



Stepwise Therapy for COA, PDA, VSD with Severe PAH

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No disclosure

19 year-old woman presented with dyspnea and hoarseness.

PE: HR 110/min regular

133/65 (85%)	126/63 (84%)
121/40 (75%)	123/80 (76%)

Very active precordium and suprasternal notch area, mild digital clubbing.

Very loud P_2 .

Gr 3-4/6 to and fro murmur LUSB.

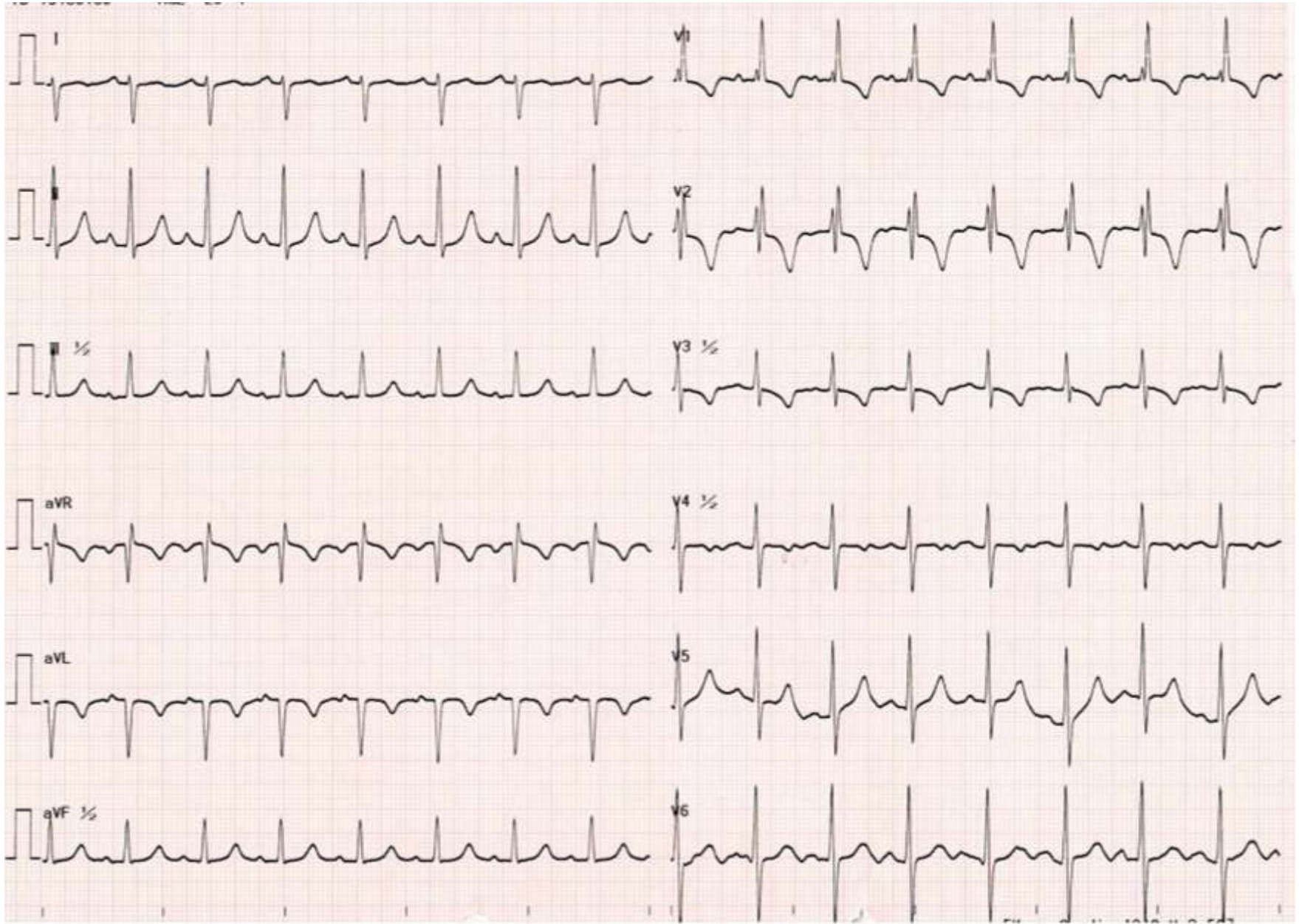
Fine crepitation bilaterally both lungs.

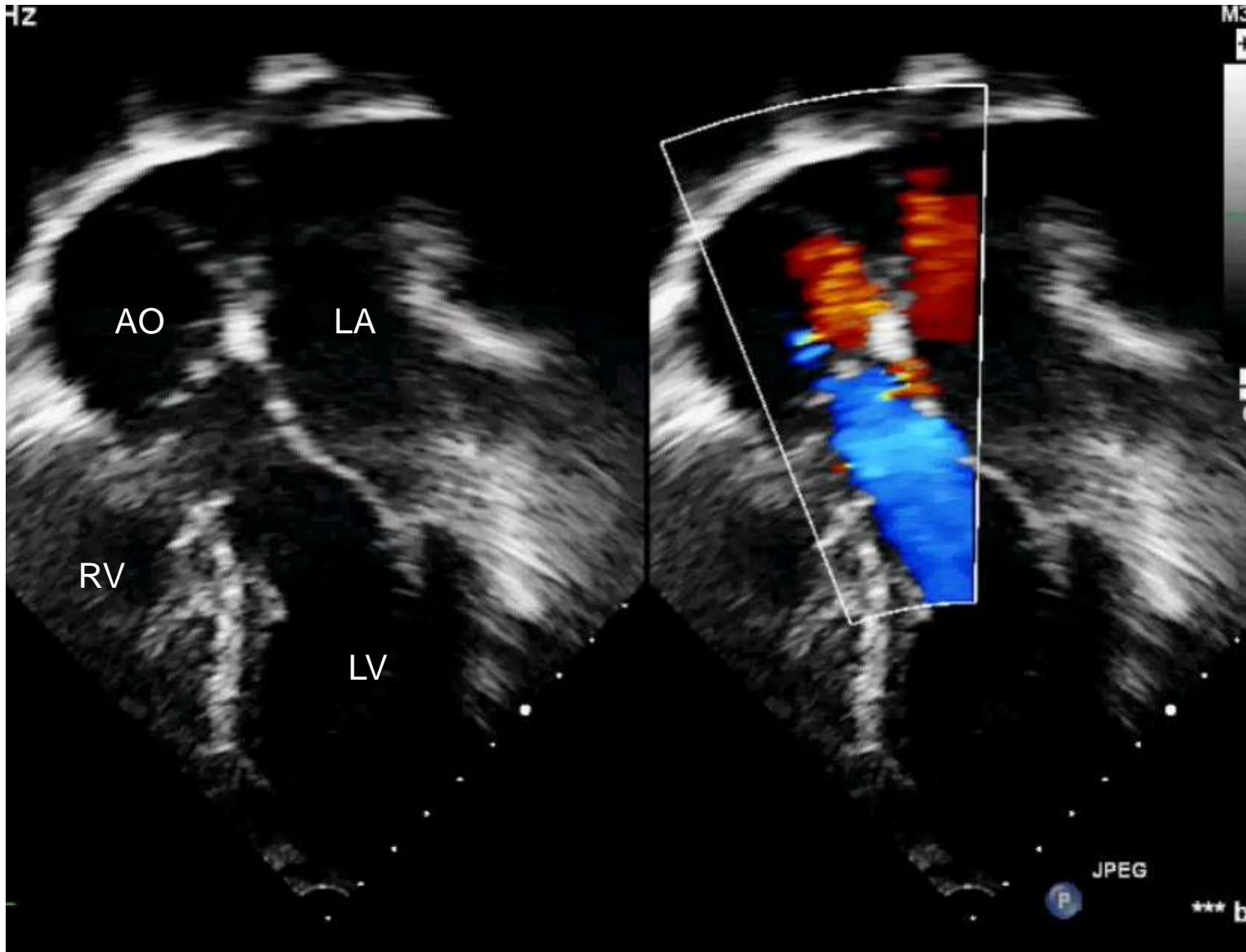
No hepatomegaly.

CXR

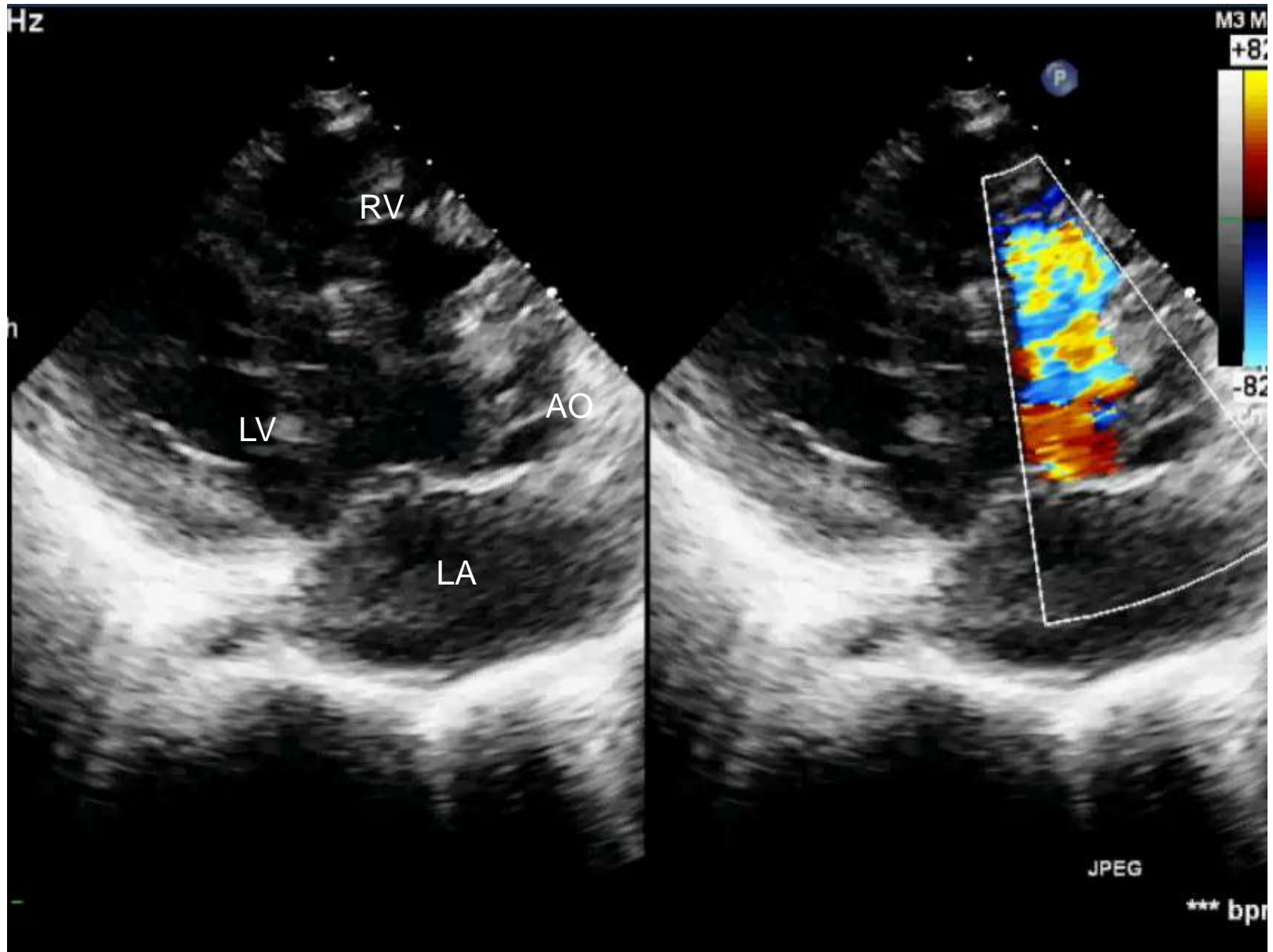


ECG

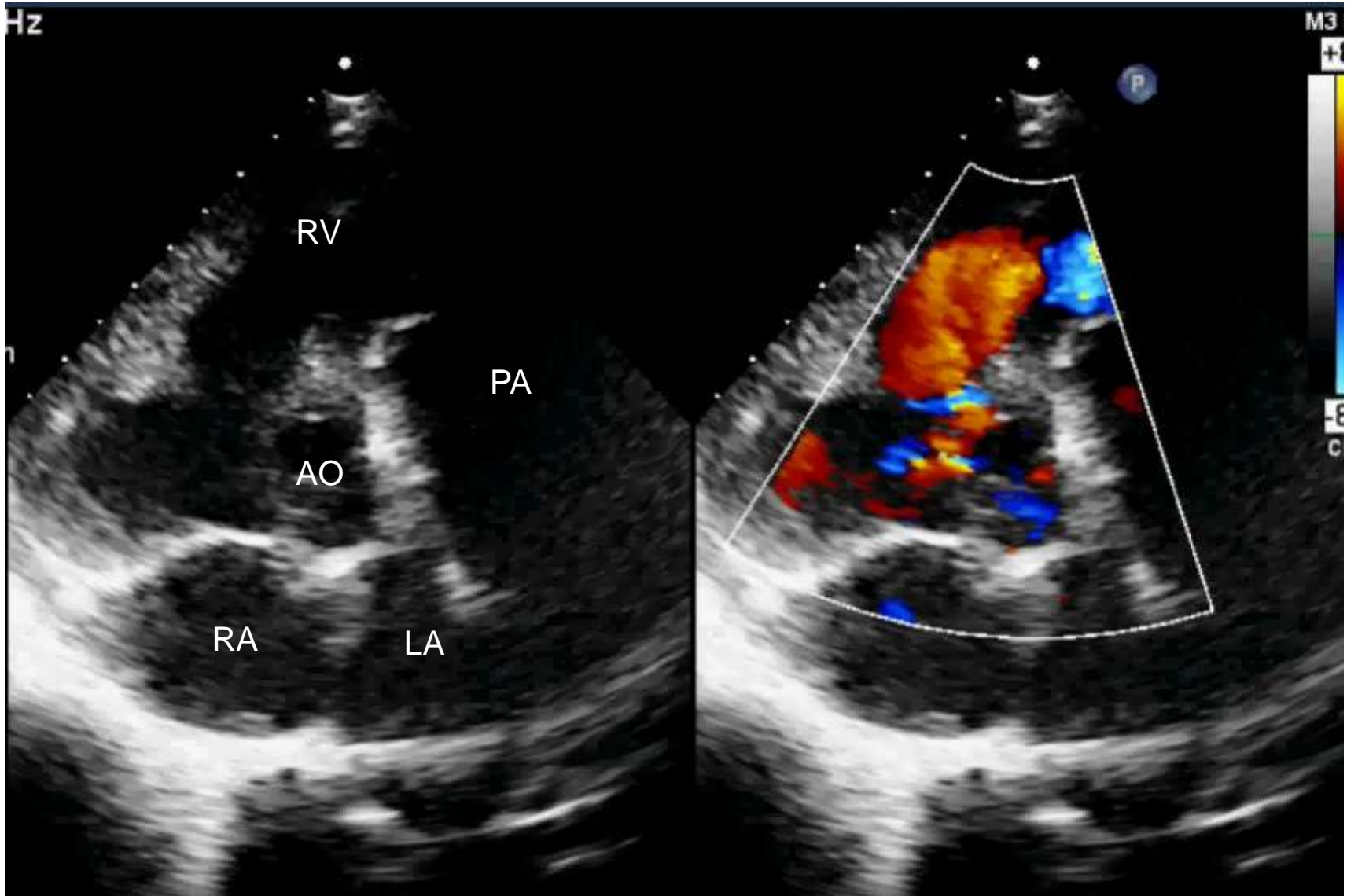




19 years old lady with hoarseness and differential cyanosis.



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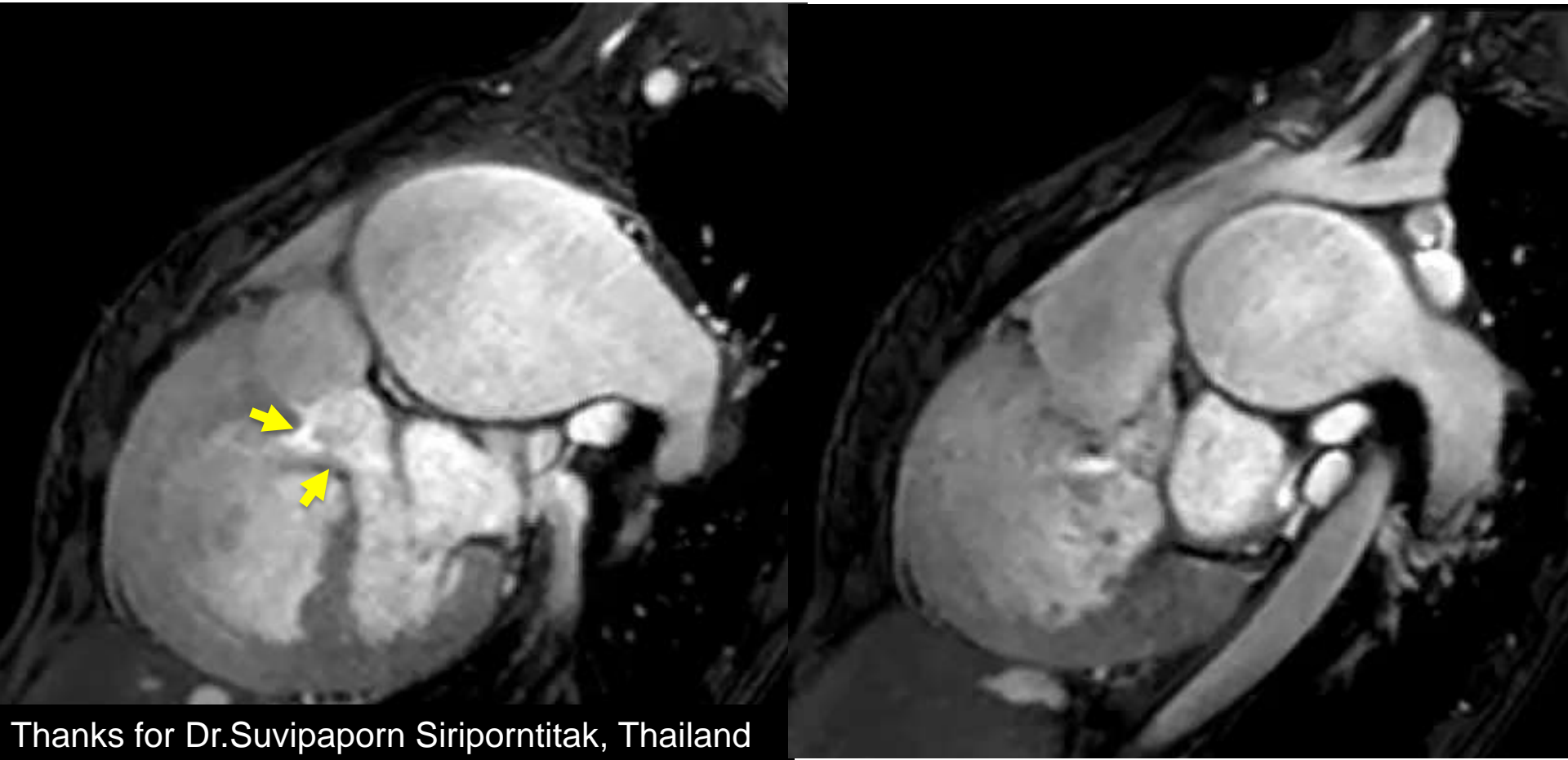


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Poor window for suprasternal notch view!

19 years old lady with hoarseness and differential cyanosis.

CMR



Thanks for Dr.Suvipaporn Siriporntitak, Thailand

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CMR



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CMR

Heart:

- o LV dilatation with LVH and normal systolic function
- o LVEF=62% 2
- o LVEDV index 172.2 ml/m
- o Paradoxical septal wall motion
- o Mild aortic regurgitation, regurgitation fraction 17%
- o RV dilation with RVH
- o RVEF = 50.4% 2
- o RVEDV index 230.4 ml/m
- o **Severe PR**; Regurgitation fraction = 55%
- o Mild TR; Regurgitation fraction =17.2%
- o **Large VSD with bidirectional shunt**;
 - o Perimembranous type, 24 mm in size.
 - o Qp:Qs for only VSD (stroke volume of RV : LV) = 1.1:1
- **Large PDA with bidirectional shunt**
 - o Length 12.9 mm, diameter 10.6 mm at aortic end and 12.7 mm at pulmonic end.
 - o Total Qp/Qs = 2.8:1? (MPA:aorta flow)

Aorta:

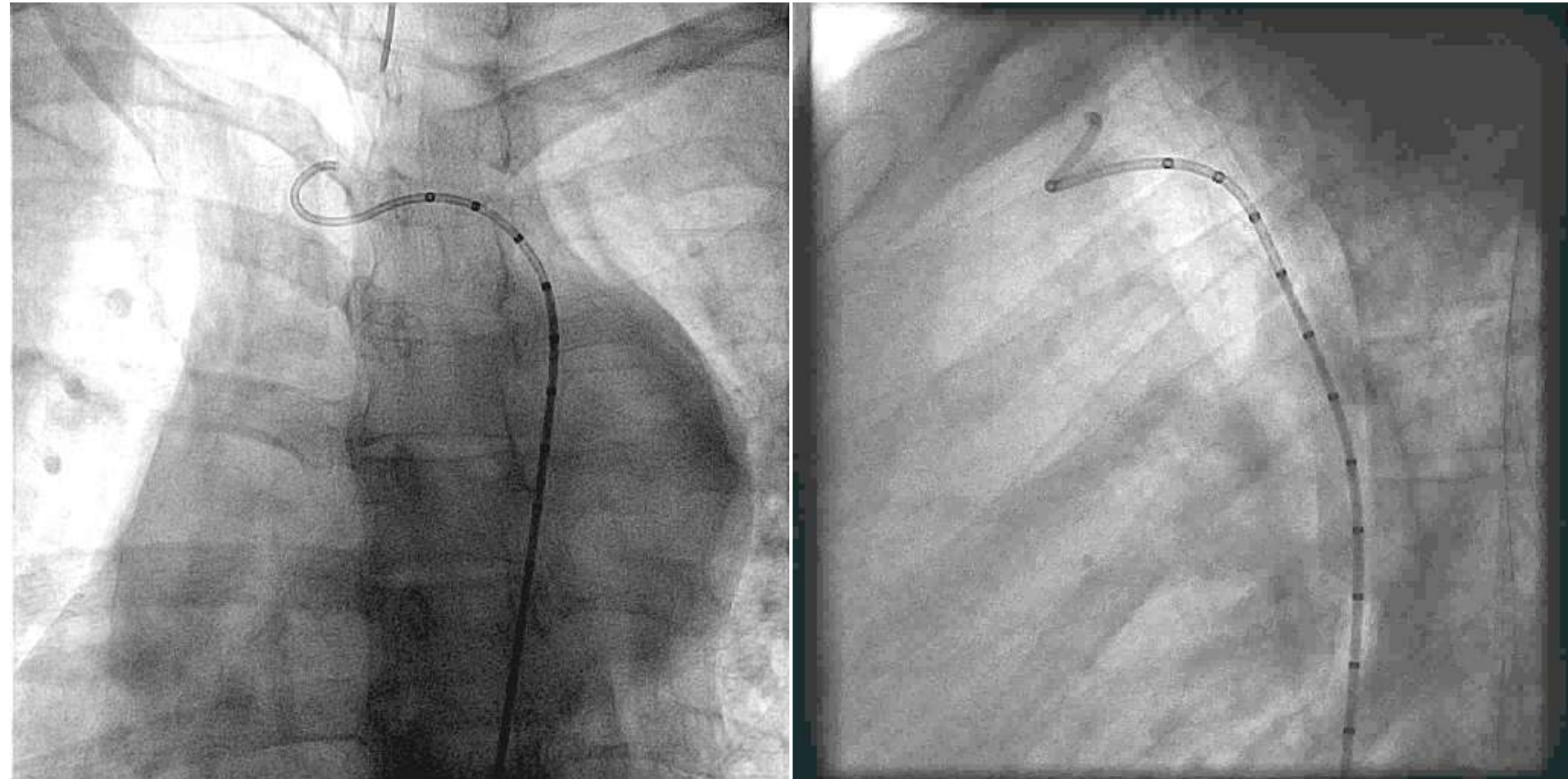
- o Left sided aortic arch with normal branching
- o Mild hypoplastic of the distal aortic arch and aortic isthmus
- o **Severe form of discrete juxtaductal coarctation** (at same level with PDA)
 - o Size 5.5 mm, at 1.4 cm distal to Lt SCA origin.
 - o Limitation in evaluation gradient across the coarctation.
 - o No significant collateral demonstrated.

MPA:

- o Large fusiform MPA aneurysm, enlarged RPA and LPA suggestive of PAHT.

Chest: Multifocal areas of air-trapping at the RLL, LUL and LLL.

Aortogram



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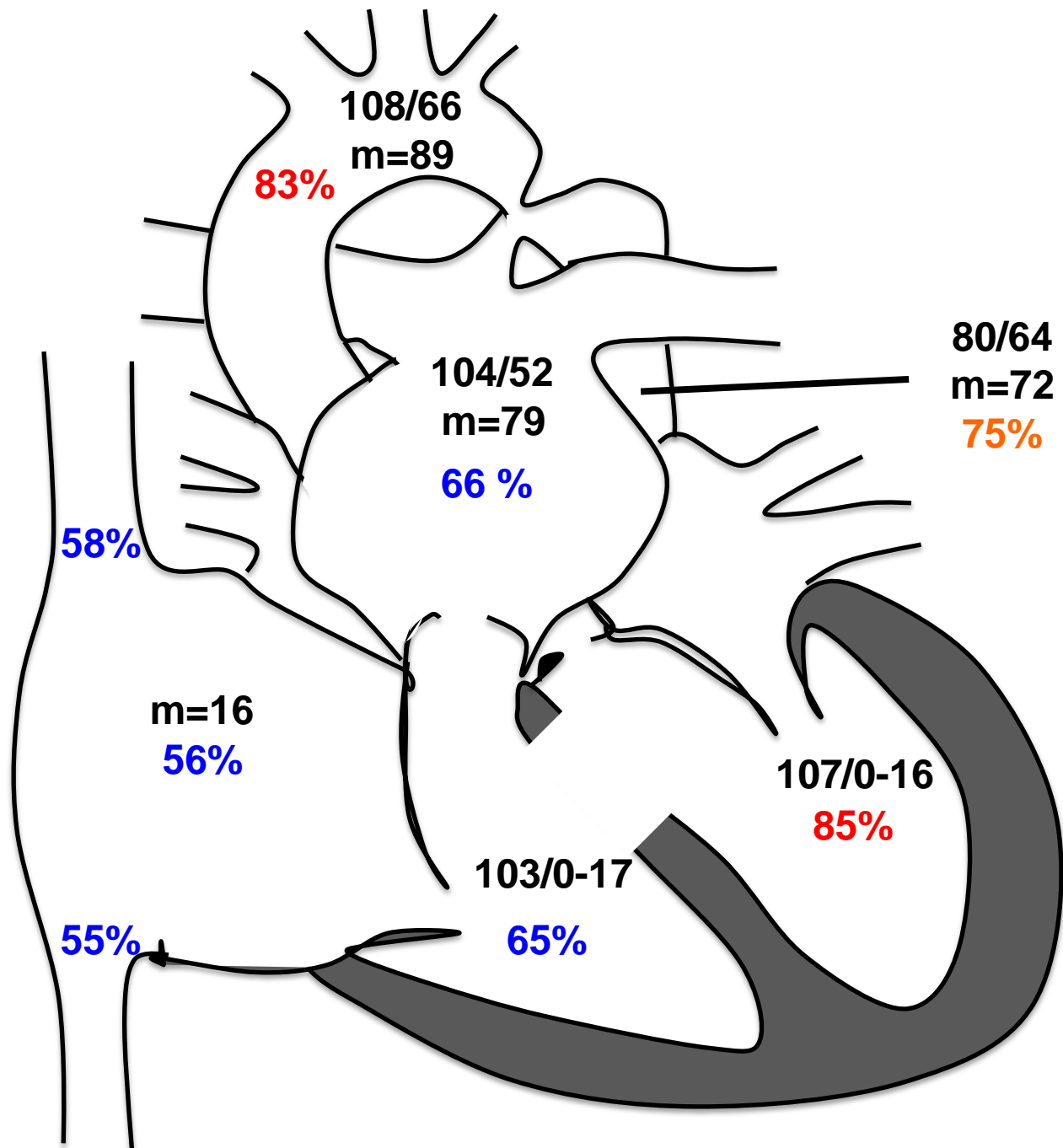
Problems and Risks

Problems

- Large ventricular shunt
- Very high PA pressure
- Huge MPA
- External compression of Lt main bronchus
- Hoarseness from Lt vocal cord paralysis
- Rt to Lt flow across PDA
- Severe juxta-ductal obstruction

Risks

- Pulmonary hypertensive crisis
- Rupture huge MPA
- Cardiac arrhythmia
- Sudden death
- Compromise airway



VSD and PDA are bidirectional shunting:

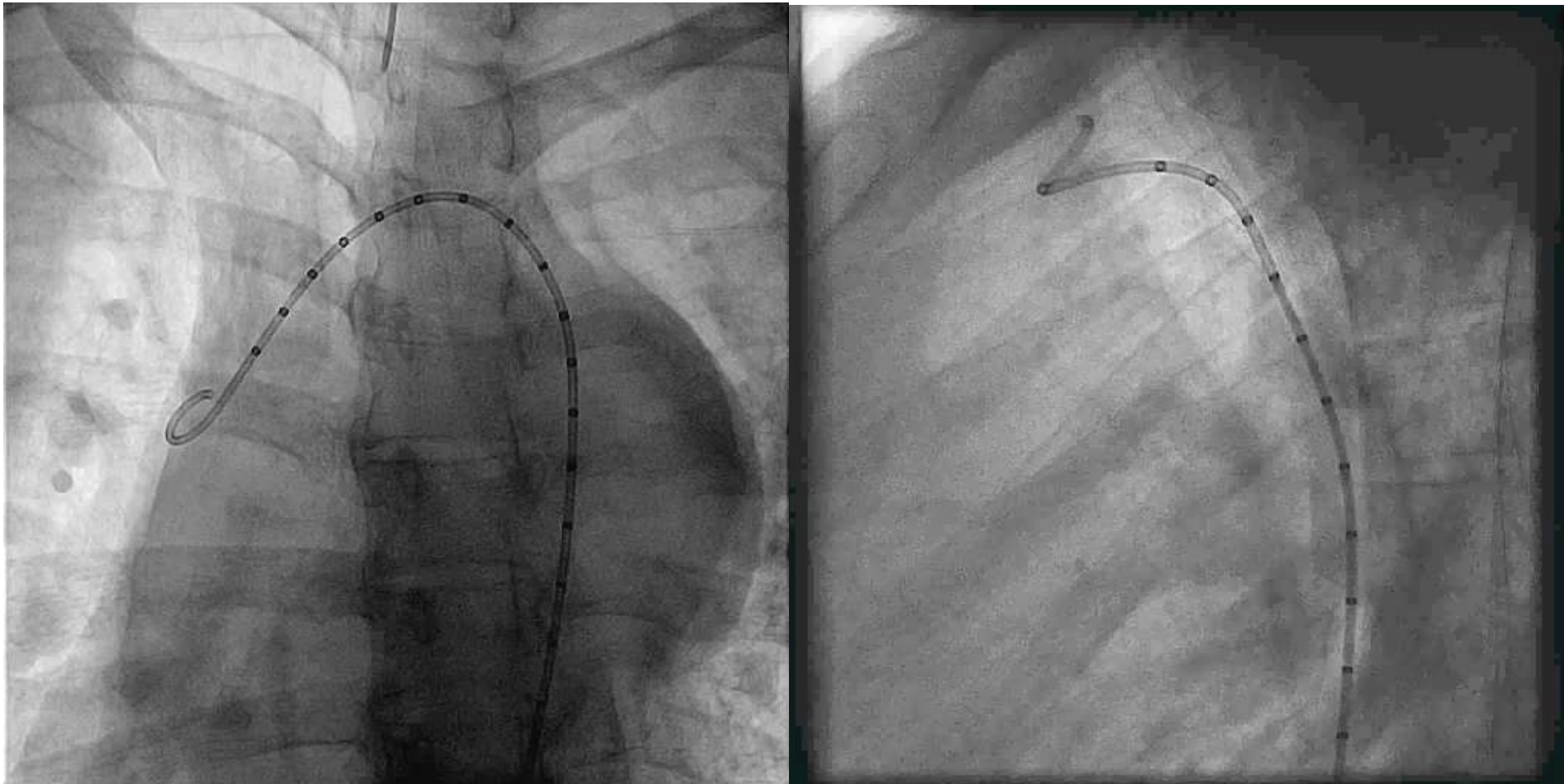
What should I do?



- 1. Leave her as she was**
- 2. Send for surgery: COAT repair and closure VSD**
- 3. Send for Heart-Lung transplantation**
- 4. Catheter or Surgical Intervention:**
 - Close COAT/PDA first, Close VSD later
- 5. Other possible options?**

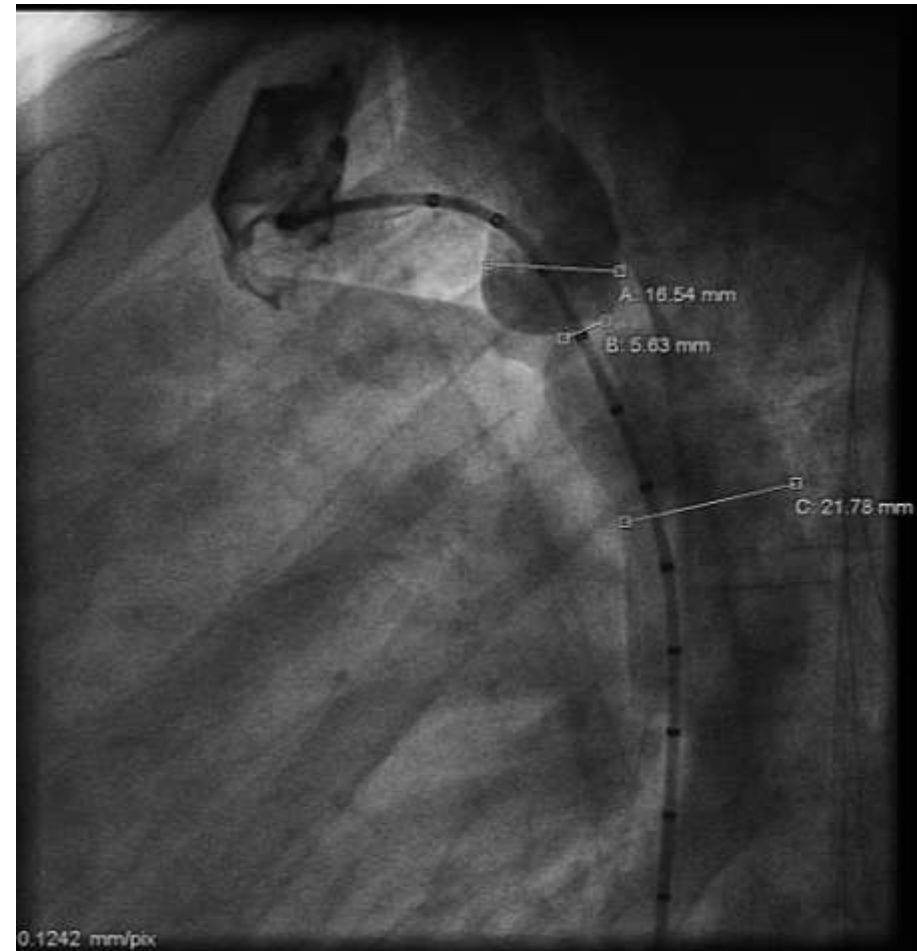
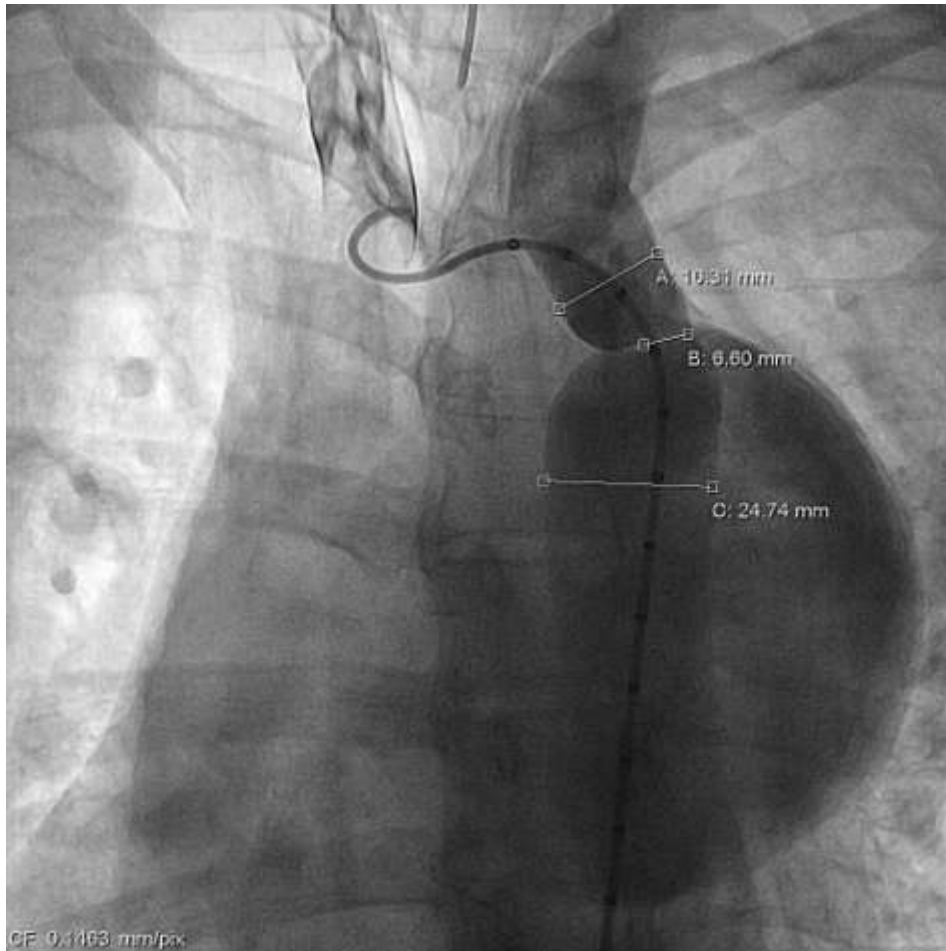
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Complex lesions: Large VSD,PDA,COAT



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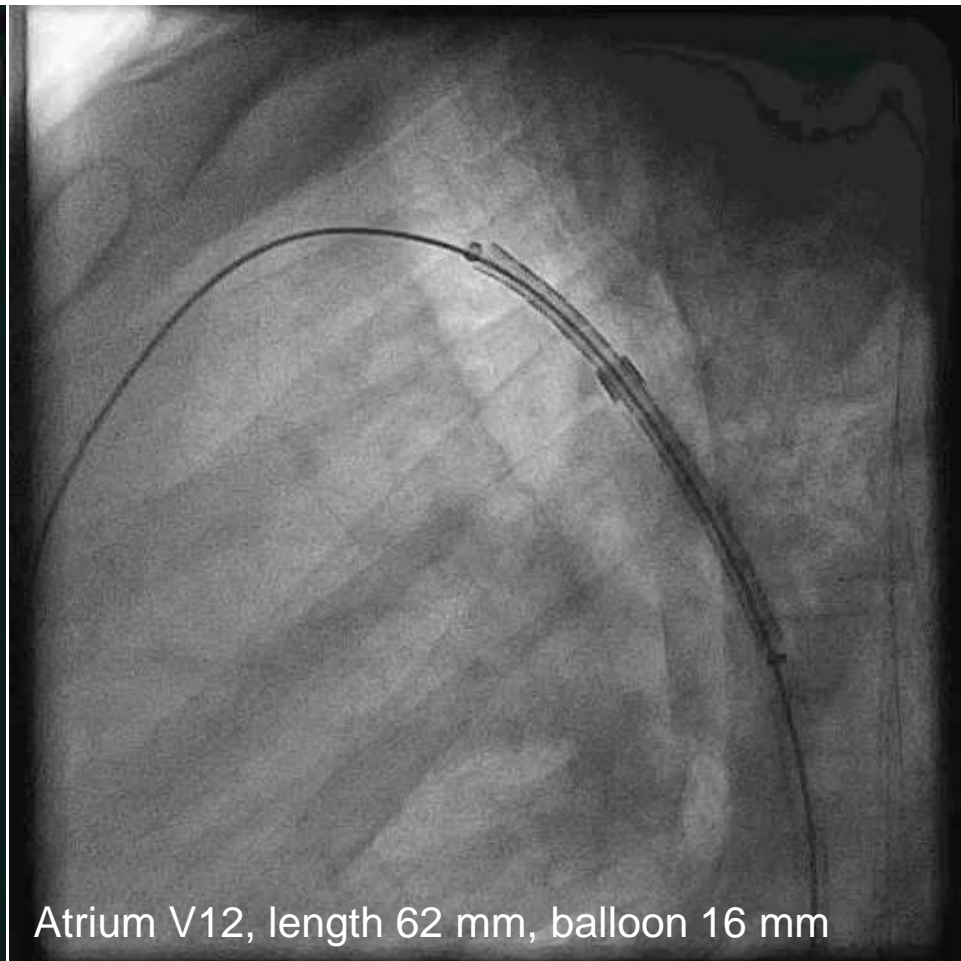
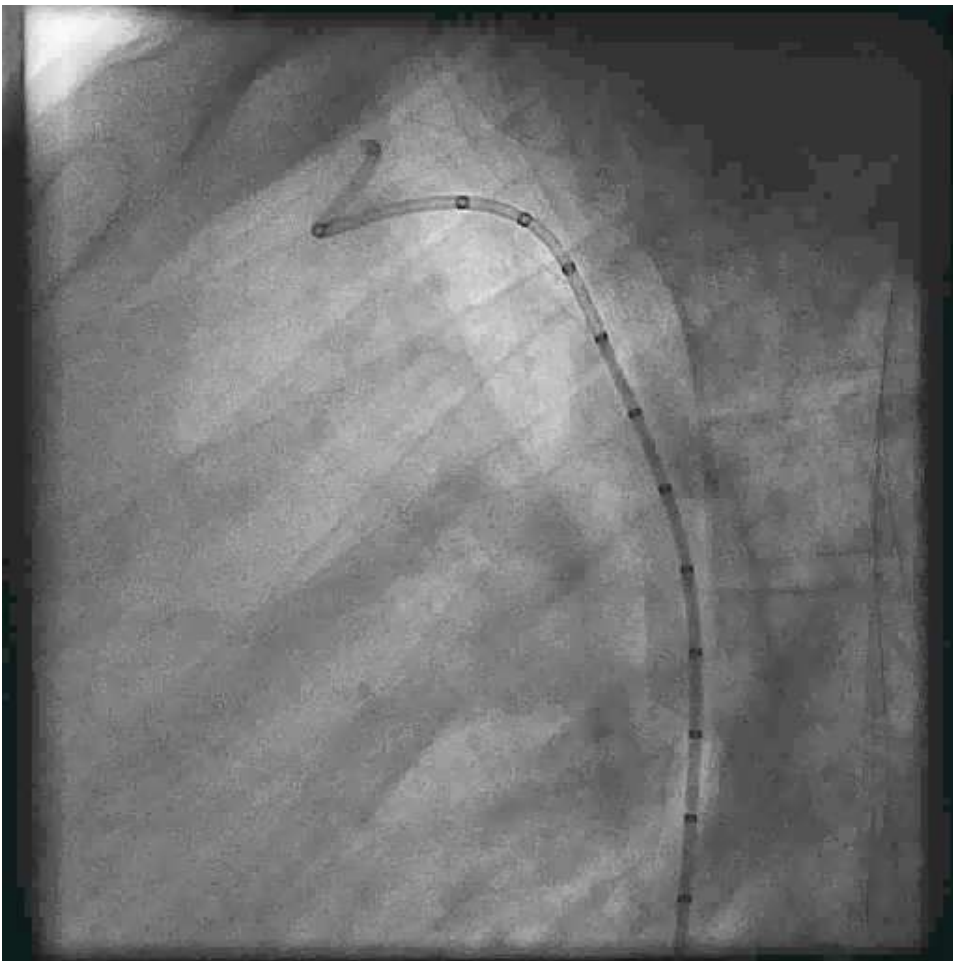


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What is (are) the right choice of device (s)?

- PDA device and bare stent.
- Covered stent.
- Other options.

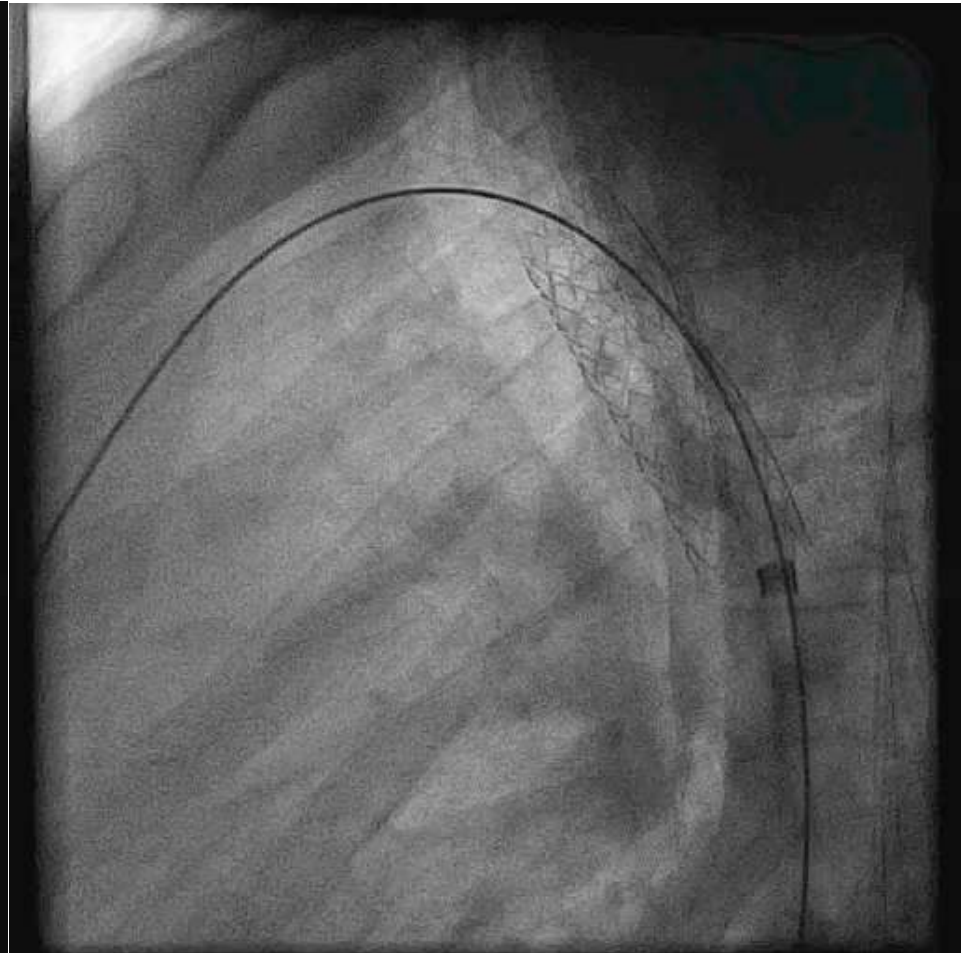
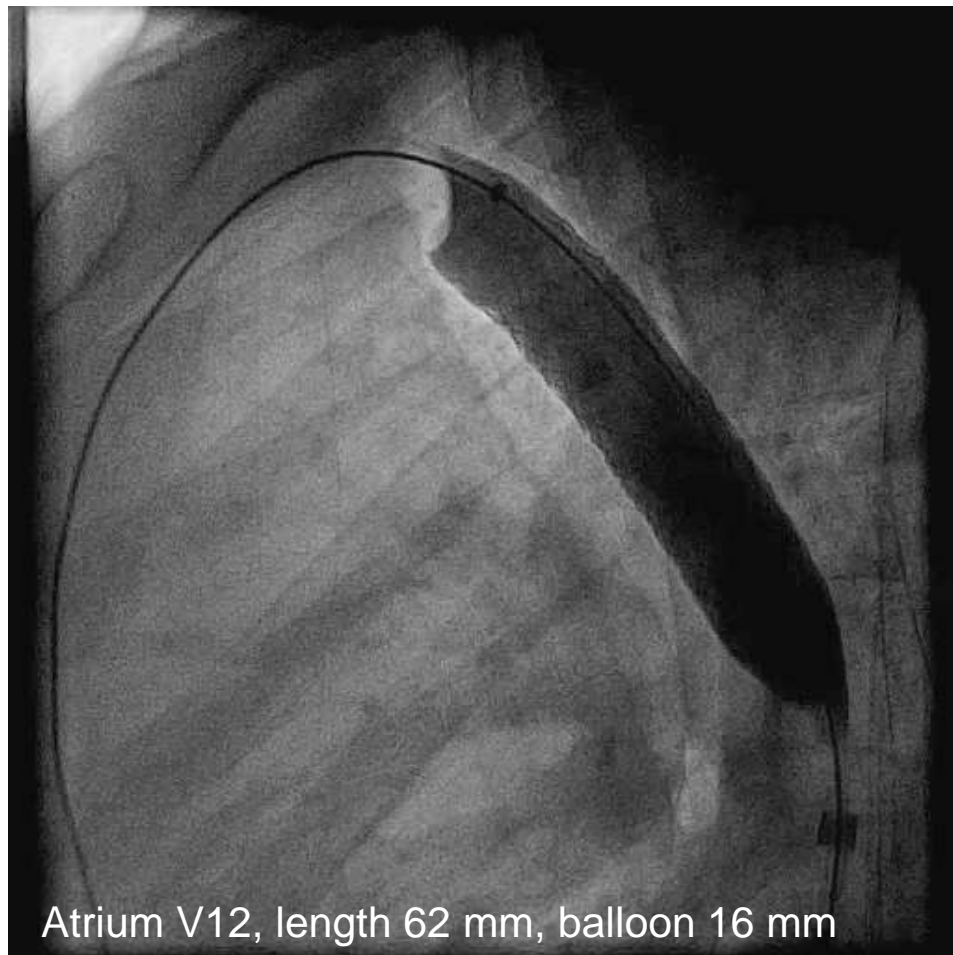
Complex lesions: Large VSD,PDA,COAT



Atrium V12, length 62 mm, balloon 16 mm

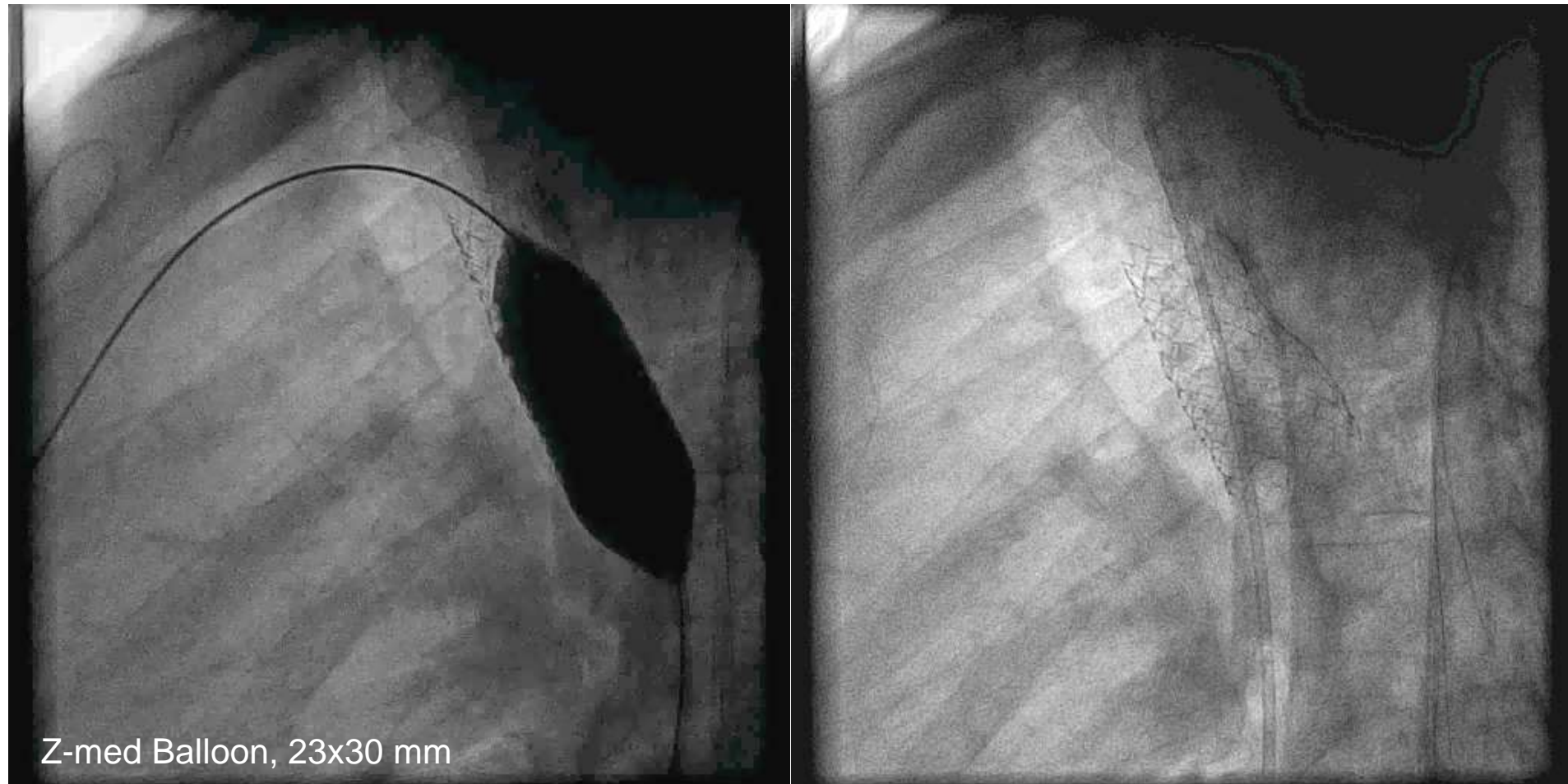
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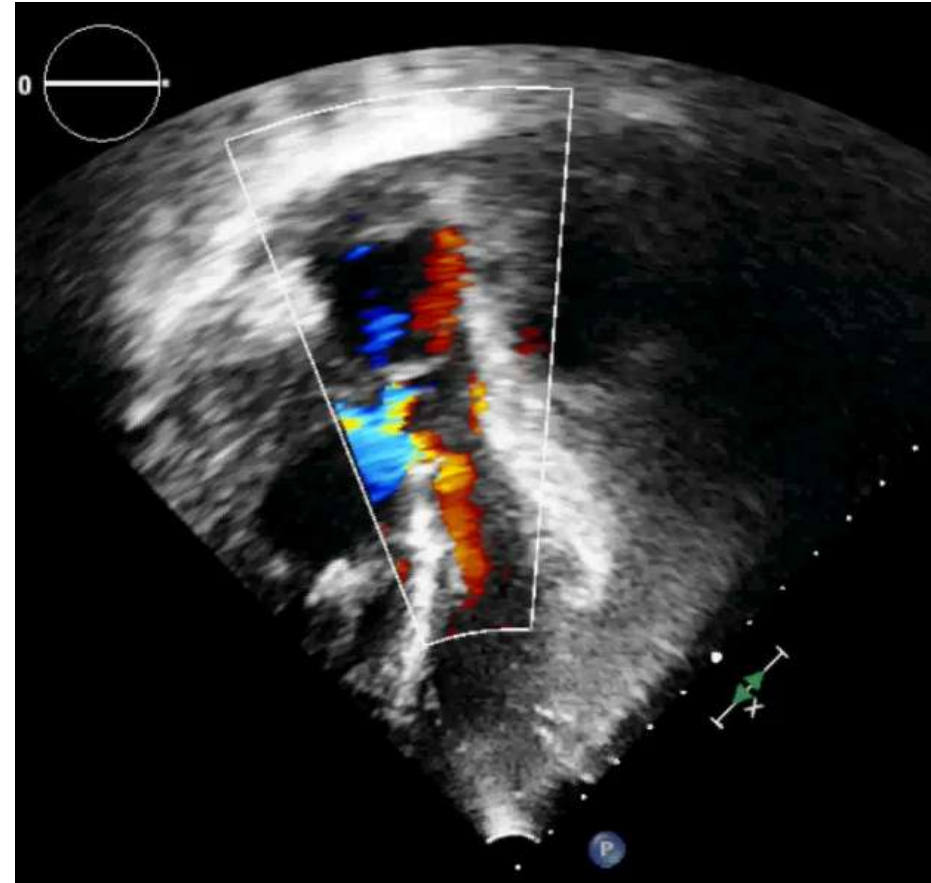
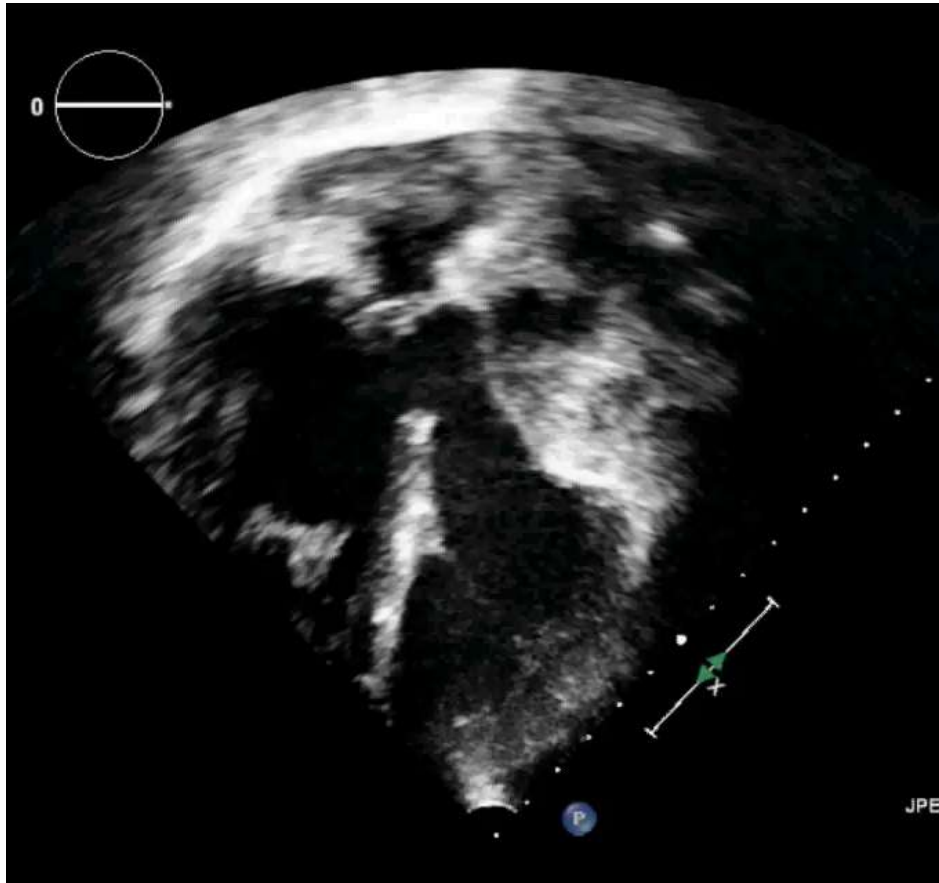


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Post Intervention

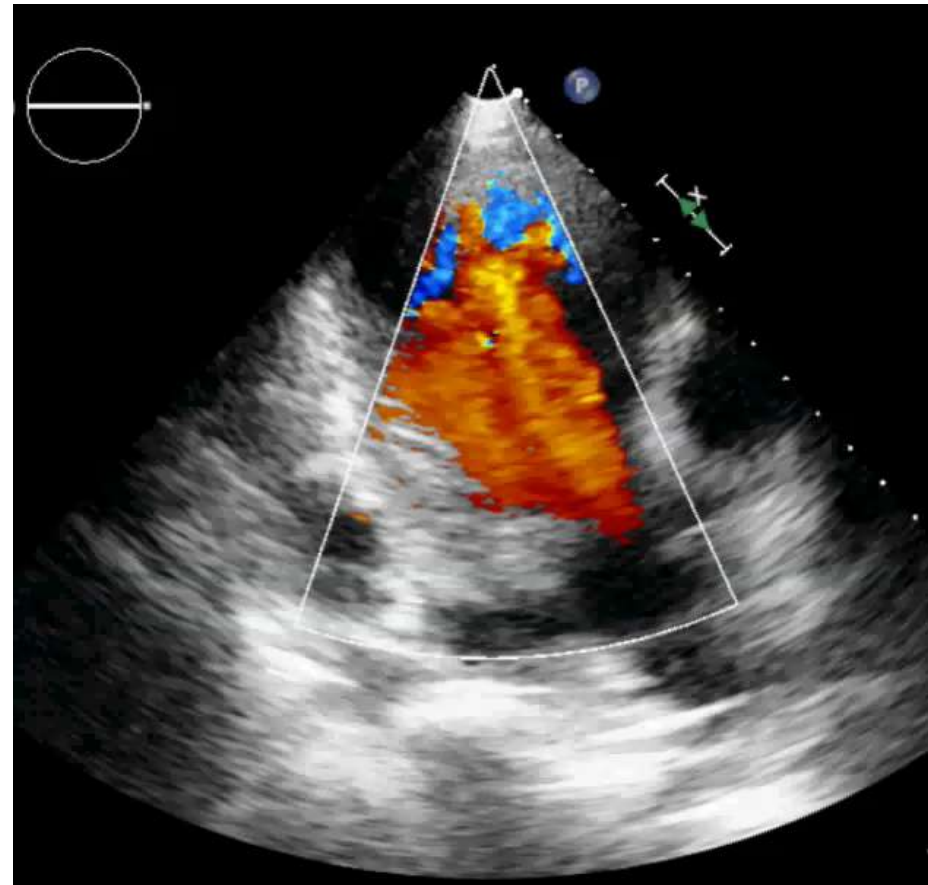
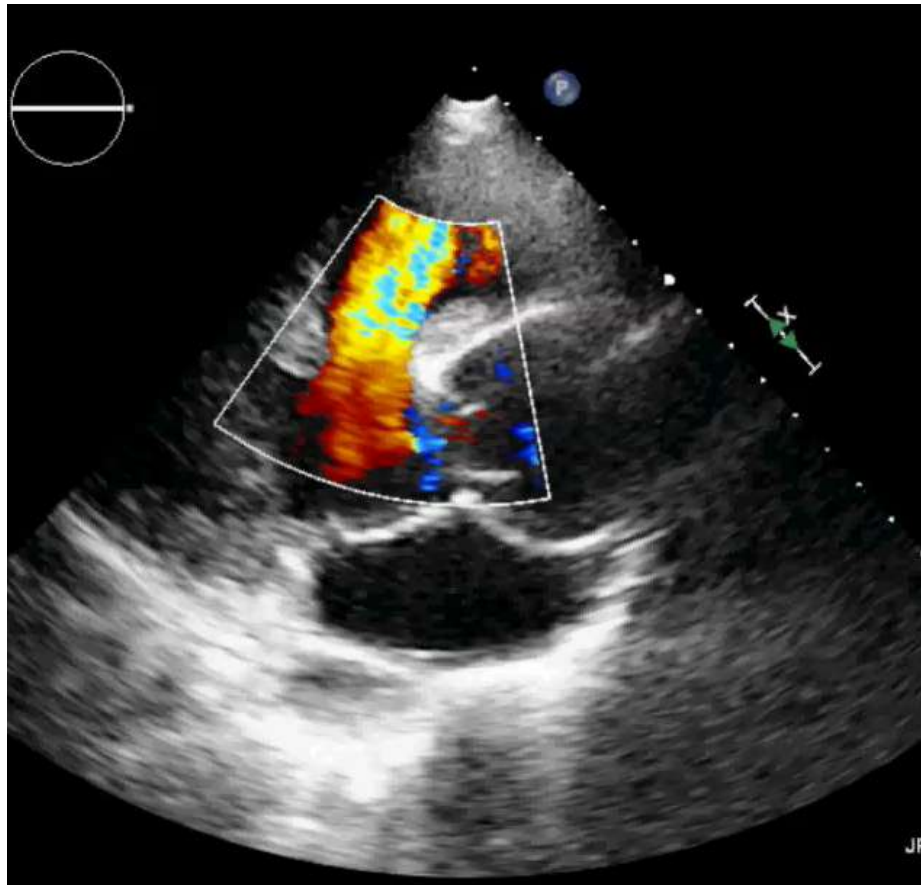
- **Compromised femoral artery.**
- **Developed persistent wheezing and Lt lung pneumonia.**
 - VSD more Rt. to Lt. at the first day ($SpO_2 \sim 78-80\%$ with $100\%O_2$)
 - Intubated for 6 days.
 - Antibiotic and bronchodilator for 10 days.
- **Still having severe PAH.**
 - Primacor IV infusion
 - Sildenafil 1.5 mg/kg/day
 - Inhaled iloprost q 2 hrs in the first
- **Discharge home 2 weeks after the procedure**
 - SpO_2 93% room air
 - Sildenafil, lanoxin and lasix

Echocardiogram – 5 months later



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Echocardiogram – 5 months later



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Plan

In the next few months:

- Reassess $Q_p:Q_s$, PVR, pulmonary vascular reactivity.
- Repeat CTA & CMR

Lesson Learned

Complex PAH-ACHD is a challenging situation.

- It's a long and winding road.
- Always have more than one option.
- Choose what “WE” (doctors-patient-parents) think it's the most suitable way for “US”.
- Every measure always have risk(s).
- Need a bit of luck (unfortunately)!

감사합니다

