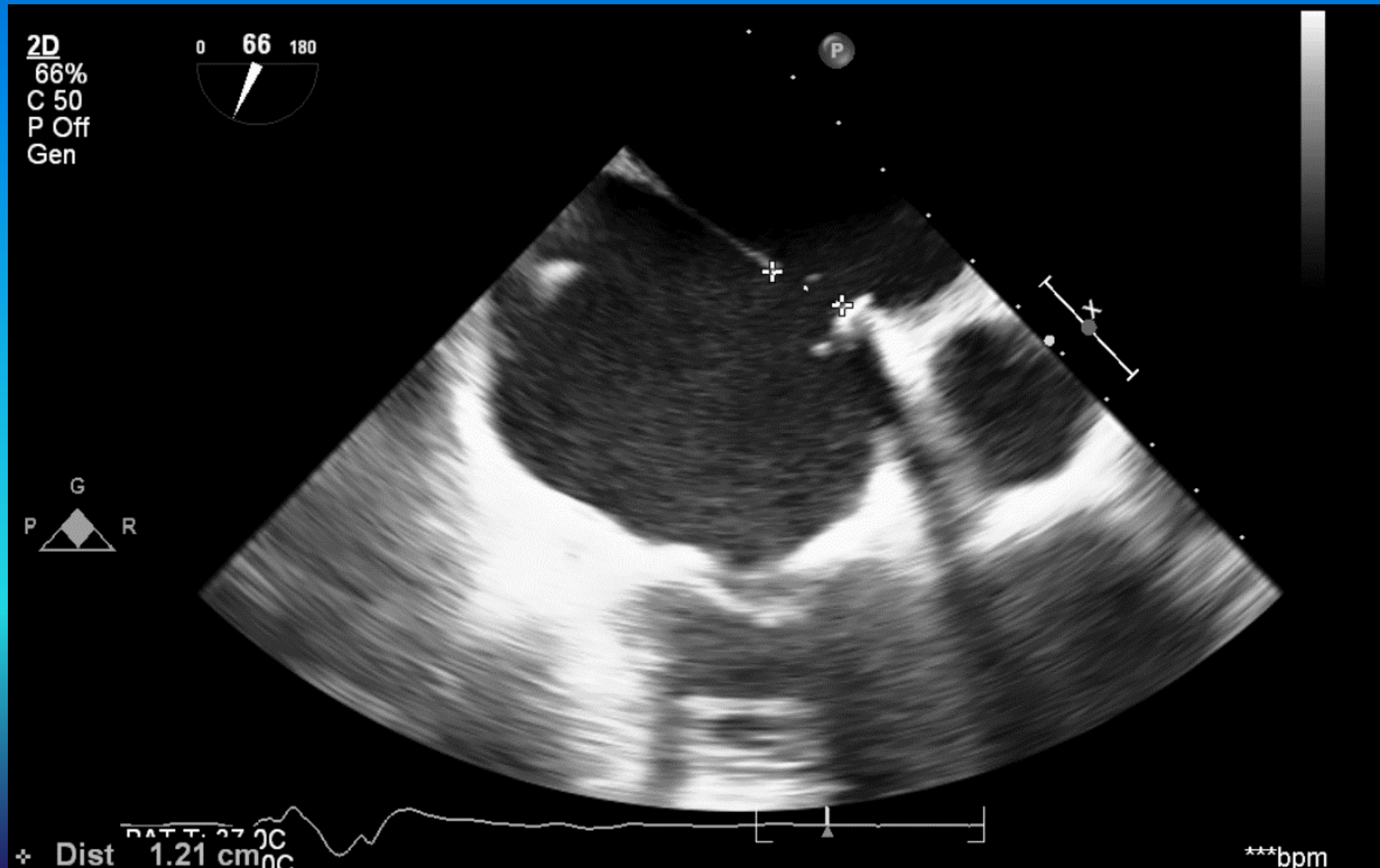
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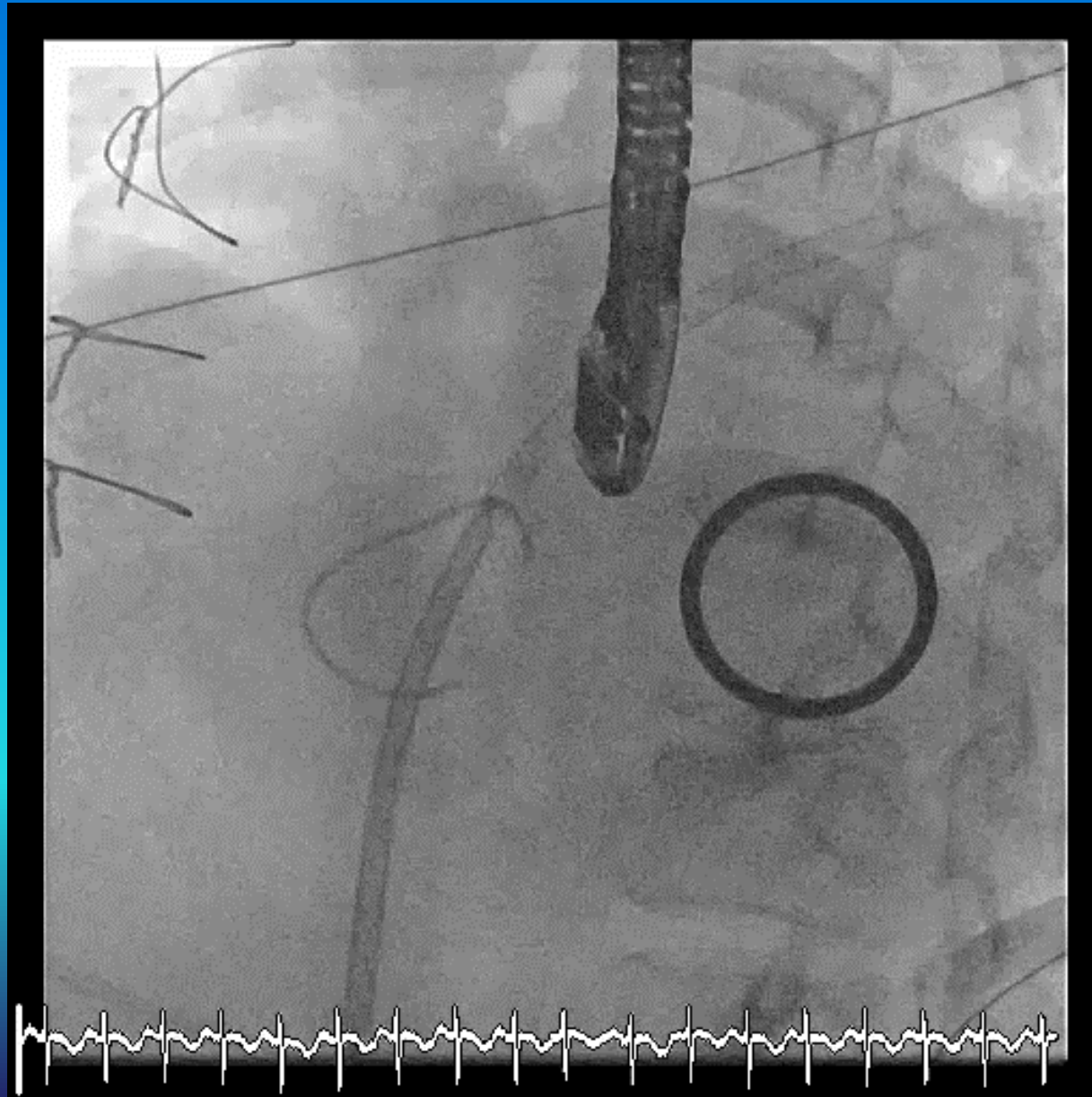


# Echocardiography: 18<sup>th</sup> Mar 2020 (3 days post Sx)



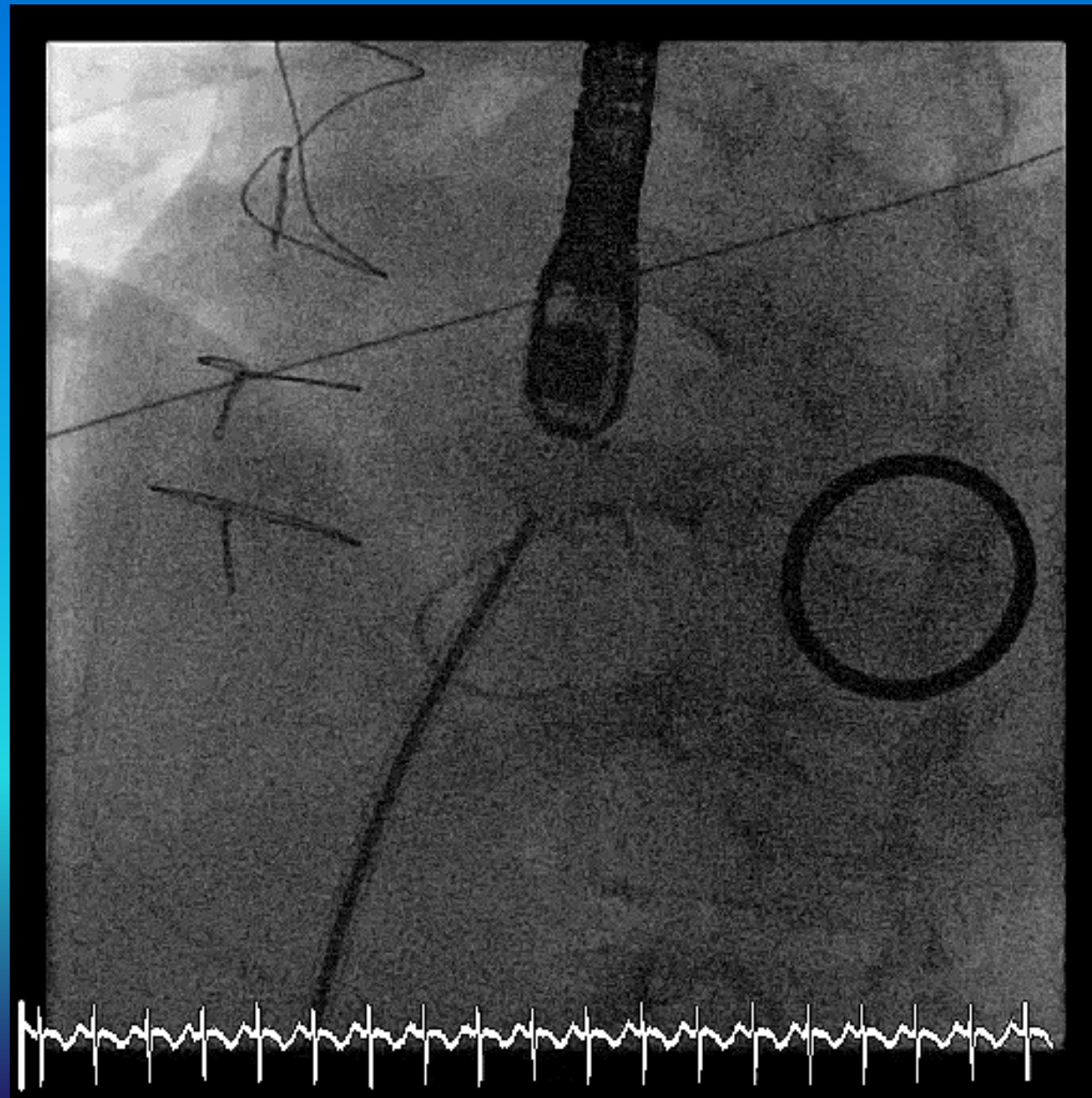


# Case presentation

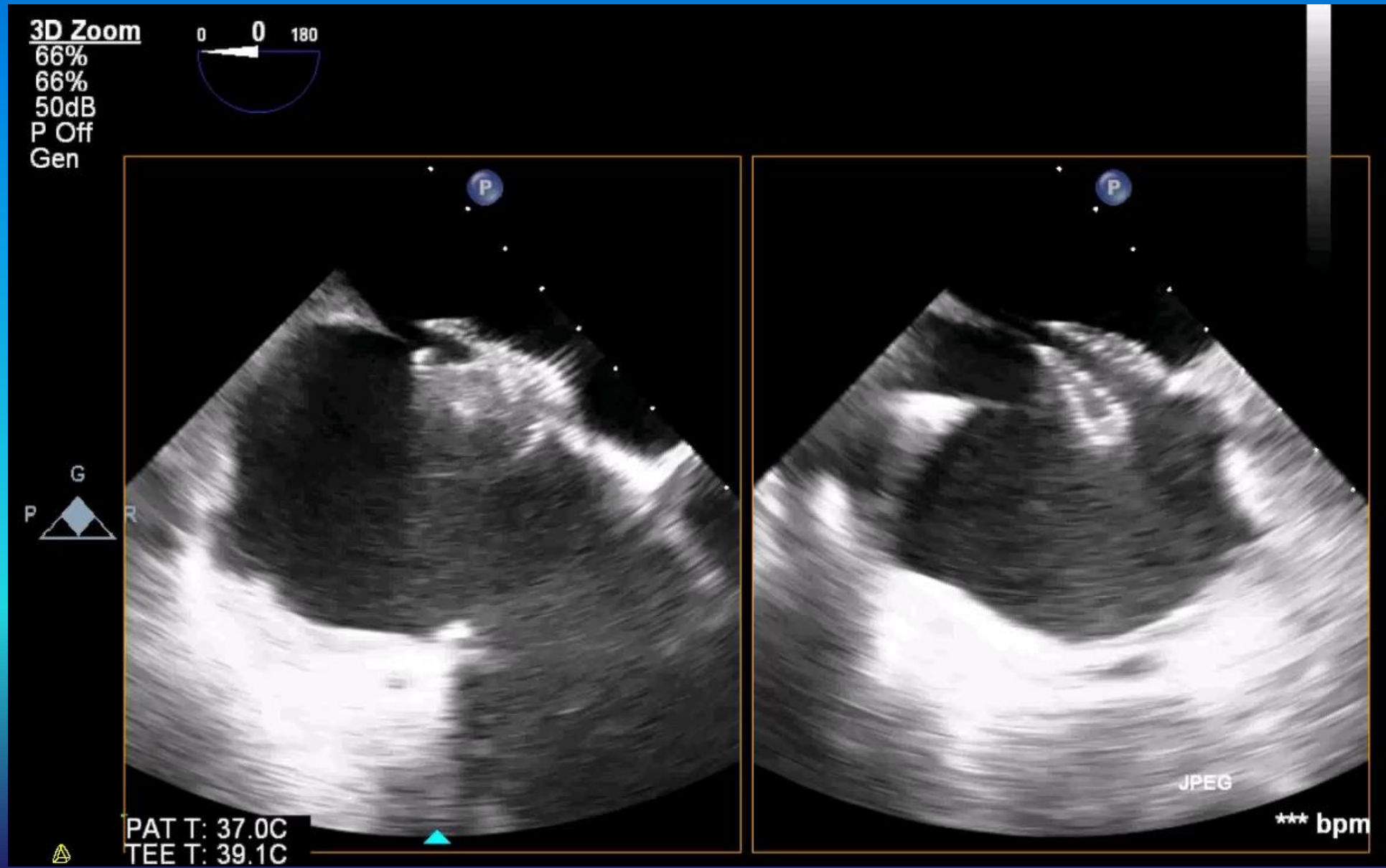




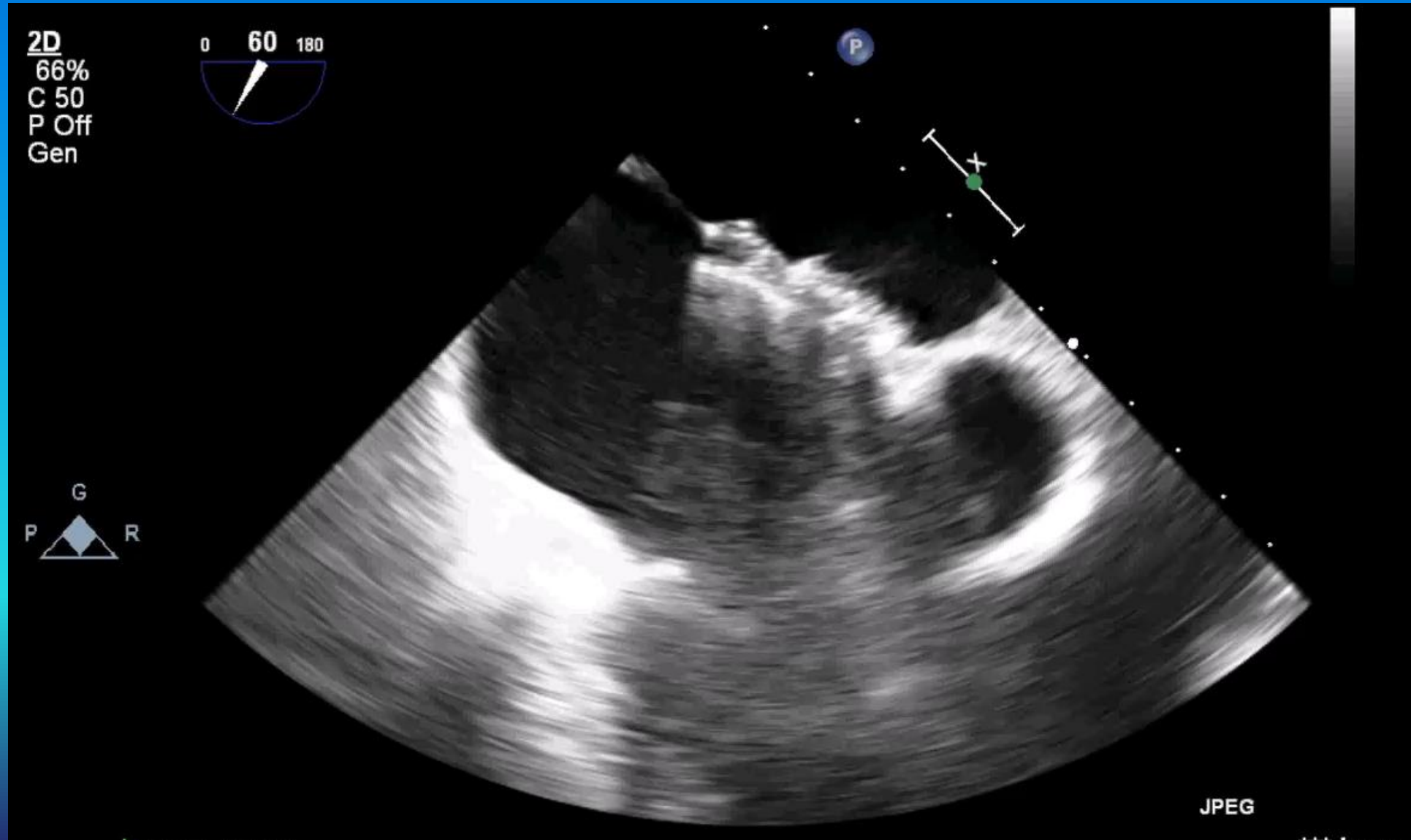
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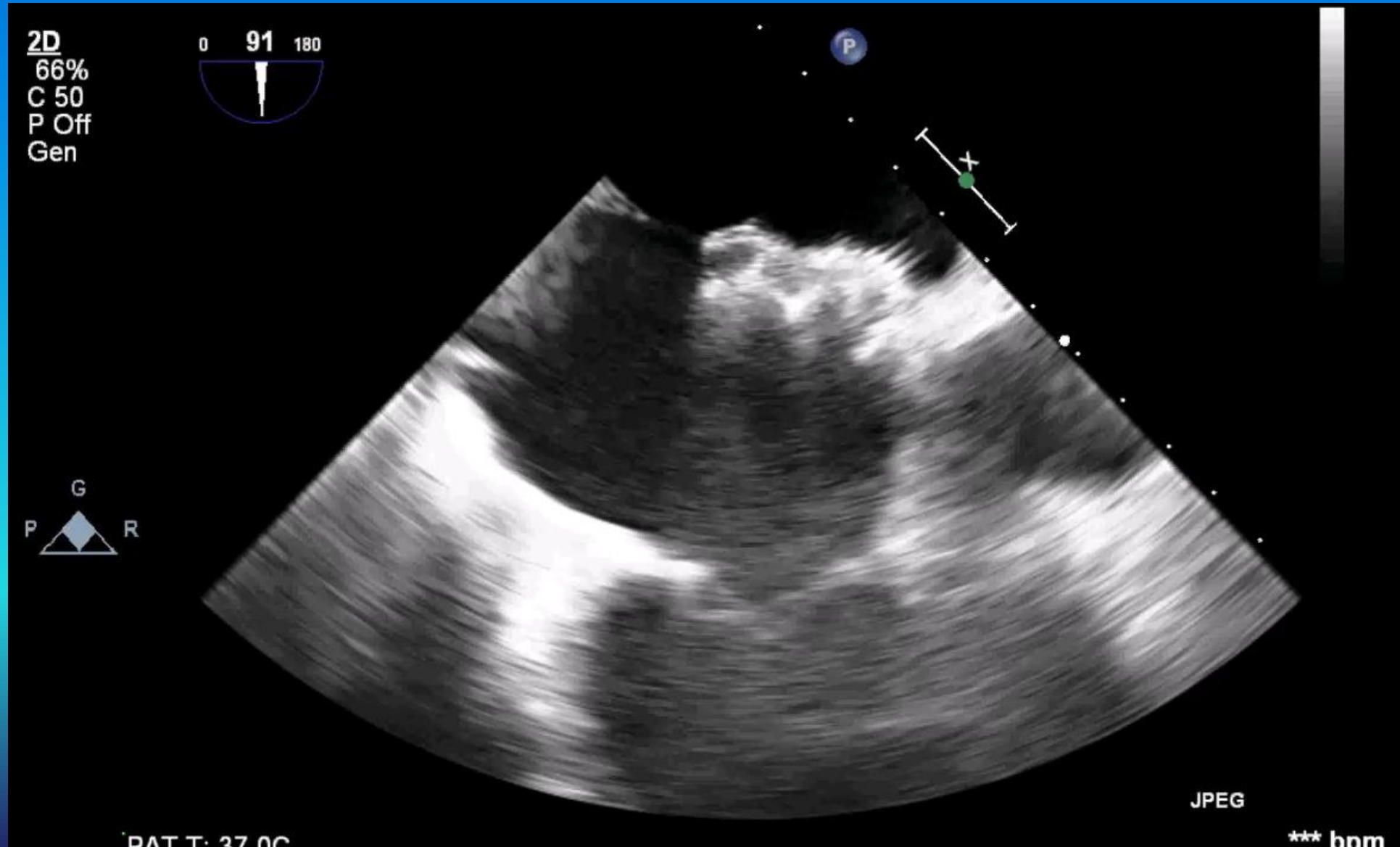
# Case presentation



# Case presentation

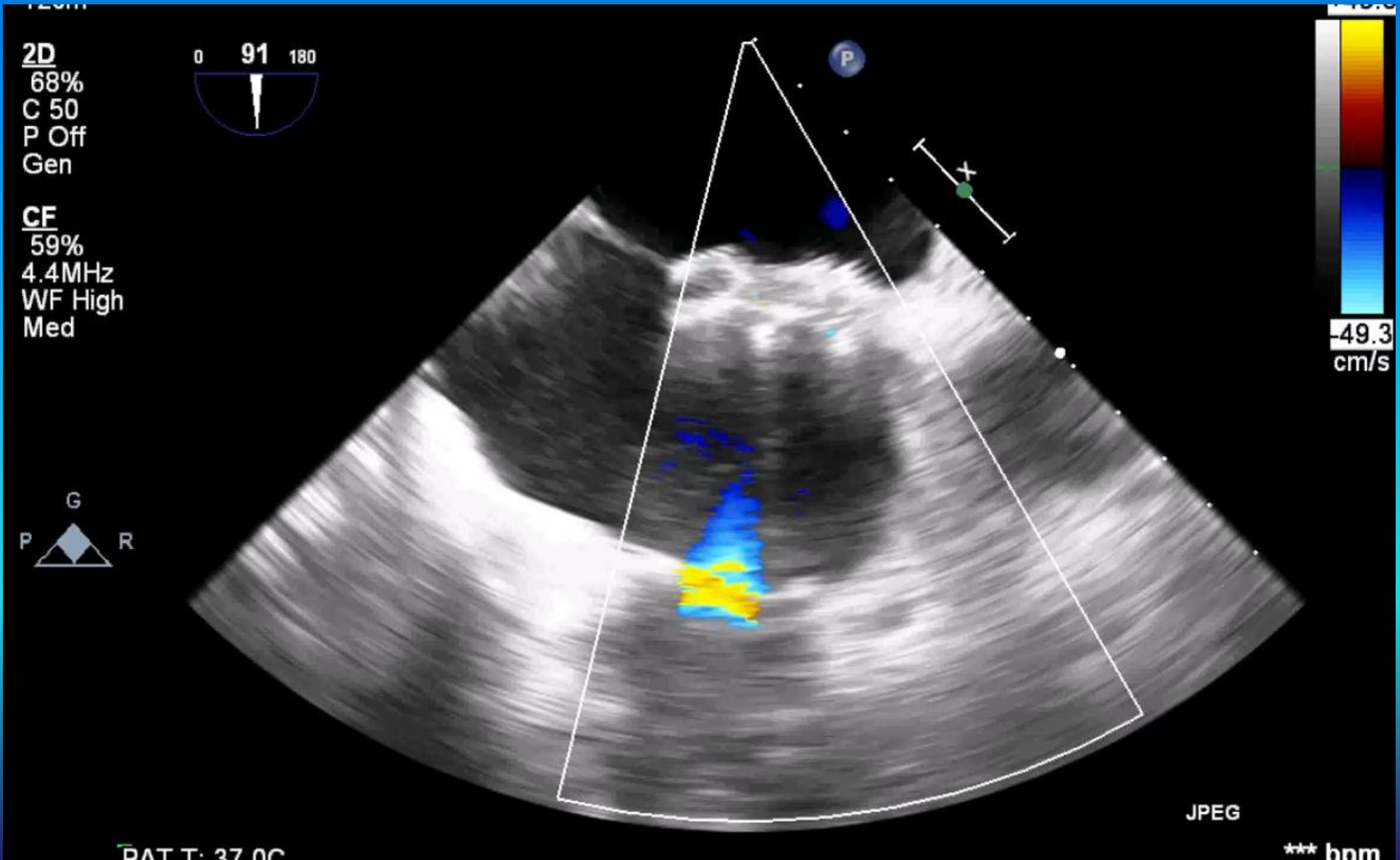


# Case presentation



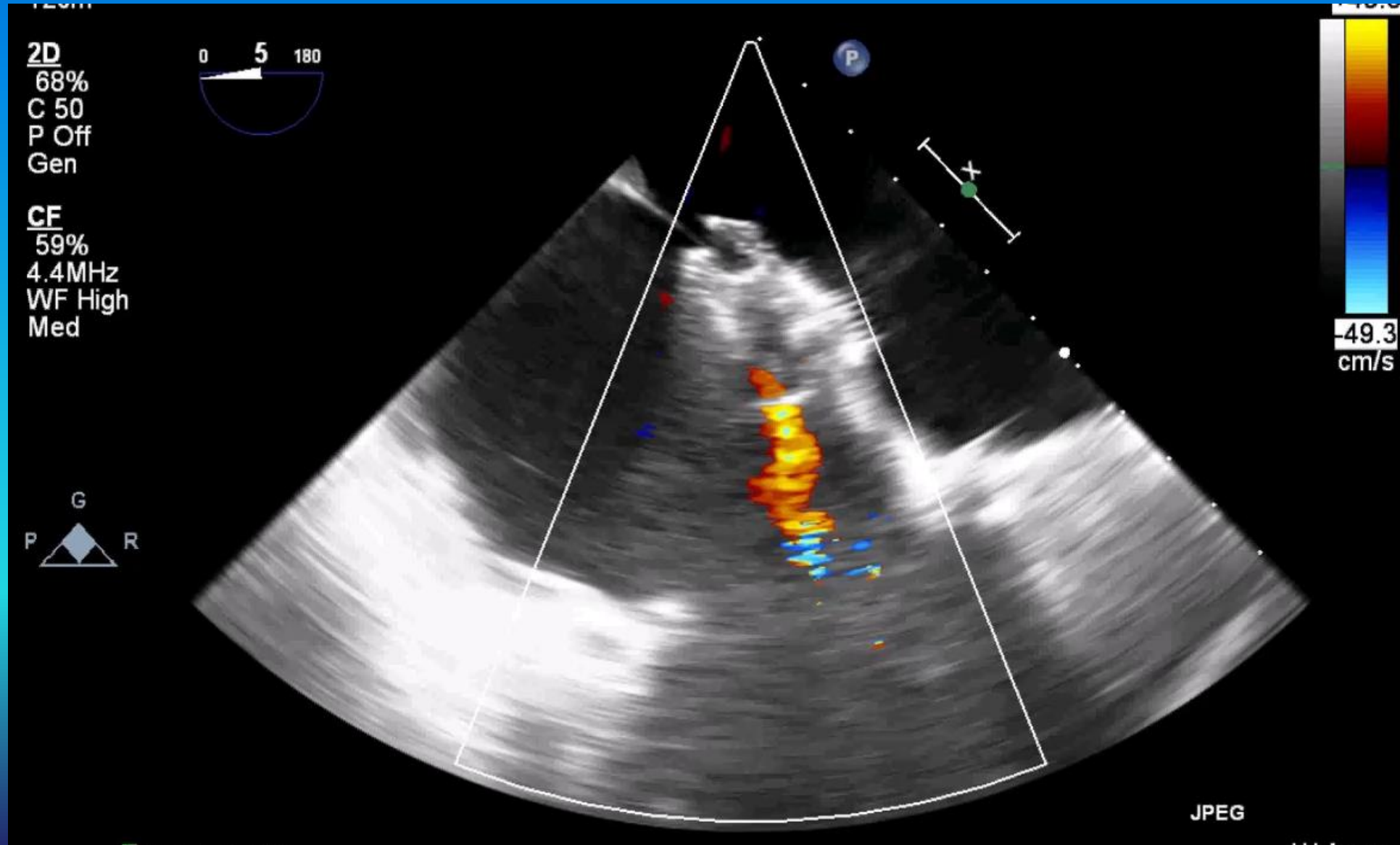


# Case presentation



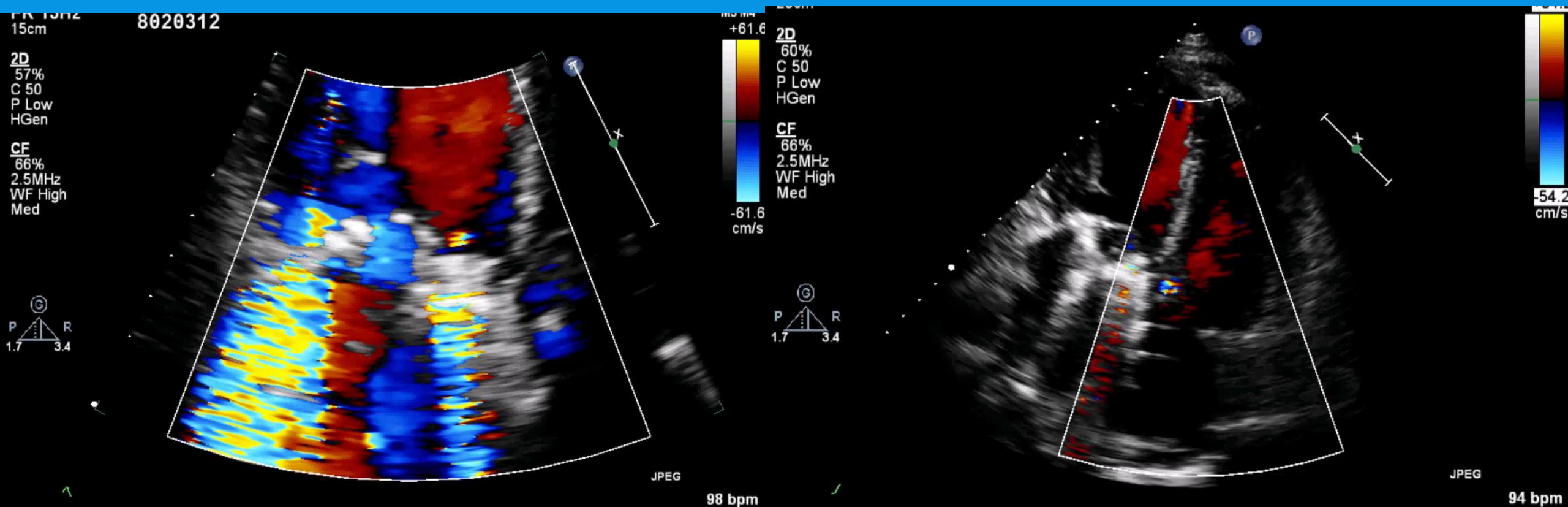


# Case presentation



# Case presentation

- Patient was discharge after ASD closure.



# Take home messages

- Heart team is important particular in very critical ill patients who need multidisciplinary management.
- Echocardiography have to extensive review to find out the cause of PAH.
- Degree and cause of PAH may determine the outcomes and prognosis of treatment.
- Early bridging ECMO before definite treatment may be needed in some situations.





*Thank you for your attention*