



CASE STUDY ON

PULMONARY ARTERIAL HYPERTENSION (PAH)

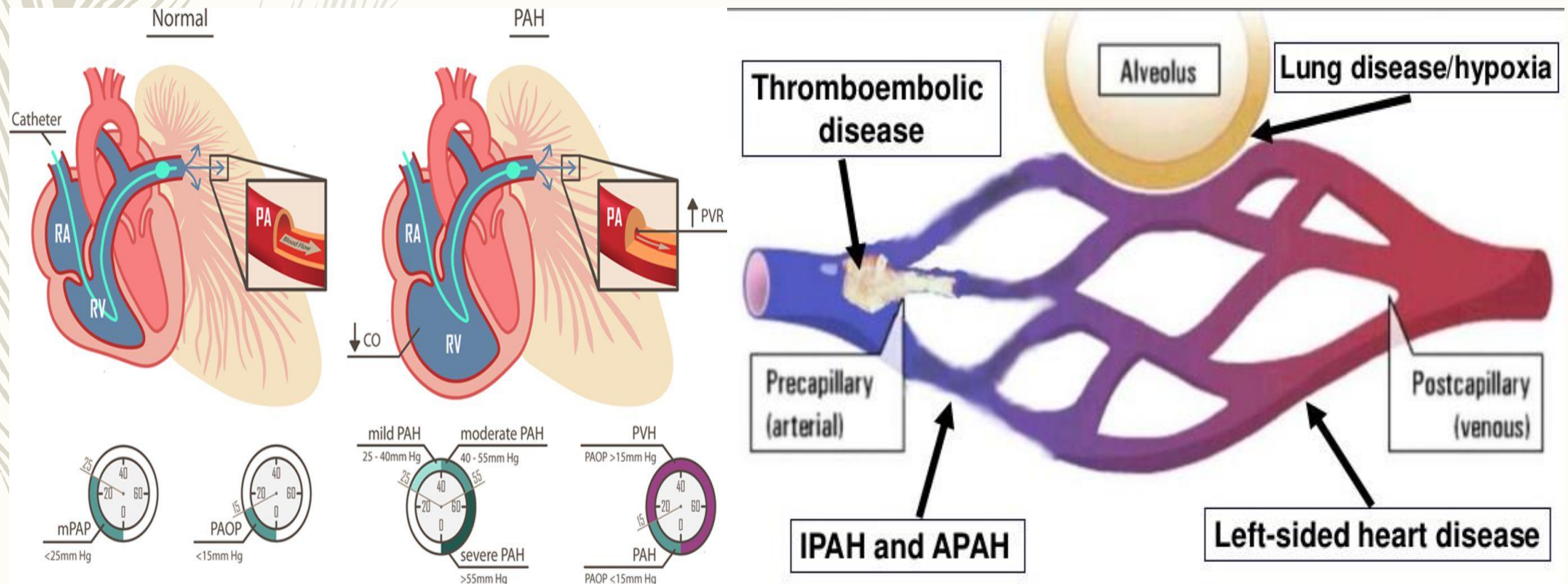
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INTRODUCTION

- Pulmonary arterial hypertension is a progressive disorder characterized by abnormally high blood pressure in the pulmonary artery, the blood vessel that carries blood from the heart to the lungs.
- It is said to be PAH if the mPAP ≥ 25 mmHg at rest or ≥ 30 mmHg with exercise and PWP < 15 mmHg.
- Hypertension occurs when most of the very small arteries throughout the lungs narrow in diameter, which increases the resistance to blood flow through the lungs. To overcome the increased resistance, pressure increases in the pulmonary artery and in the heart chamber that pumps blood into the pulmonary artery.

- Shortness of breath (dyspnea) during exertion and fainting spells are the most common symptoms of pulmonary arterial hypertension.
- Other symptoms include dizziness, swelling (edema) of the ankles or legs, chest pain, and a racing pulse.



SUBJECTIVE DATA

- A 45 year old female patient presented to the emergency with complaints of Severe cough, palpitations, generalised weakness and difficulty in breathing during household work.

OBJECTIVE DATA

- The patient is a known case of pulmonary arterial hypertension with **complete hoarsening of voice** from past 10 years.
- The patient was on medication:
 - ✓ Viagra 50mgt.i.d,
 - ✓ Lasix 40mgb.d,
 - ✓ Aldactone 25mgb.d,
 - ✓ Warf 2mgo.d and Pantodac 40mgo.d.



On examination, the vital signs were:

Temperature: Afebrile

Blood pressure: 130/80mmHg

RBS: 124mg/dl

Pulse: 90beats/min.

Respiratory rate: 25/min

CVS: S1S2+

SP_O₂: 88%



The physician ordered the following examinations:

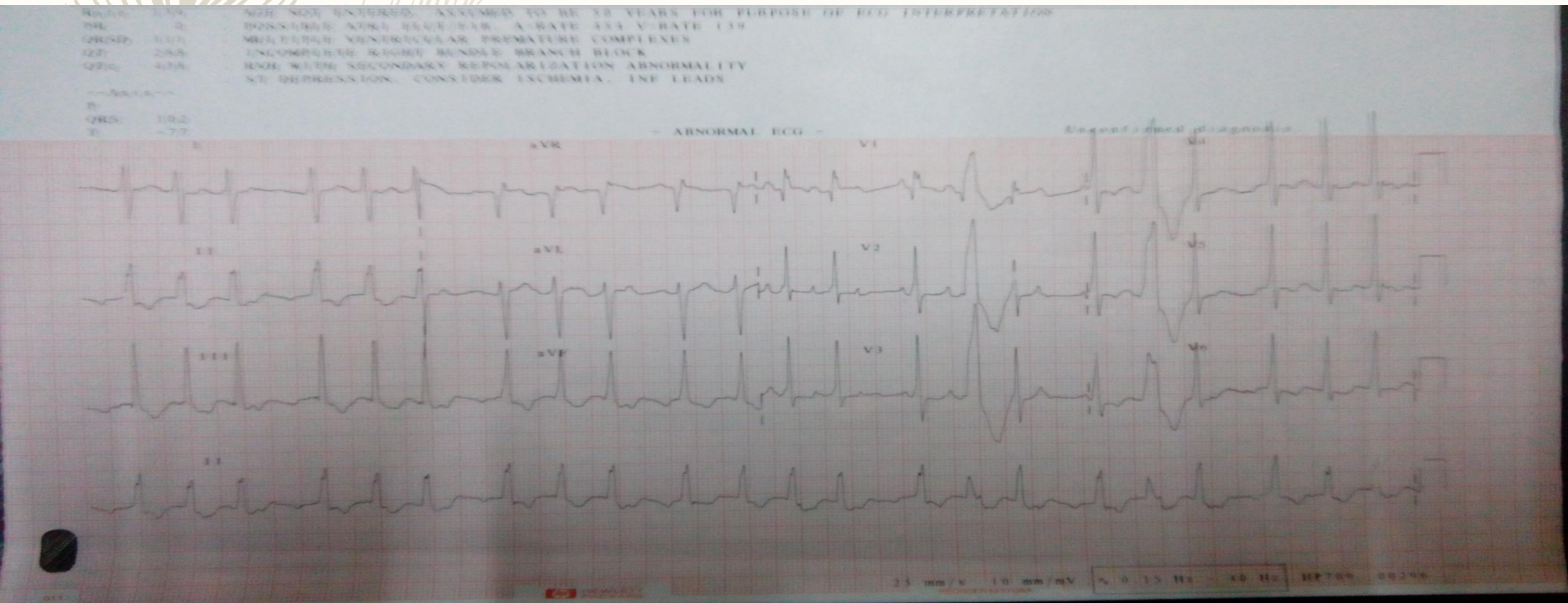
Complete Blood Picture:

Normal

Erythrocyte Sedimentation Rate: Normal

Possible Atrial Fibrillation
Multiple ventricular premature complexes
Incomplete right bundle branch block
RVH with secondary ST depression

ST depression



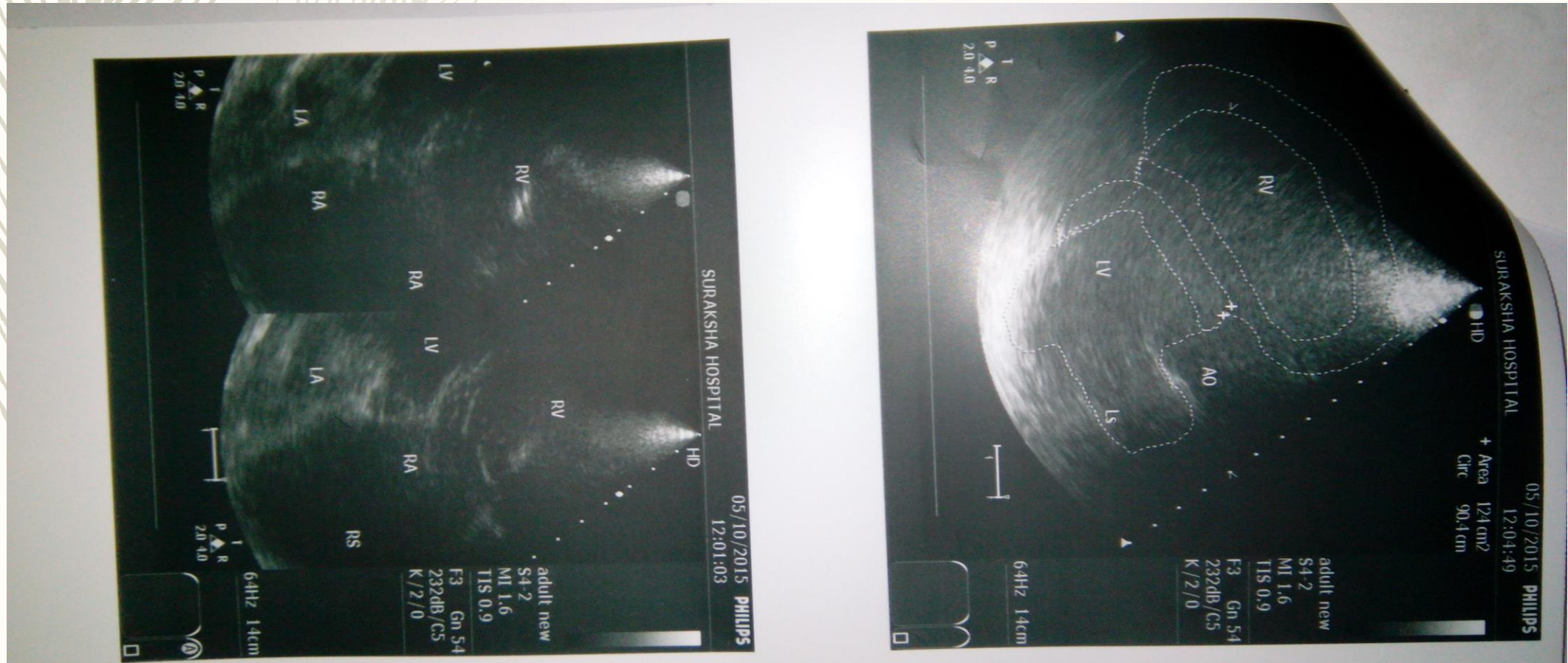
2D-Echo showed:

RA/RV/LA dilatation.

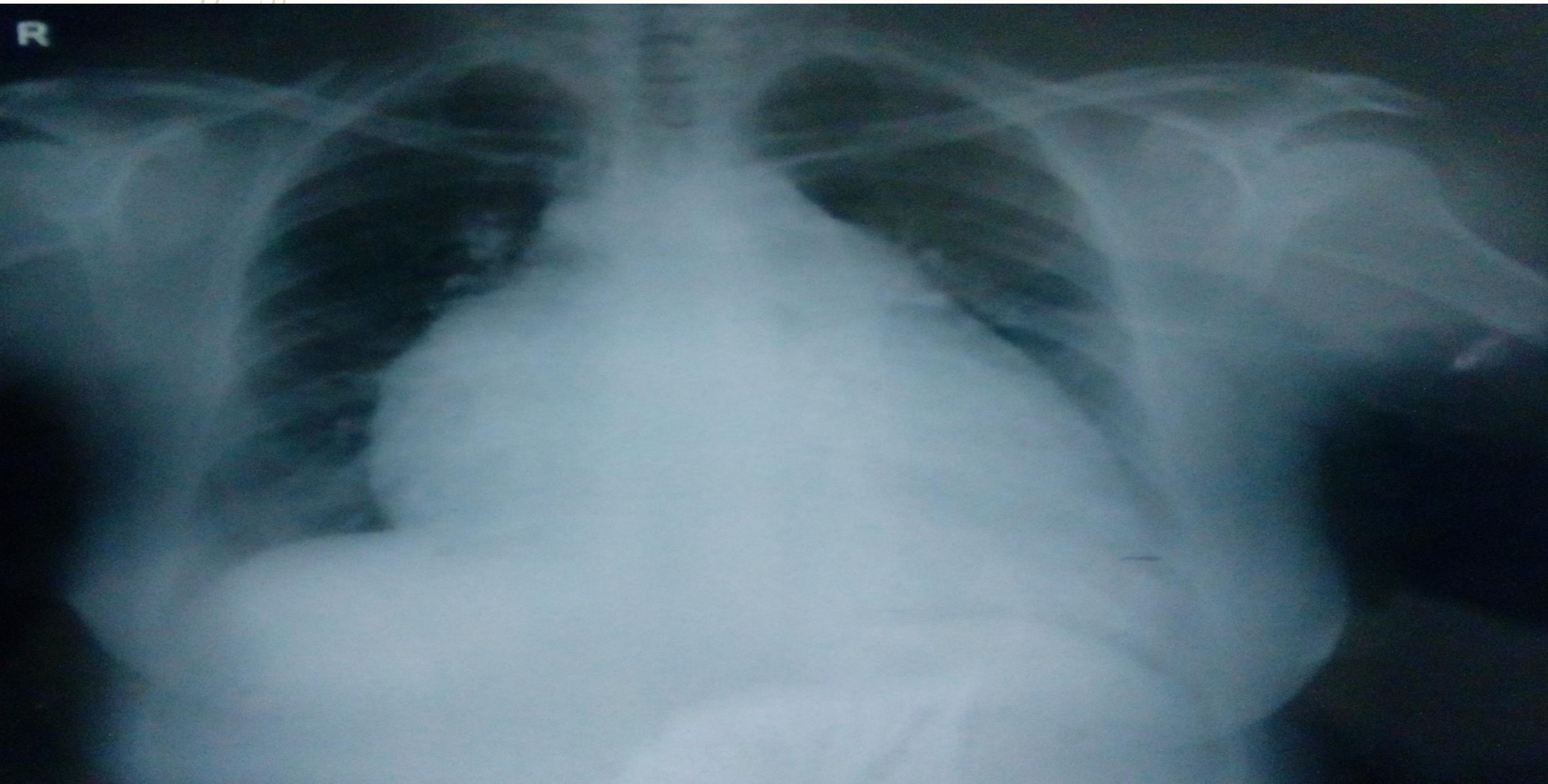
paradoxical motion of IVS

MPA, LPA, RPA enlarged.

EF=65%



X-ray showed: Cardiomegaly





ASSESSMENT DATA



- The above examinations were suggestive of pulmonary arterial hypertension(PAH)
- Apart from the past medications, Pulmonext 5mgo.d, lanoxin 0.25mgo.d were added and the sildenafil tablet was maintained at 20mgt.i.d.
- The patient was kept on oxygen for a week since her partial pressure was not contained.

STANDARD THERAPY FOR PAH


- Currently, there are four main classes of medications used to treat PAH,

Calcium-channel blockers	Phosphodiesterase type 5 inhibitors	Endothelin receptor antagonists	Prostacyclin analogues	Anticoagulant
Amlodipine Diltiazem Nifedipine	Sildenafil Tadalafil	Bosentan Ambrisentan *	Epoprostenol Iloprost Treprostinil*	Warfarin sodium

TREATMENT PLAN

DRUG(GENERIC NAME)	DOSE	MECHANISM OF ACTION	SIDE EFFECTS
ASSURANCE (SILDENAFIL CITRATE)	20MG T.I.D 	Sildenafil is a selective potent inhibitor of PDE type 5 which specifically degrades cGMP which is found in high concentrations in pulmonary arteries.	headache, flushing, dyspepsia, abnormal vision.
PULMONEXT (AMBRISENTAN) Approved June 2007 Approved for combination therapy with tadalafil in 2015.	5MG O.D 	ambrisentan) is a potent type-A selective endothelin receptor antagonist. Endothelin is a peptide made by the body in the endothelium	Headache, peripheral edema.

<p>LASIX(FUROSEMIDE)</p>	<p>40MG O.D.</p> 	<p>Diuretics are commonly used to help treat heart failure. Diuretics help eliminate excess fluid in the body</p>	<p>deep venous thrombosis, hyponatremia.</p>
<p>ALDACTONE (SPIRONOLACTONE)</p>	<p>25MG B.D.</p> 	<p>Spironolactone is a specific antagonist of aldosterone, that increased amounts of sodium and water to be excreted.</p>	<p>Hyperkalemia, hyponatremia.</p>
<p>WARF (WARFARIN SODIUM)</p>	<p>2.5MG O.D.</p> 	<p>Warfarin is an anticoagulant, which is given to allow free flow of blood.</p>	<p>Alopecia, haemorrhage.</p>



TOLVASKA (TOLVAPTAN)	15MG O.D/S.O.S	Tolvaptan is a vasopressin antagonist that causes increased water excretion with low sodium excretion,	thirst, dry mouth, asthenia, constipation.
PANTODAC (PANTOPRAZOLE)	40MG O.D.	It is a proton pump inhibitor that reduces gastric acid secretion.	Abdominal pain, nausea, headache.

RECENT DRUG DEVELOPMENT

- **Riociguat (Adempas®)**, which acts on the nitric oxide pathway and further downstream as a direct guanylate cyclase stimulator, as well as sensitizing the receptor to endogenous nitric oxide. Because it acts on the nitric oxide pathway, it should not be given with phosphodiesterase-5 (PDE-5) inhibitors. It was developed by Bayer In 2014.
- **Macitentan (Opsumit®)** which is an endothelin receptor antagonist (ERA). It was developed by actelion in 2013.
- **Treprostinil**, marketed as Orenitram™ is a prostacyclin analogue which is a vasodilator.
- Considerable additional research is needed to better refine the most appropriate therapies for individuals with the pulmonary arterial hypertension.



Thank you for
joining us!