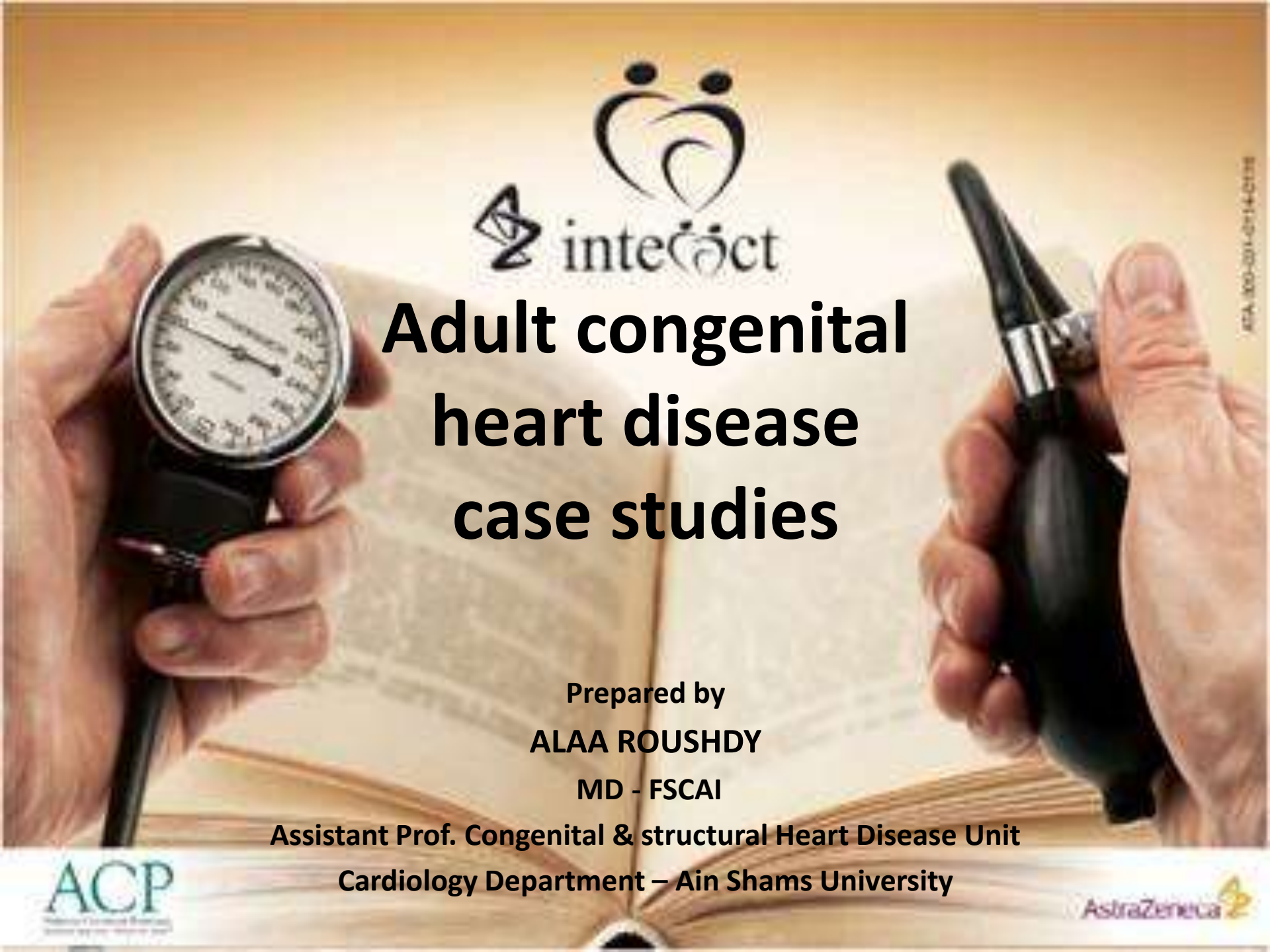


Adult congenital heart disease case studies

Prepared by

ALAA ROUSHDY

MD - FSCAI

Assistant Prof. Congenital & structural Heart Disease Unit

Cardiology Department – Ain Shams University

What is GUCH ??

- The term “grown up congenital heart disease or GUCH” refers to the group of patients who passes from the childhood period to adolescence and adulthood having some form of congenital heart disease whether repaired or non repaired by surgery or catheterization.
- Sometimes it can be referred to as “Adult Congenital Heart Disease”

- GUCH patients can broadly be divided into three categories:

1. Those who have undergone some sort of reparative operation
2. Those who have undergone a palliative Operation
3. A group of patients who have not previously undergone a corrective or palliative procedure.

Scale of the problem

- Remarkable improvement in survival of patients with congenital heart disease (CHD) has occurred over the past half century since reparative surgery has become commonplace.
- Since the advent of neonatal repair of complex lesions in the 1970s, an estimated 85% of patients survive into adult life.

Scale of the problem

- Given modern surgical mortality rates of less than 5%, one would expect that in the next decade, almost 1 in 150 young adults will have some form of CHD.
- In particular, there are a substantial number of young adults with single-ventricle physiology, systemic right ventricles (RVs), or complex intra cardiac baffles who are now entering adult life and starting families.

Clinical presentation

- Asymptomatic
- Symptoms of heart failure
- Manifestations of pulmonary hypertension
- Low cardiac output manifestations
- Manifestations of cyanotic heart disease
- Arrhythmias
- Associated conditions
- Complications

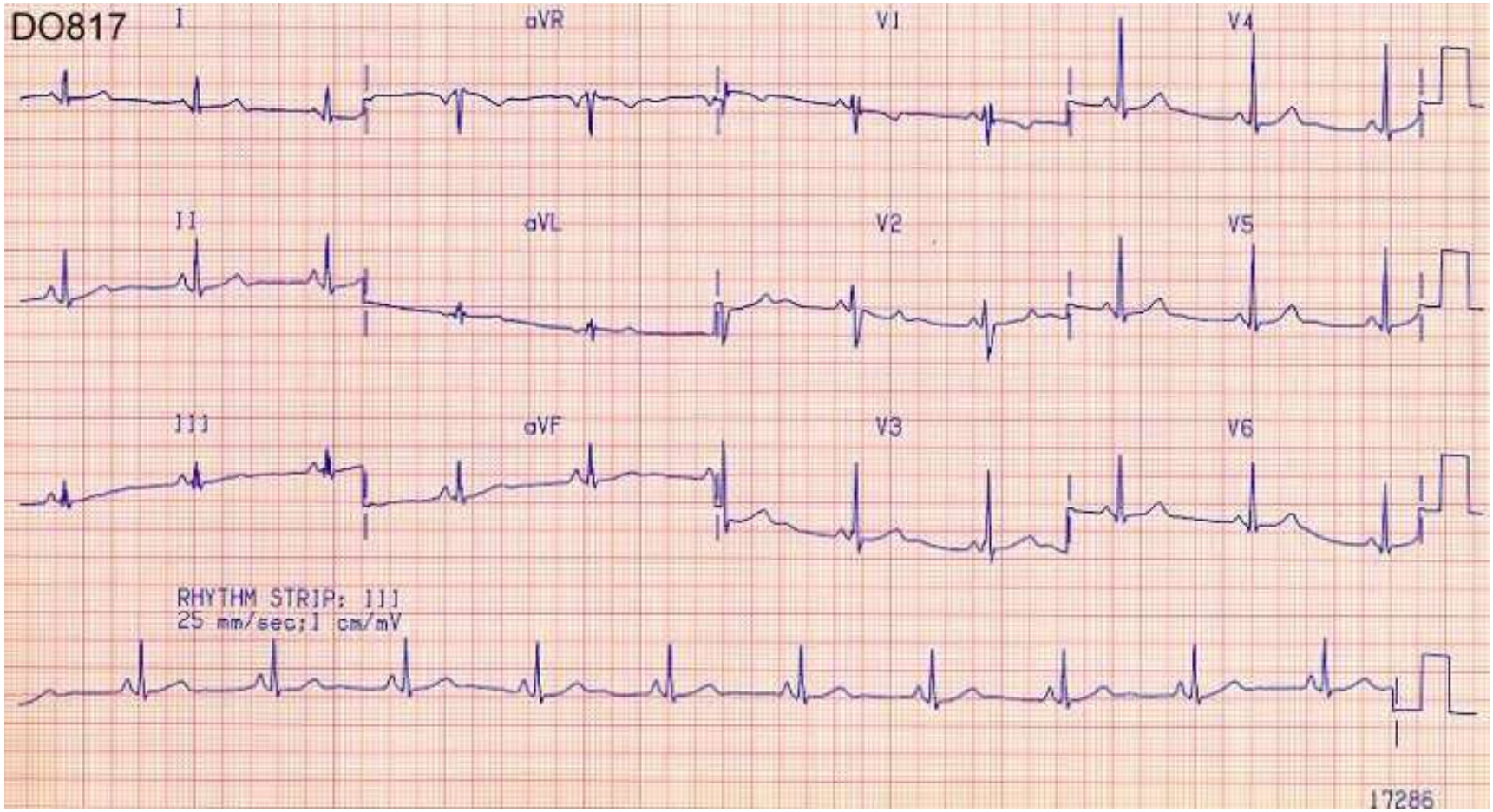
Adult congenital heart disease interactive case presentation 1

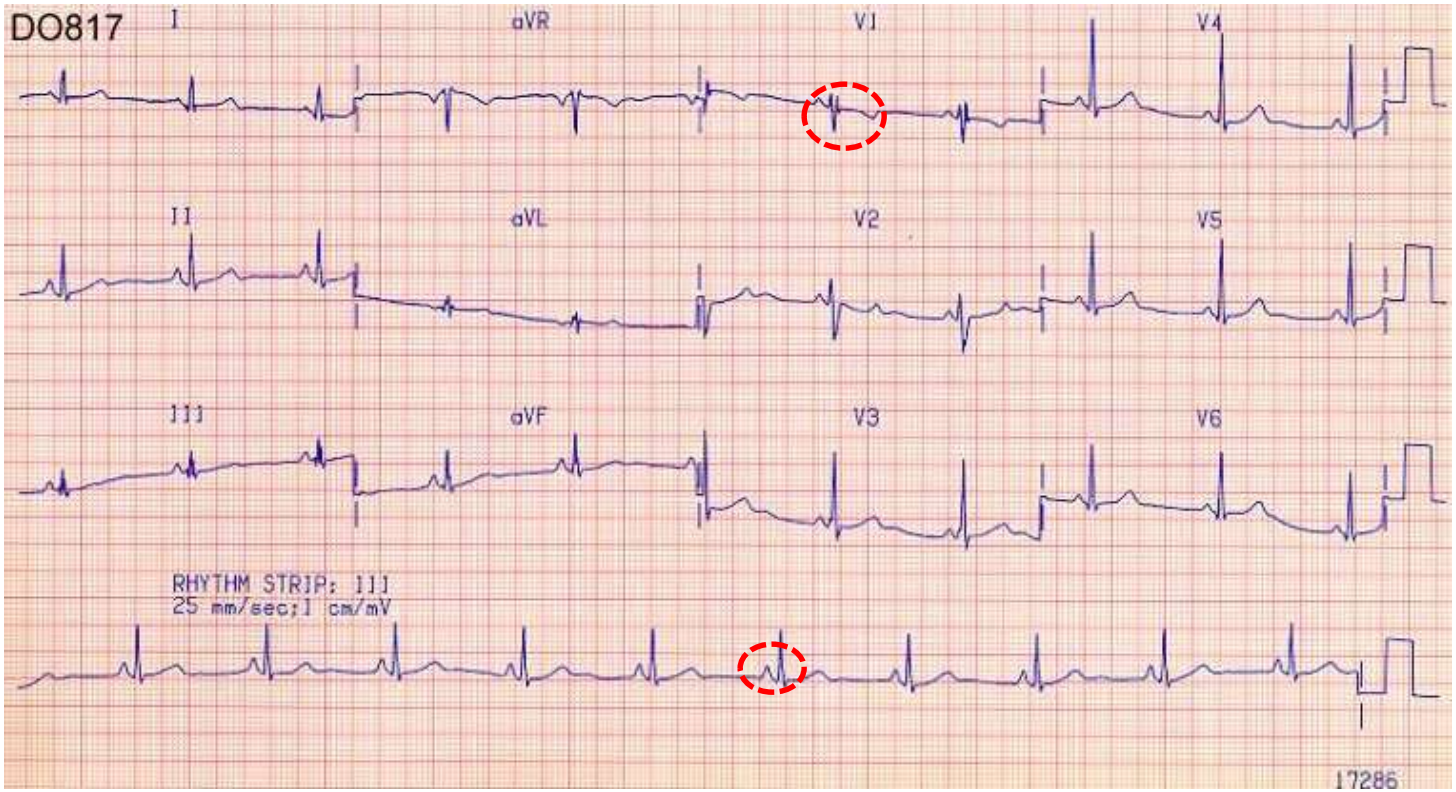
Case 1

- Fatma is a 25 years old female patient married 5 years ago and has 2 children the youngest is 2 years old.
- She used to complain of shortness of breath on severe exertion since she was 18 years old
- She also reports atypical chest pain, headache and easy fatigability of the same duration

- She sought medical advice and lab investigations were done as well as ECG and Chest x ray 3 years ago.
- Her CBC showed HB of 10 g%
- Her ECG and CXR were as follows

ECG


DO817
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
RHYTHM STRIP: III
25 mm/sec;1 cm/mV
17286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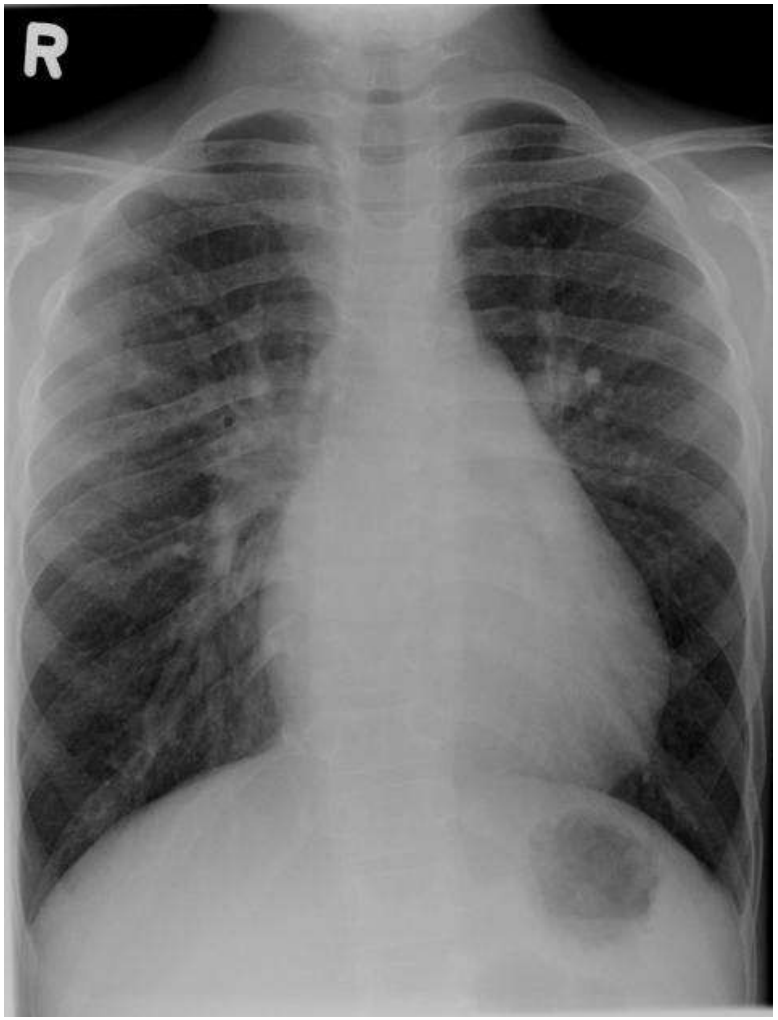
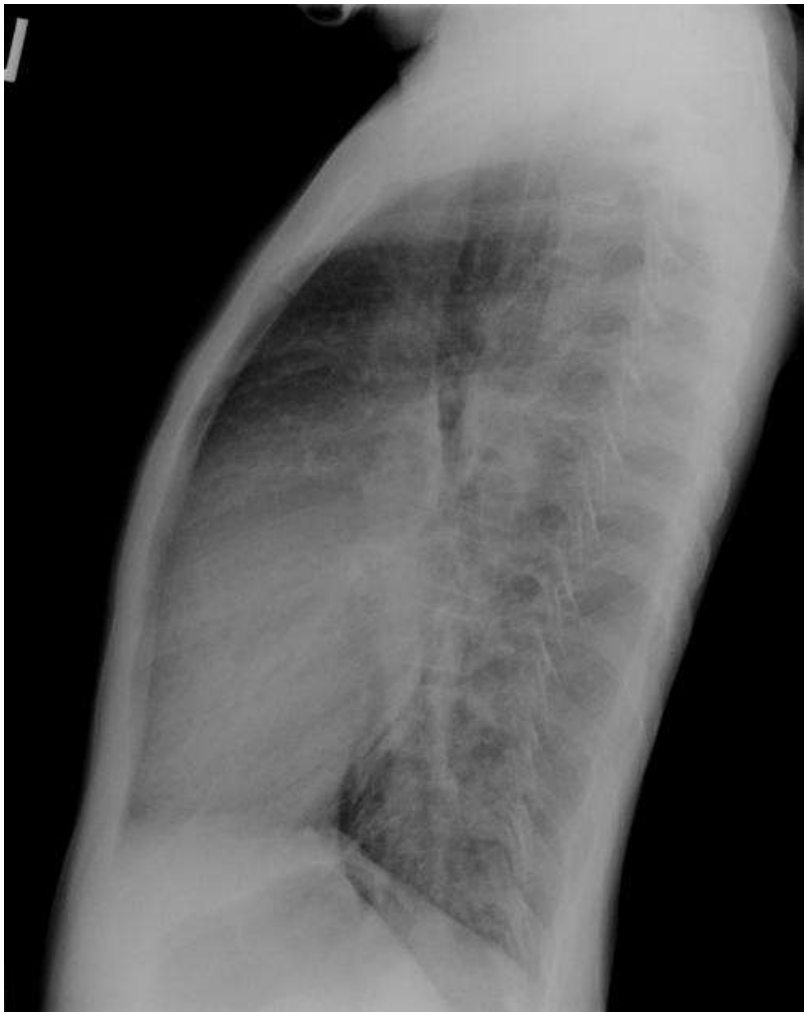
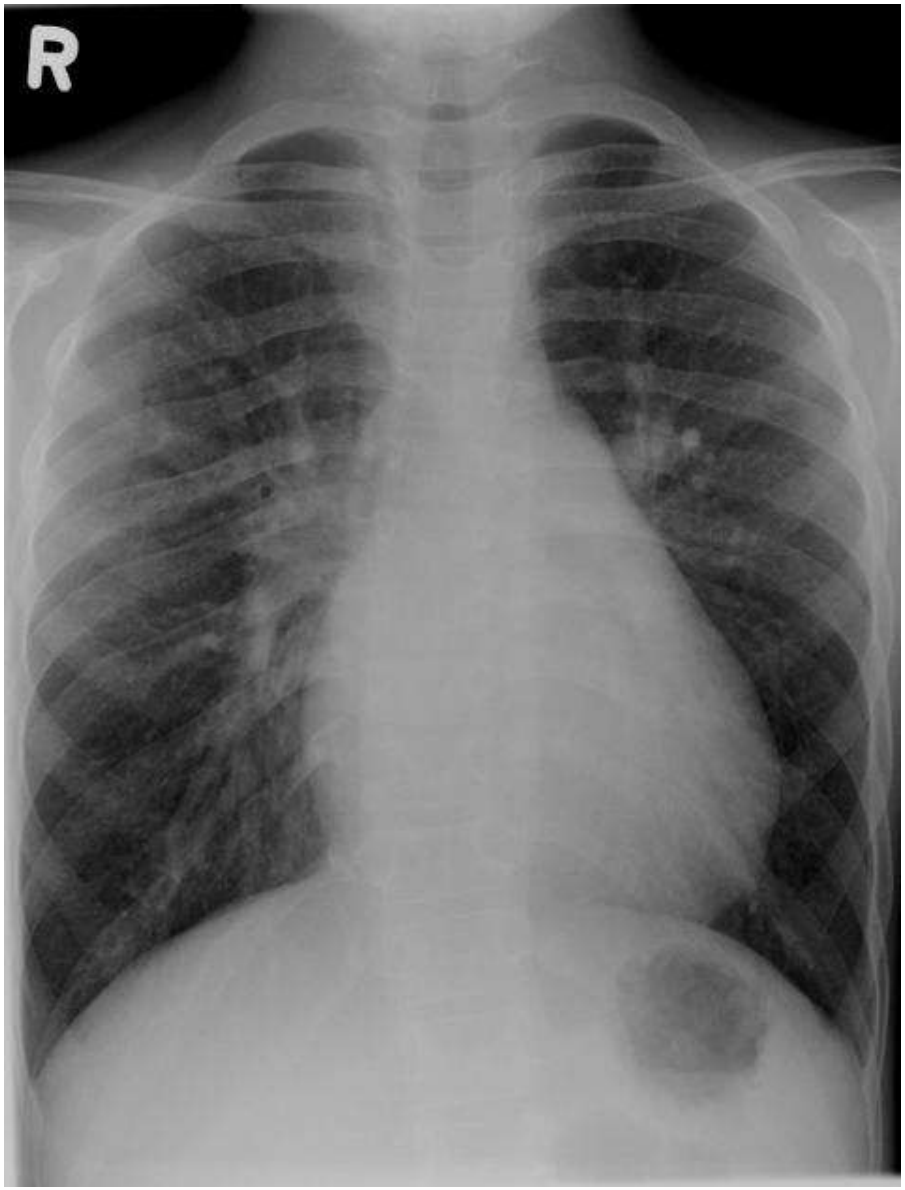
RHYTHM STRIP: III
25 mm/sec;1 cm/mV
17286

CXR






R

- Her physician attributed her complains to reduced Hb level and prescribed her vitamins and reassured her.
- He considered her ECG to be a normal variant and did not comment on the CXR.

What do you think?

- After her last pregnancy 2 years ago she started to complain of progressive dyspnea with NYHA class 2 and her easy fatigability increased as well
- Her recent CBC showed a Hb level of 10 g% and her other lab investigations was unremarkable

General examination

- She was alert conscious oriented to time place and person.
- Blood pressure 110/70
- Pulse 90 beat per minute equal on both sides and peripherally felt
- No pallor no jaundice no cyanoses
- No lower limb edema or congested neck veins

What do you think?

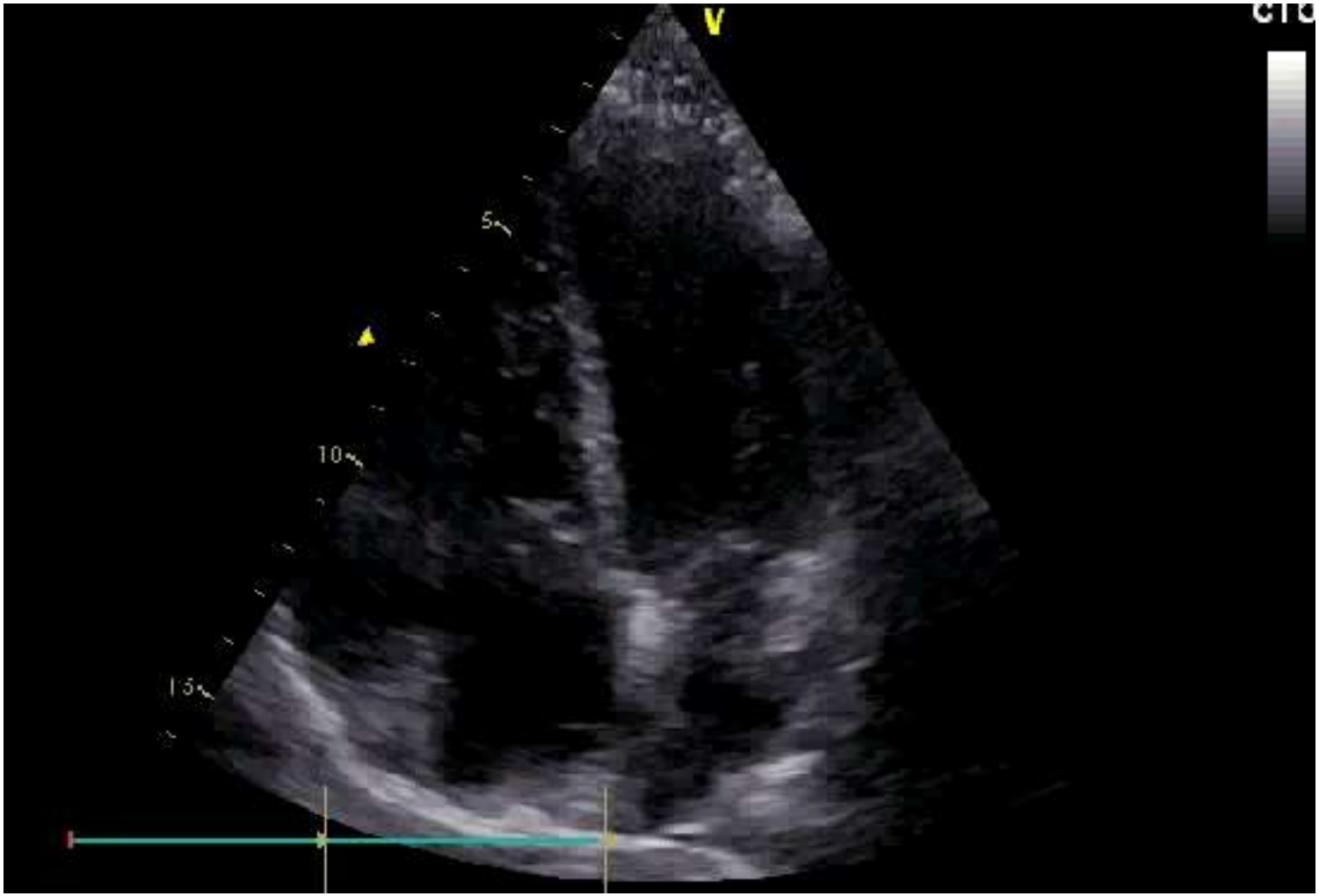
Local examination

- Normal S1
- Wide fixed splitting of S2 over the pulmonary area
- Soft systolic murmur grade 3 over the pulmonary area
- Short mid diastolic rumble over the tricuspid area

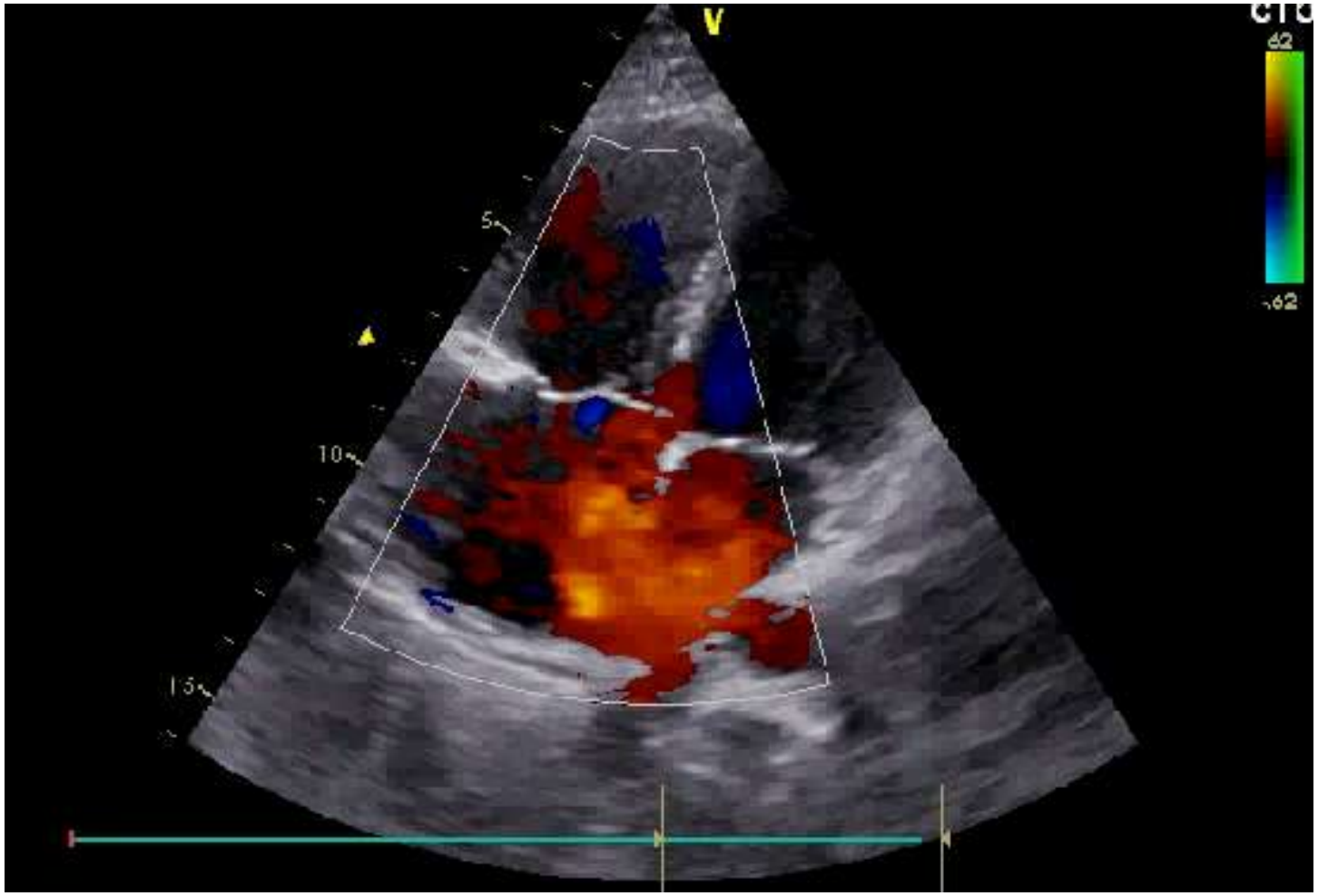
What is your probable diagnoses?

What is your next step?

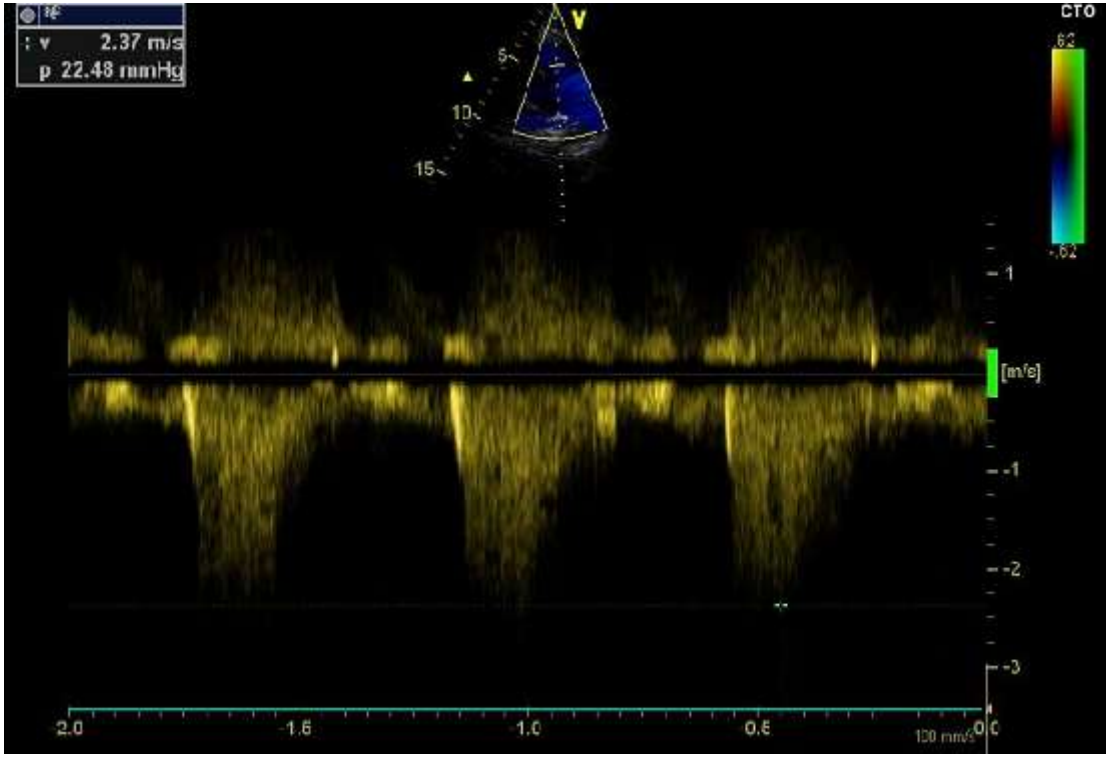
Echocardiogram



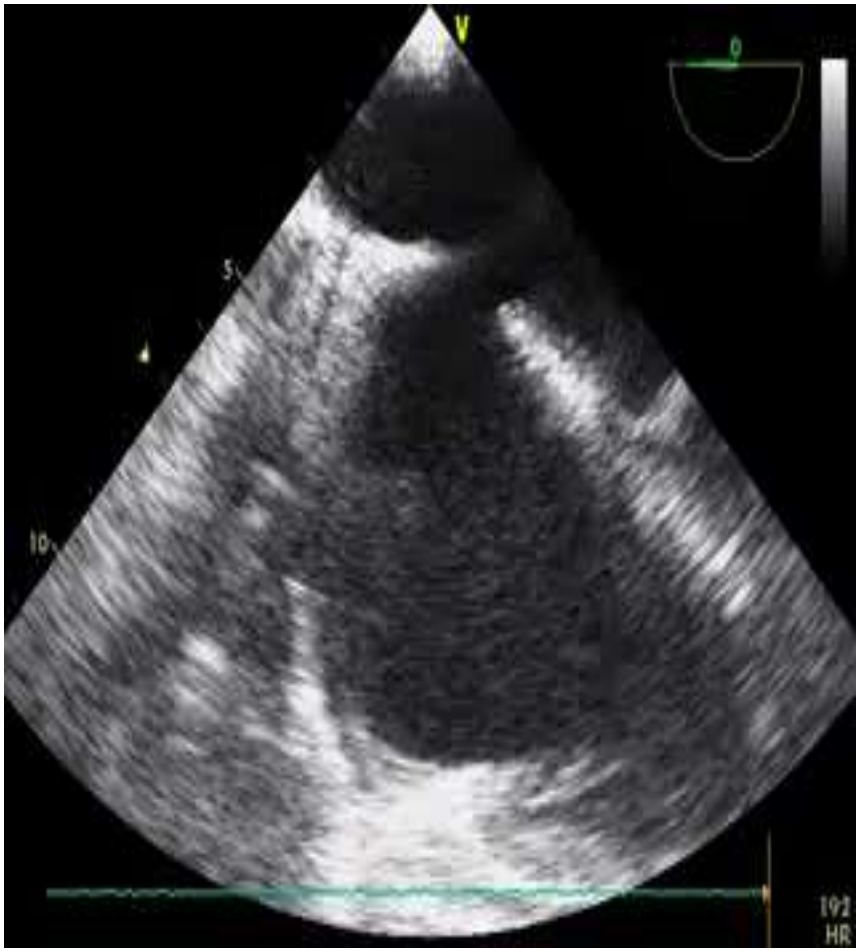
Echocardiogram

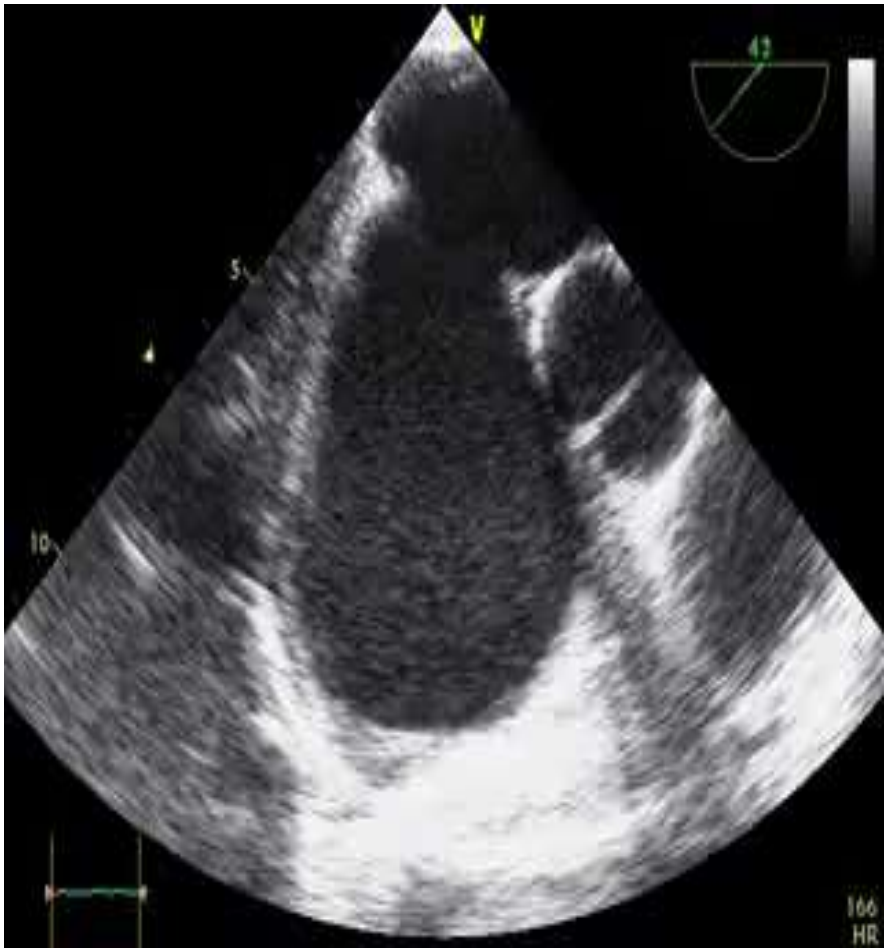


Echocardiogram

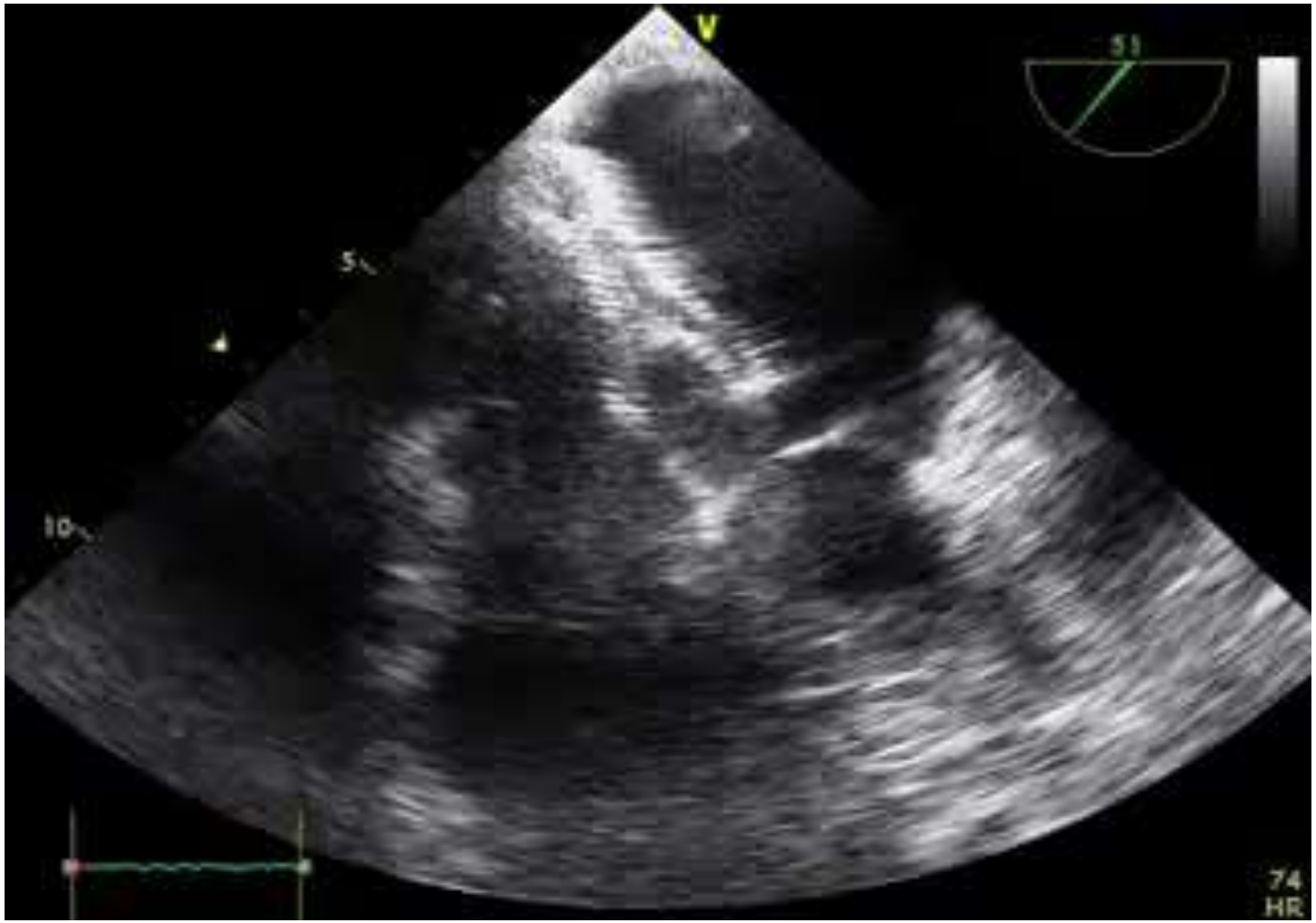


TEE

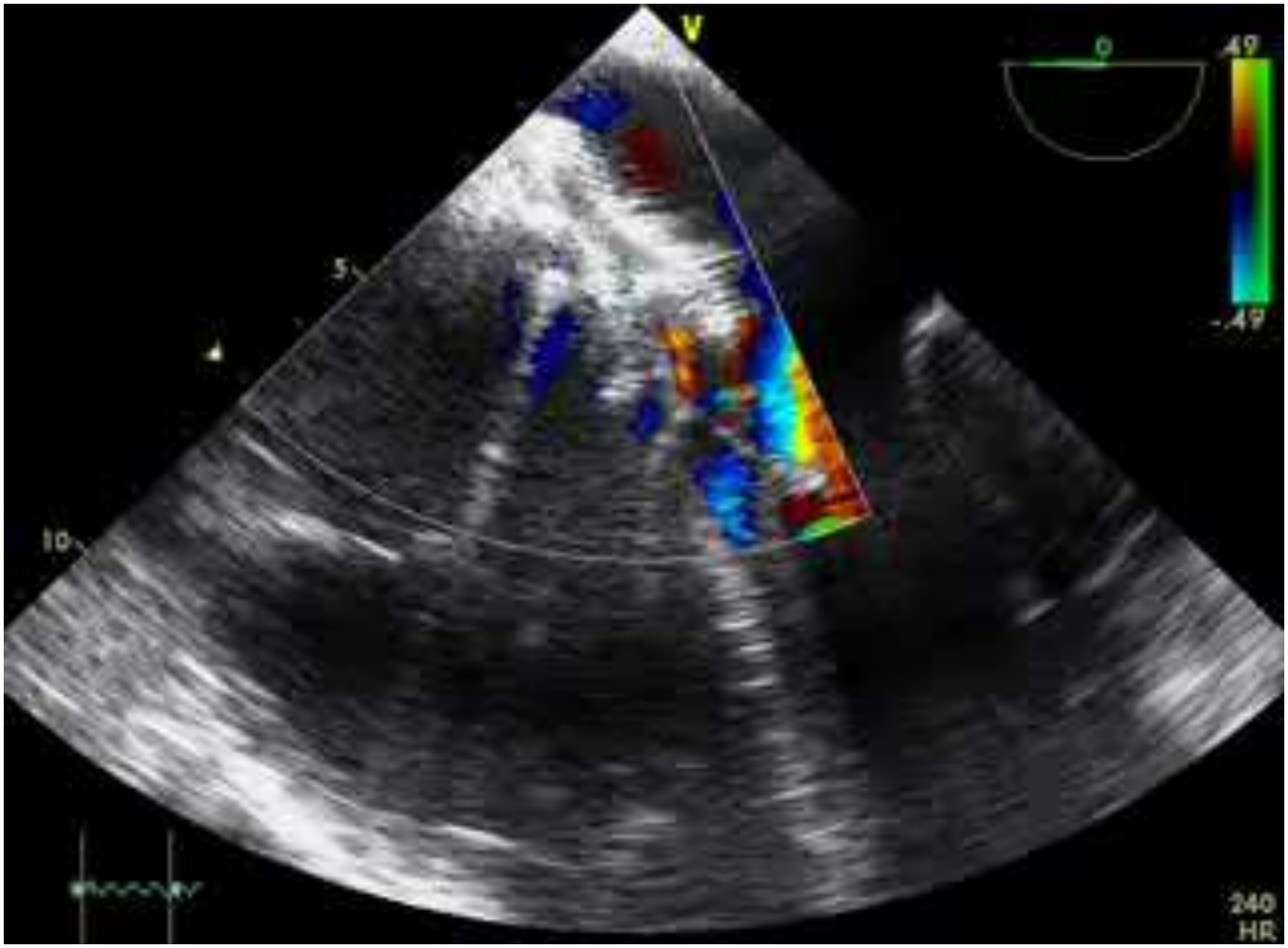




ASD device closure



ASD device closure



Final diagnoses

- Progressive dyspnea and easy fatigability
- ECG: Incomplete RBBB
- CXR: Lung plethora +/- cardiomegaly
- ECHO: Dilated RV/RA + increasedPASP
- TEE: Inter atrial communication

25 years old female – secundum ASD – Mild PHT – adequate rims and defect size by TEE → for device closure versus Surgery

Epidemiology

- ASD accounts for 10% of all congenital heart disease and as much as 20-40% of congenital heart disease presenting in adulthood.
- Depending on the size of the defect, size of the shunt, and associated anomalies, it can result in a spectrum of disease from no significant cardiac sequelae to right-sided volume overload, pulmonary arterial hypertension, and even atrial arrhythmias.
- By the age of 40 years, 90% of untreated patients have symptoms of exertional dyspnea, fatigue, palpitation, sustained arrhythmia, or even evidence of heart failure.

Types of ASD

- The most common types of ASD include the following:
- **Ostium secundum:** The most common type of ASD accounting for 75% of all ASD cases, representing approximately 7% of all congenital cardiac defects and 30-40% of all congenital heart disease in patients older than 40 years.
- **Ostium primum:** The second most common type of ASD accounts for 15-20% of all ASDs.
- **Sinus venosus:** is seen in 5-10% of all ASDs. The defect is located along the superior aspect of the atrial septum. Anomalous connection of the right-sided pulmonary veins is common and should be expected.

Hemodynamics

- The magnitude of the left-to-right shunt across the ASD depends on the defect size, the relative compliance of the ventricles, and the relative resistance in both the pulmonary and systemic circulation.
- The chronic left-to-right shunt results in increased pulmonary blood flow and diastolic overload of the right ventricle.
- Resistance in the pulmonary vascular bed is commonly normal in children with ASD, and the volume load is usually well tolerated even though pulmonary blood flow may be more than 2 times systemic blood flow.

Hemodynamics

- Altered ventricular compliance with age can result in an increased left-to-right shunt contributing to symptoms. The chronic significant left-to-right shunt can alter the pulmonary vascular resistance leading to pulmonary arterial hypertension, even reversal of shunt and Eisenmenger syndrome.
- Because of an increase in plasma volume during pregnancy, shunt volume can increase, leading to symptoms. Pulmonary artery pressure usually remains normal.

Causes of clinical deterioration

1. An age-related decrease in left ventricular compliance augments the left-to-right shunt.
2. Atrial arrhythmias, especially atrial fibrillation, but also atrial flutter or paroxysmal atrial tachycardia, increase in frequency after the fourth decade and can precipitate right ventricular failure.
3. Most symptomatic adults older than 40 years have mild-to-moderate pulmonary arterial hypertension in the presence of a persistent large left-to-right shunt; therefore, the aging right ventricle is burdened by both pressure and volume overload.

Physical findings

- **A hyperdynamic RV impulse** due to increased diastolic filling and large stroke volume.
- **Palpable pulsation of the pulmonary artery** and an ejection click can be detected because of a dilated pulmonary artery.
- **S_1 is typically split**, and the second component may be increased in intensity, reflecting forceful RV contraction and delayed closure of the tricuspid leaflets.
- **S_2 is often widely split and fixed** because of reduced respiratory variation due to delayed pulmonic valve closure (seen only if pulmonary artery pressure is normal and pulmonary vascular resistance is low).
- ASD with moderate-to-large left-to-right shunts result in increased RV stroke volume across the **pulmonary** outflow tract creating a **crescendo-decrescendo systolic ejection murmur**.
- A **rumbling middiastolic murmur at the lower left sternal border** because of increased flow across the tricuspid valve.

INVESTIGATIONS

- ECG
- CXR
- TRANSTHORACIC ECHOCARDIOGRAM
- TRANSOESOPHOGEAL ECHOCARDIOGRAM
- +/- CARDIAC CATHETERIZATION

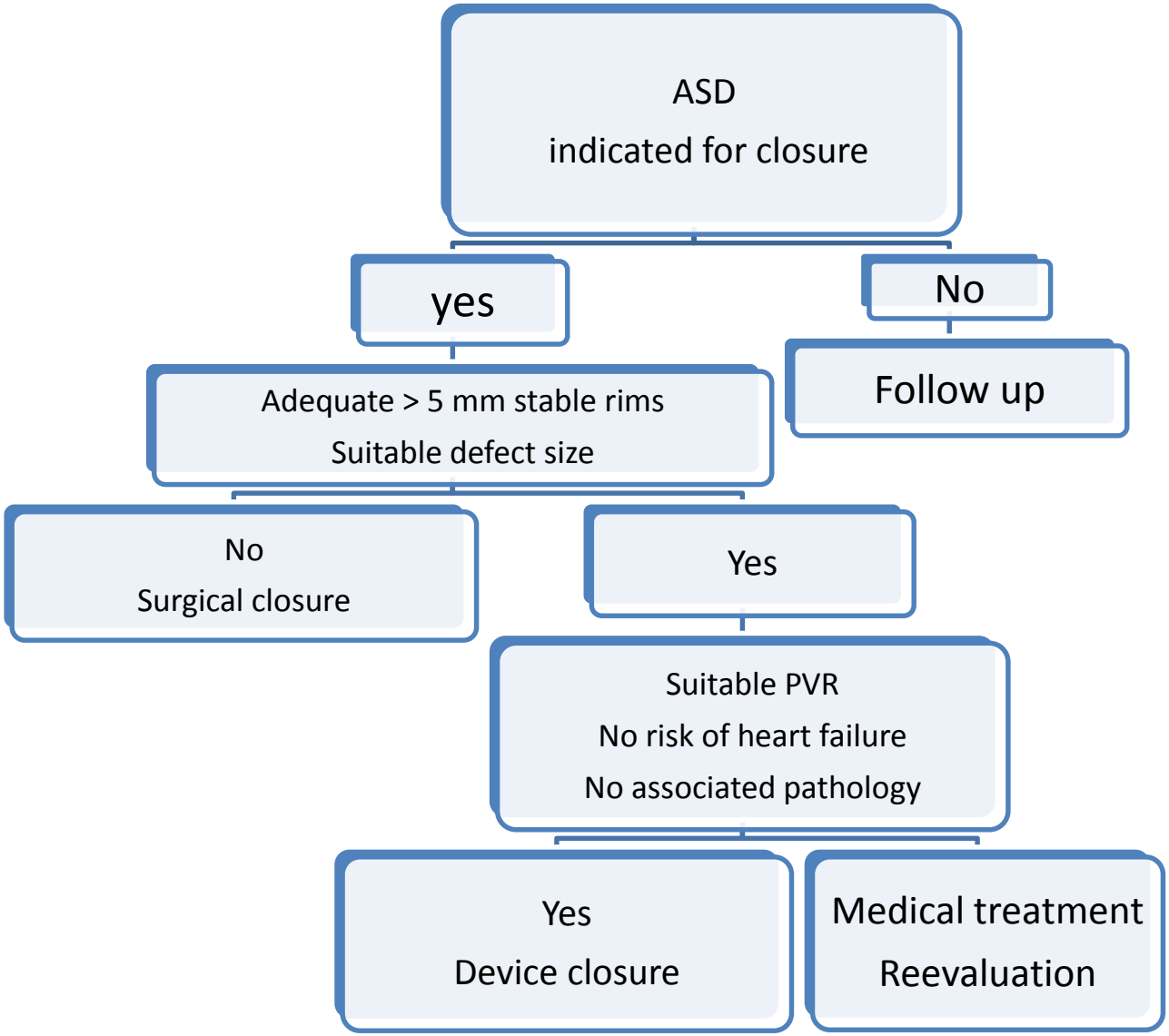
WHEN TO CLOSE AN ASD?

- In general, elective closure is advised for all ASDs with evidence of right ventricular overload or with a clinically significant shunt (pulmonary flow [Qp]–to–systemic flow [Qs] ratio >1.5).
- The widespread use of catheter closure of secundum ASD with lower mortality and without cardiopulmonary bypass has raised the question regarding the need to close even small defects.

When not to close an ASD?

1. A clinically insignificant shunt (Qp-Qs ratio 0.7 or below)
2. In those who have severe pulmonary arterial hypertension or irreversible pulmonary vascular occlusive disease who have a reversed shunt with at-rest arterial oxygen saturations of less than 90%.
3. Whether the patient whose condition is diagnosed well in the sixth decade of life would benefit from surgical closure remains controversial.

TREATMENT STRATEGY



Adult congenital heart disease interactive case presentation 2

Case 2

- Ahmed is a 27 years old engineer not married and has no special habits of medical importance.
- At the age of 21 he was first diagnosed as having high blood pressure.
- Over the last 5 years he sought repeated medical advice with 4 different physicians because of uncontrolled BP.

WHAT INVESTIGATIONS WOULD YOU RECOMMEND FOR THIS PATIENT ?

- Repeated Lab work up including VMA, Serum cortisol, renal functions, lipid profile and serum electrolytes were within normal range.
- Pelvi-abdomenal ultrasound was normal
- He was kept on ARBs, calcium channel blocker and beta blocker with his BP kept in the range of 150/100 on this antihypertensive combination

- 1 year ago he started to complain of occasional leg cramps on climbing the stars.
- In his visit to the last physician, he was asked to do renal duplex
- Renal arterial duplex: no significant stenosis of either the left or right renal arteries but there was blunted Doppler signal of both vessels.

General examination

- He was alert conscious oriented to time place and person.
- Blood pressure 150/100
- Pulse 90 beat per minute equal on both sides
- Peripheral pulsation was not felt
- No pallor no jaundice no cyanoses
- No lower limb edema or congested neck veins

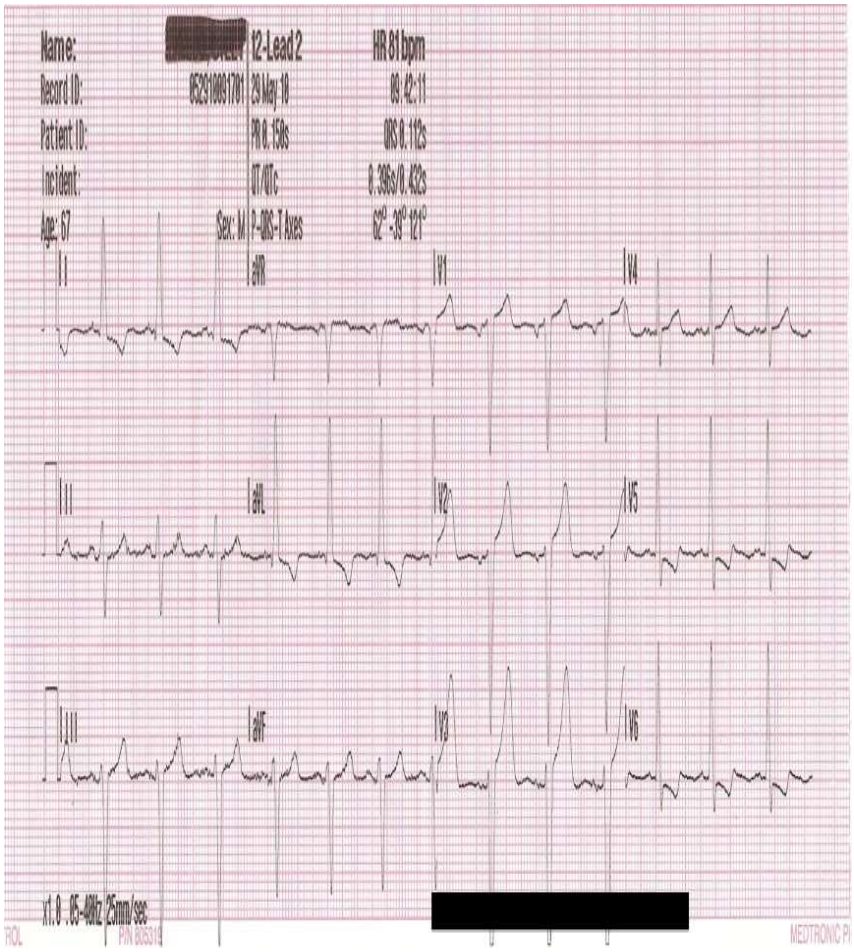
What are the positive findings so far?

- Young male
- Uncontrolled BP on 3 antihypertensive drugs
- Leg cramps on exertion
- Peripheral pulsations not felt
- Blunted renal arterial duplex waves

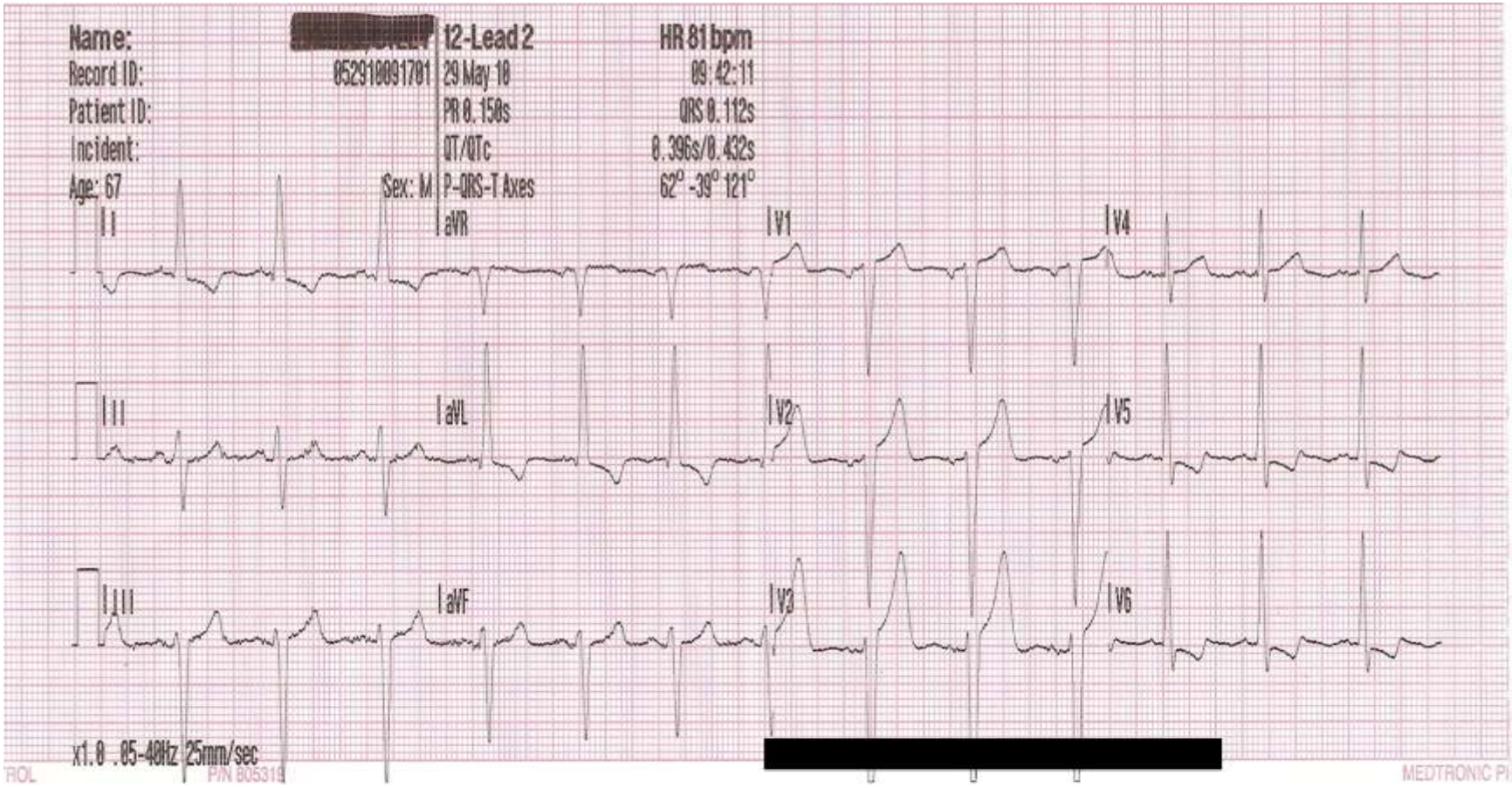
Local examination

- Normal S1, S2
- No additional sounds or murmurs over the precordium
- Soft hemic murmur over the inter scapular area in the back

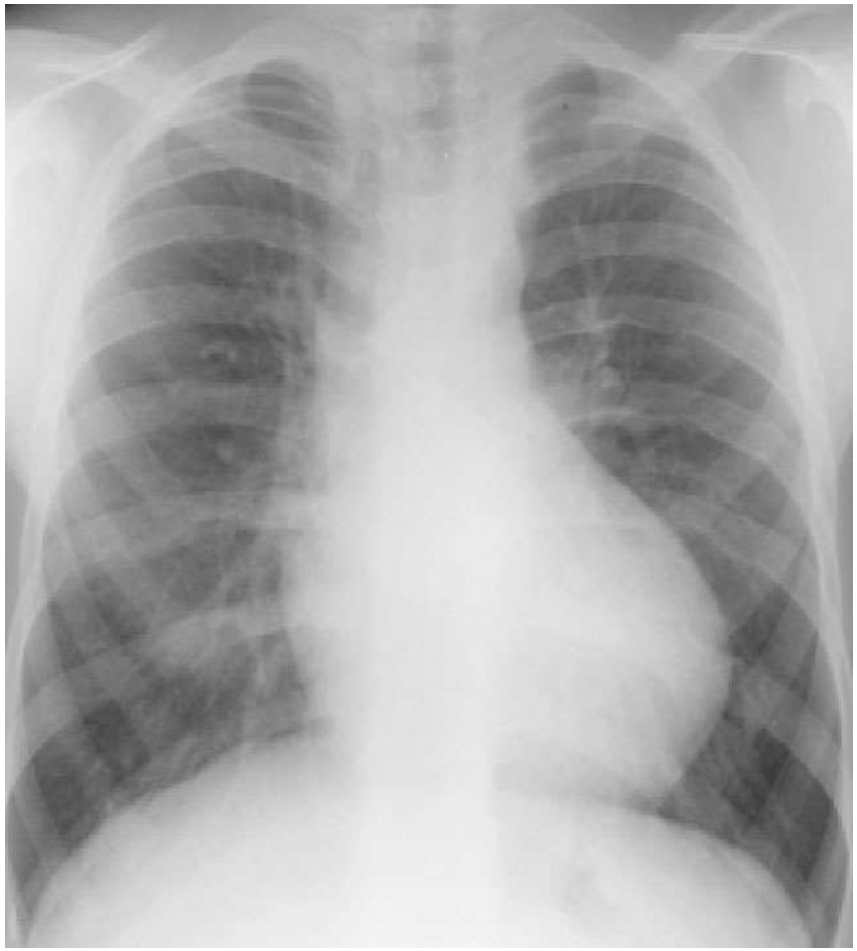
ECG


Name:
Record ID:
Patient ID:
Incident:
Age: 67
12-Lead 2
HR 81 bpm
25 May 18
09:42:11
PR 0.158s
QRS 0.112s
QT/QTc
0.396s/0.452s
Sex: M
P-QRS-T Axes
62° -30° 121°
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
x1.0 .05-40Hz 25mm/sec
MEDTRONIC PI

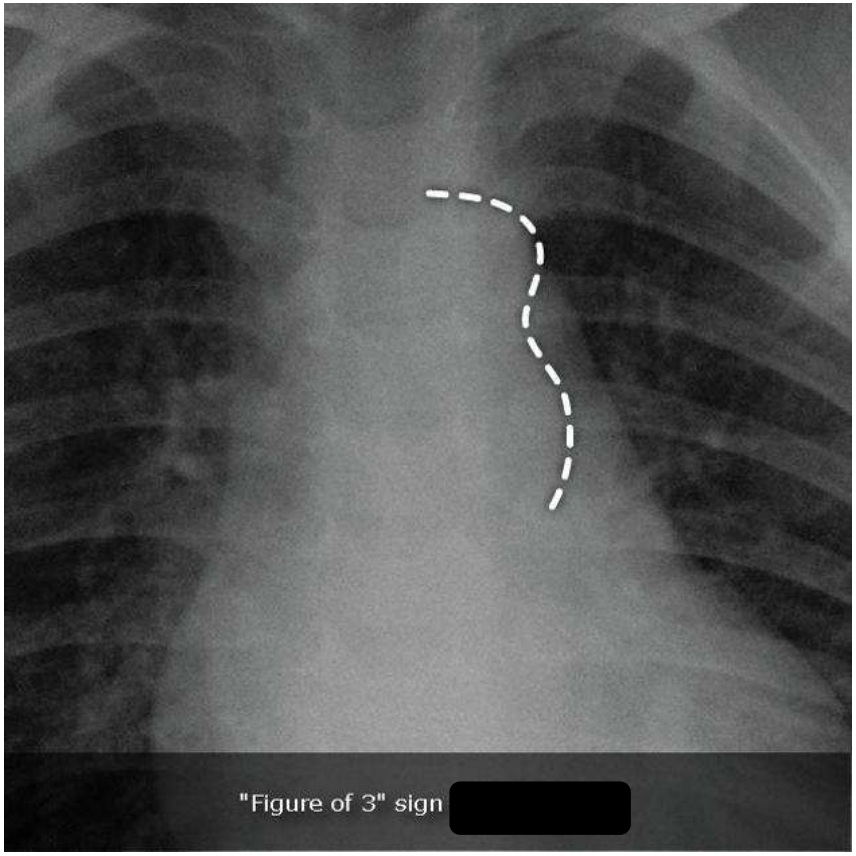
ECG

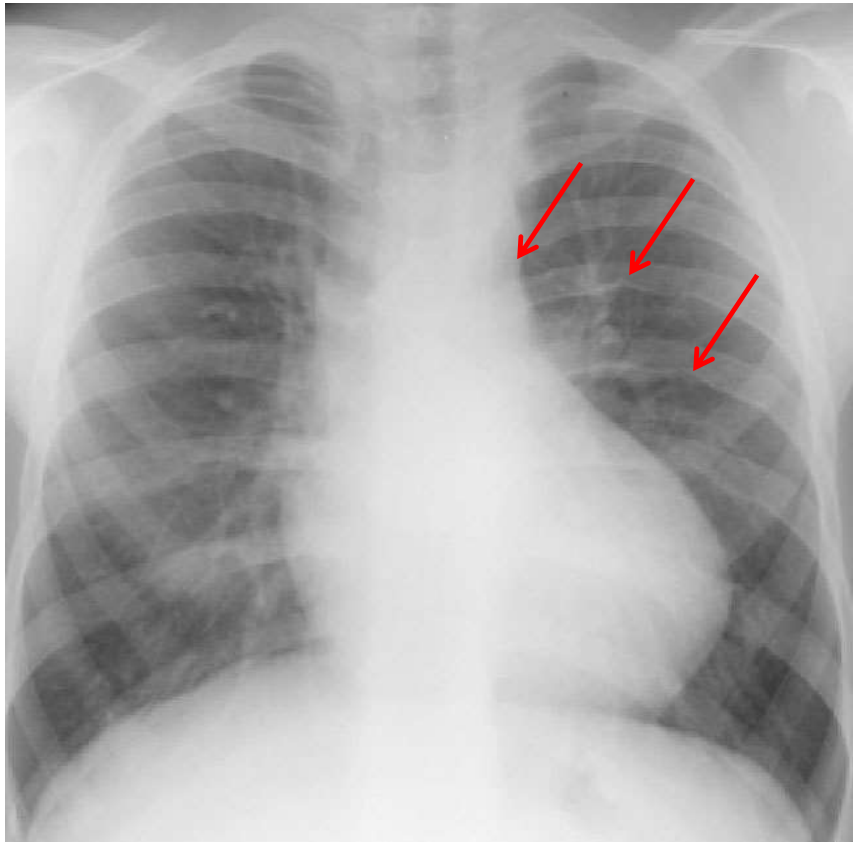

Name:
Record ID: 052918091701
Patient ID:
Incident:
Age: 67 Sex: M
12-Lead 2
29 May 18
PR 0.158s
QT/QTc
P-QRS-T Axes
HR 81 bpm
09:42:11
QRS 0.112s
0.396s/0.432s
62° -39° 121°
I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
x1.0 .05-40Hz 25mm/sec
MEDTRONIC

CXR



CXR





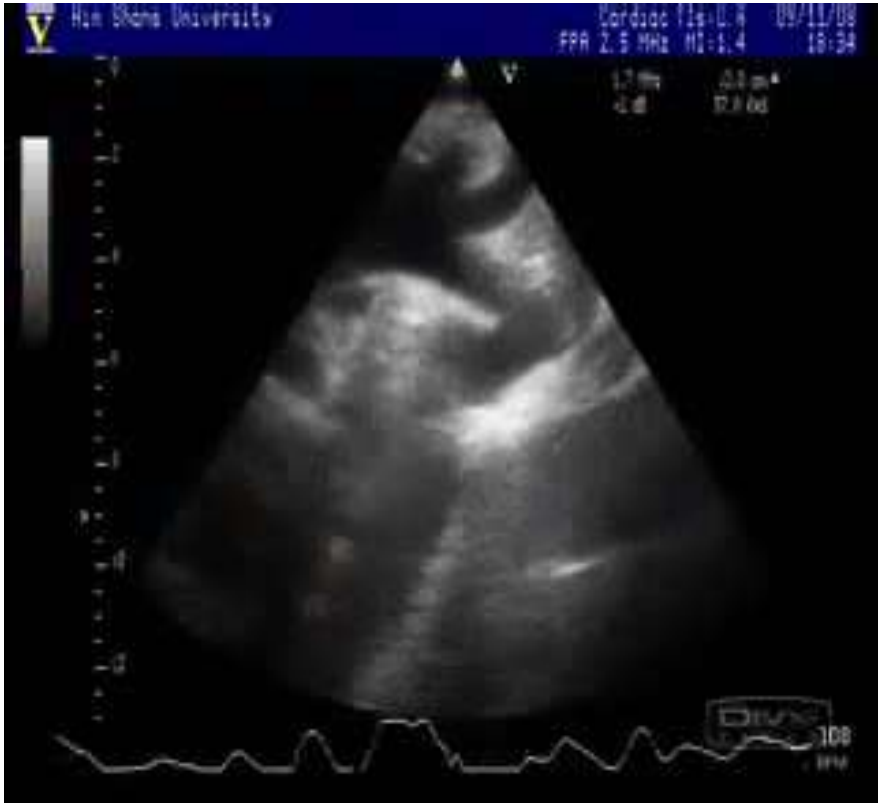
What are the positive findings so far?

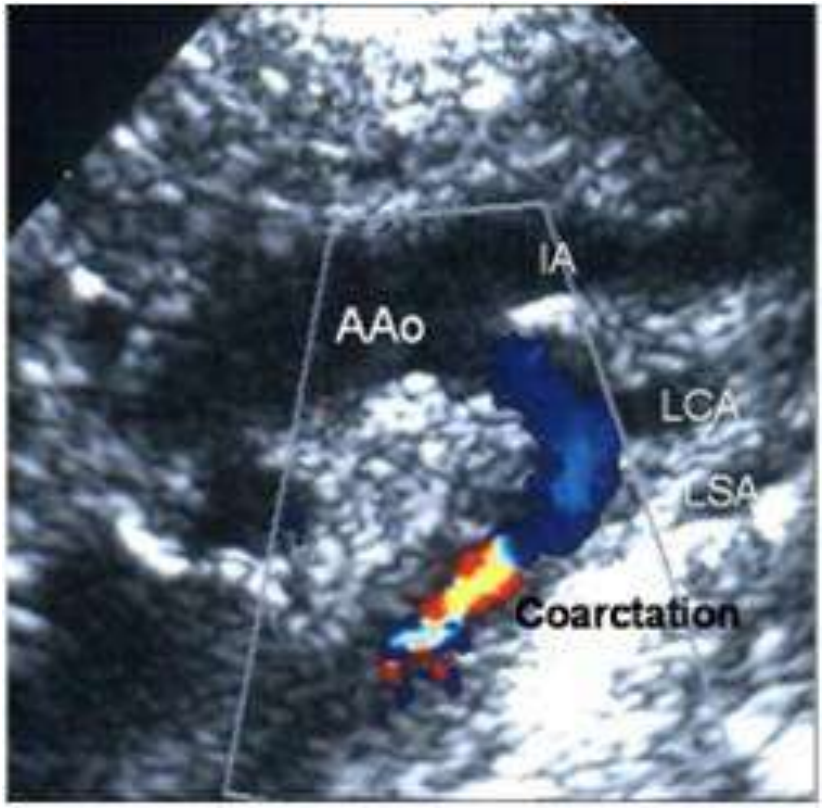
- Young male
- Uncontrolled BP on 3 antihypertensive drugs
- Leg cramps on exertion
- Peripheral pulsations not felt
- Blunted renal arterial duplex waves
- ECG: LVH
- CXR: Figure of 3 sign and rib notches

What is your probable diagnosis?

What is your next step?

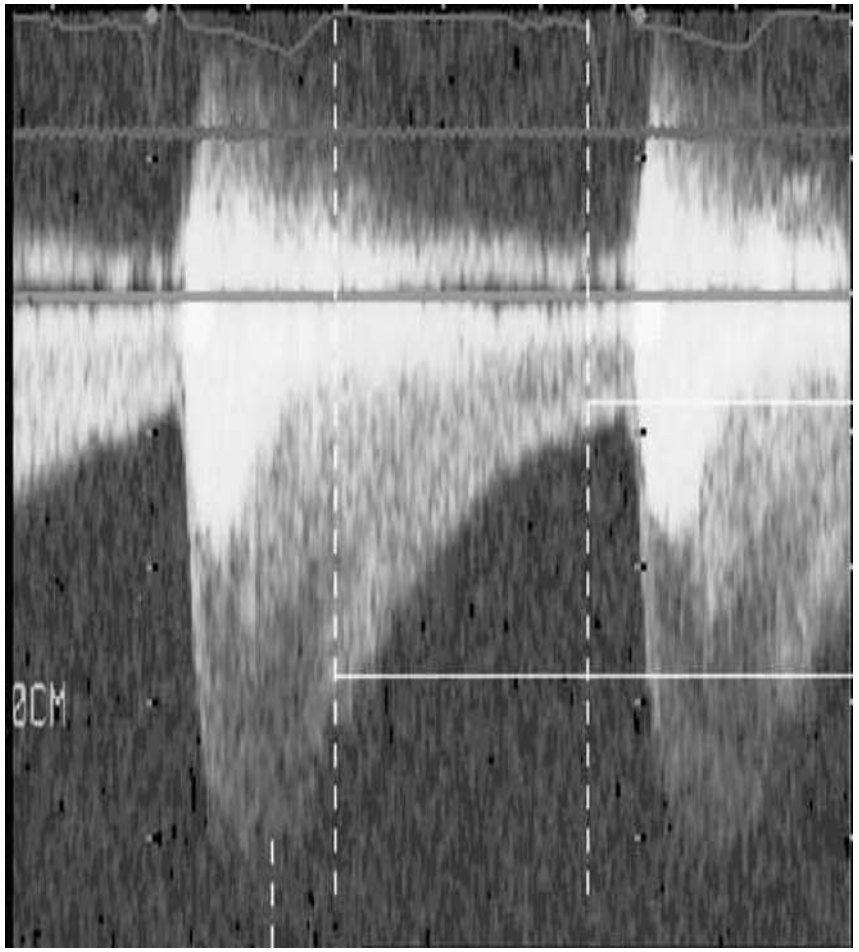
ECHOCARDIOGRAM



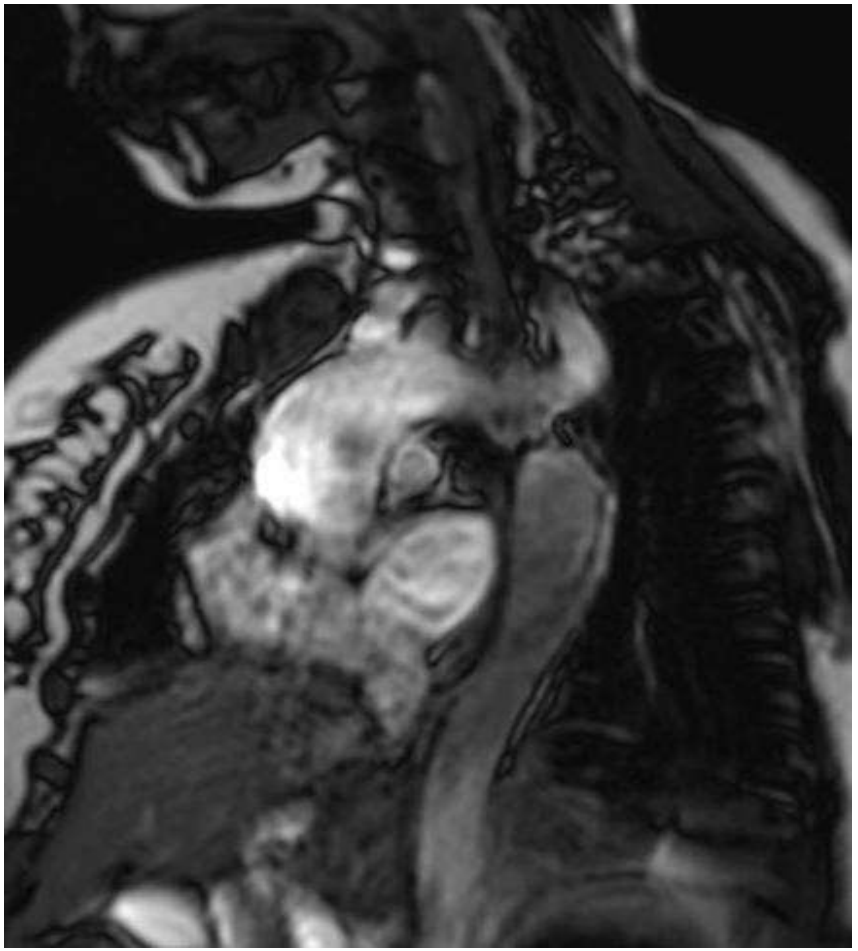


DOPPLER EXAMINATION

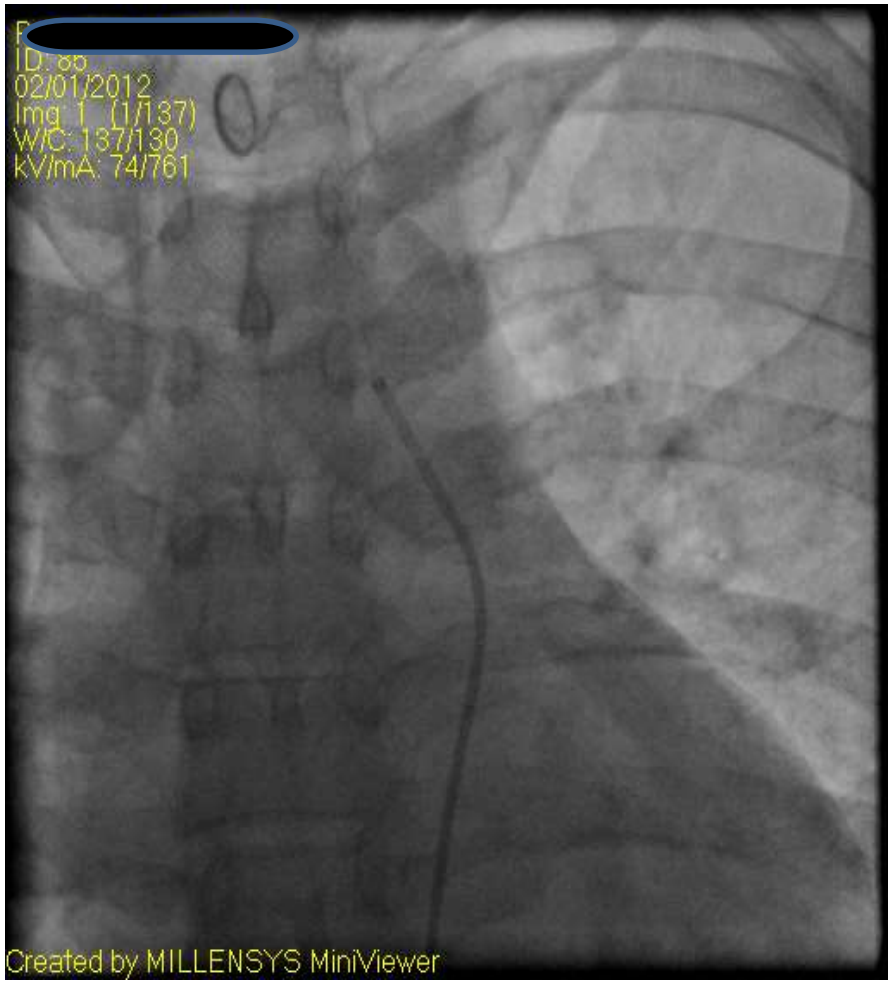
Increased systolic gradient across the coarctation segment with prominent diastolic tailing suggests severe coarctation with loss of the normal pulsatile arterial wave form



MSCT versus MRI



What should be done next for this patient?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W/C: 137/130
kV/mA: 74/761
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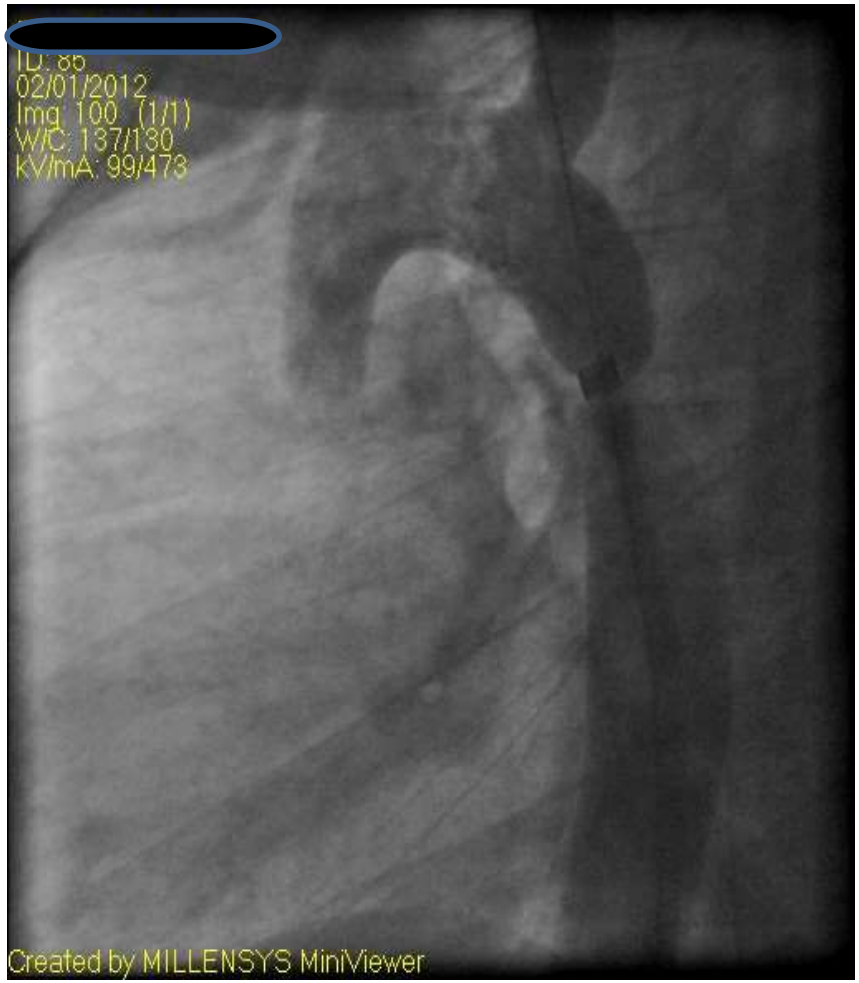
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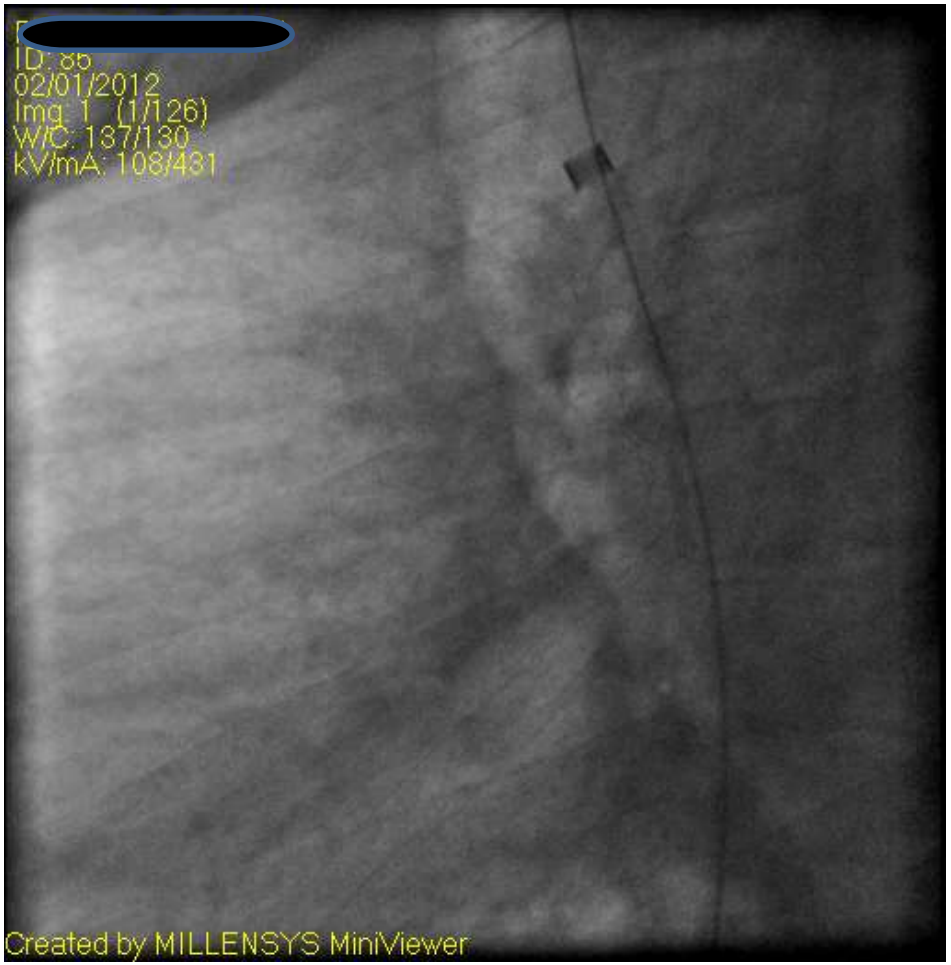
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Img 1 (1/126)
W/C: 137/130
kV/mA: 108/431
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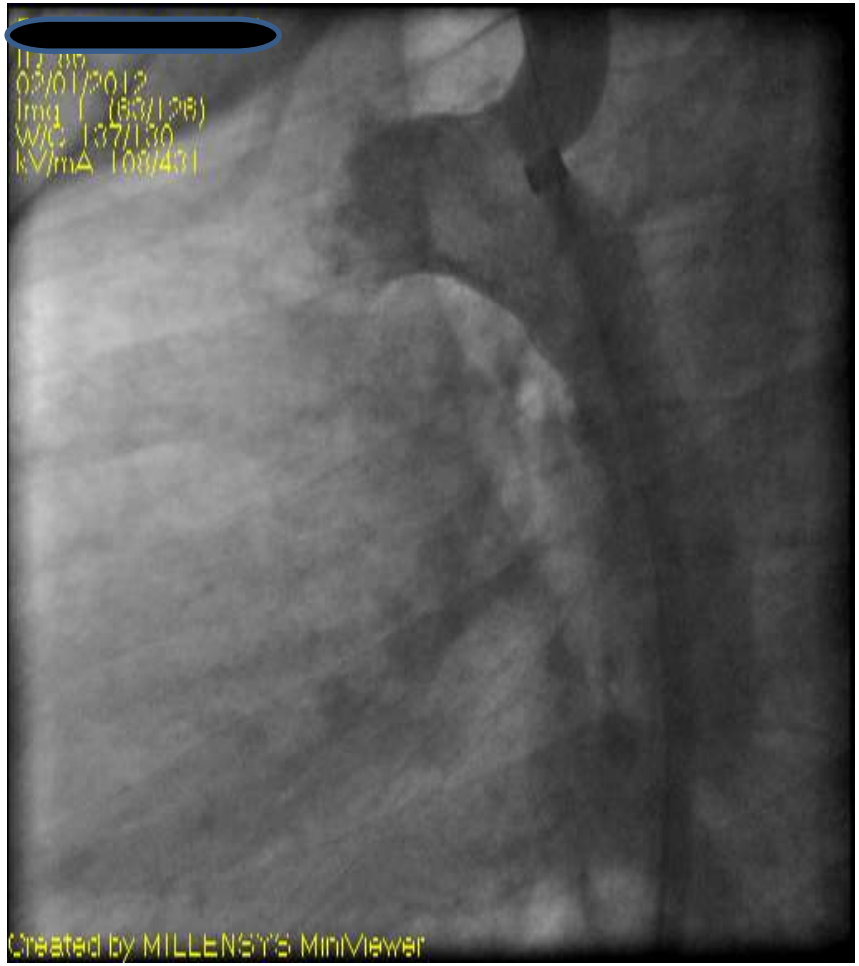
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02/01/2012
W/C: 137/130
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Indications for Intervention in Coarctation of the Aorta

- All patients with a non-invasive pressure difference > 20 mmHg between upper and lower limbs, regardless of symptoms but with upper limb hypertension (> 140/90 mmHg in adults), pathologic blood pressure response during exercice, or significant LVH should have intervention.
- Independent of the pressure gradient, hypertensive patients with ≥ 50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT or invasive angiography) should be considered for intervention.
- Independent of the pressure gradient and presence of hypertension, patients with ≥ 50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT or invasive angiography) may be considered for intervention.

Class[a]	Level[b]
I	C
IIa	C
IIb	C

a = class of recommendation. b = level of evidence.
CMR = cardiac magnetic resonance; CoA = coarctation of the aorta; CT = computed tomography; LVH = left ventricular hypertrophy.

European Heart Journal 2010; doi:10.1093/eurhearj/ehq249

Coarctation
Follow-up

- All pts. require regular follow-up at least every second year including evaluation in specialized GUCH centers. Imaging of the aorta (preferably with CMR) is required. Imaging intervals depend on baseline pathology.
- Residua, sequelae and complications include:
  - arterial hypertension at rest or during exercise,
  - recurring or residual CoA may induce or aggravate systemic arterial hypertension,
  - aneurysms of the ascending aorta or at the intervention site (risk of rupture and death),
  - attention is required for BAV, mitral valve disease, premature CAD, berry aneurysms of the circle of Willis (currently, no routine screening is recommend).

European Heart Journal 2010; doi:10.1093/eurhearj/ehq249

Adult congenital heart disease interactive case presentation 3

Case 3

- Laila is a 35 years old female patient who reports progressive dyspnea over the last 5 years which was sometimes attributed to having bronchial asthma.
- Over the last year the dyspnea was progressive up to dyspnea on mild exertion and easy fatigability with effort intolerance with a tinge of cyanoses on exertion

General examination

- The patient was alert conscious oriented to time place and person
- BP 100/70
- HR 90/Min regular of average volume equal on both sides and well felt peripherally
- Cyanoses
- First degree clubbing

What are the positive findings so far?

- Dyspnea on mild exertion
- Easy fatigability
- Effort intolerance
- Cyanoses
- First degree clubbing

What is your differential diagnoses?

What are the positive findings so far?

- Dyspnea on mild exertion
- Easy fatigability
- Effort intolerance
- Cyanoses
- First degree clubbing

Local examination

- Right ventricular heave with palpable, loud pulmonary component of the second heart sound
- Loud second heart sound with a narrow split
- A diastolic murmur audible along the left sternal border a high-pitched, decrescendo murmur, loudest during inspiration

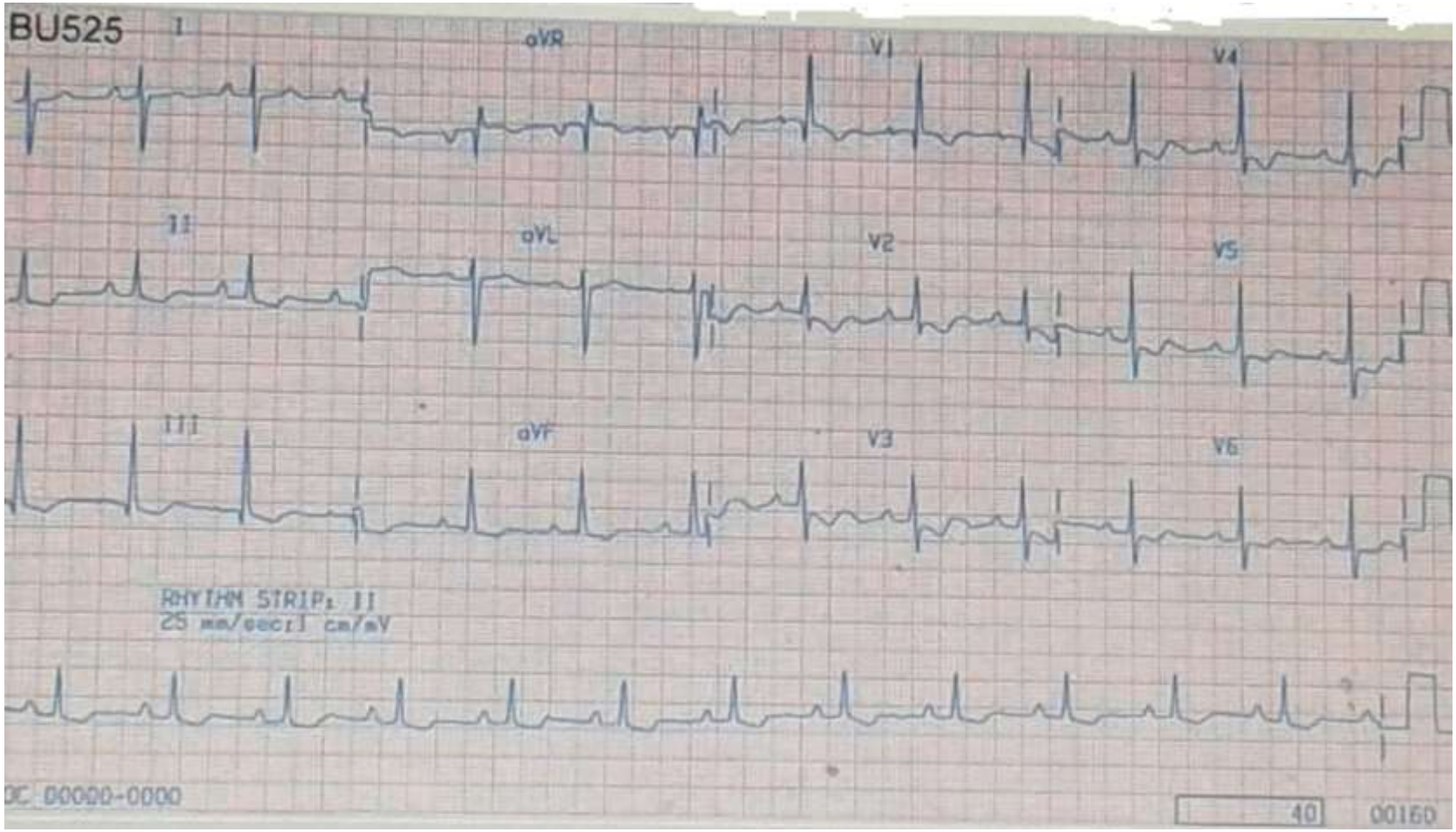
What are the positive findings so far?

- Dyspnea on mild exertion
- Easy fatigability
- Effort intolerance
- Cyanoses
- First degree clubbing
- Pulmonary hypertension

Investigations

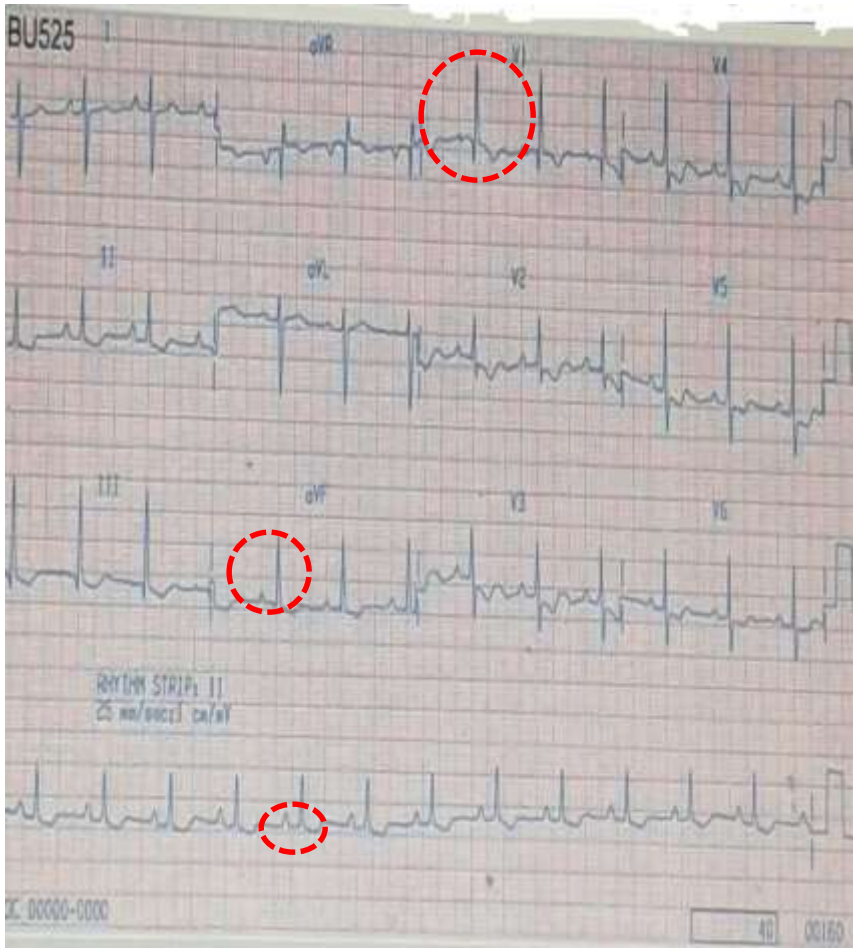
- Resting pulse oximetry showed oxygen saturation 85%
- Her CBC showed Hb 17g%
- Mildly elevated liver enzymes
- Renal functions and serum electrolytes normal
- Clotting profile normal

ECG

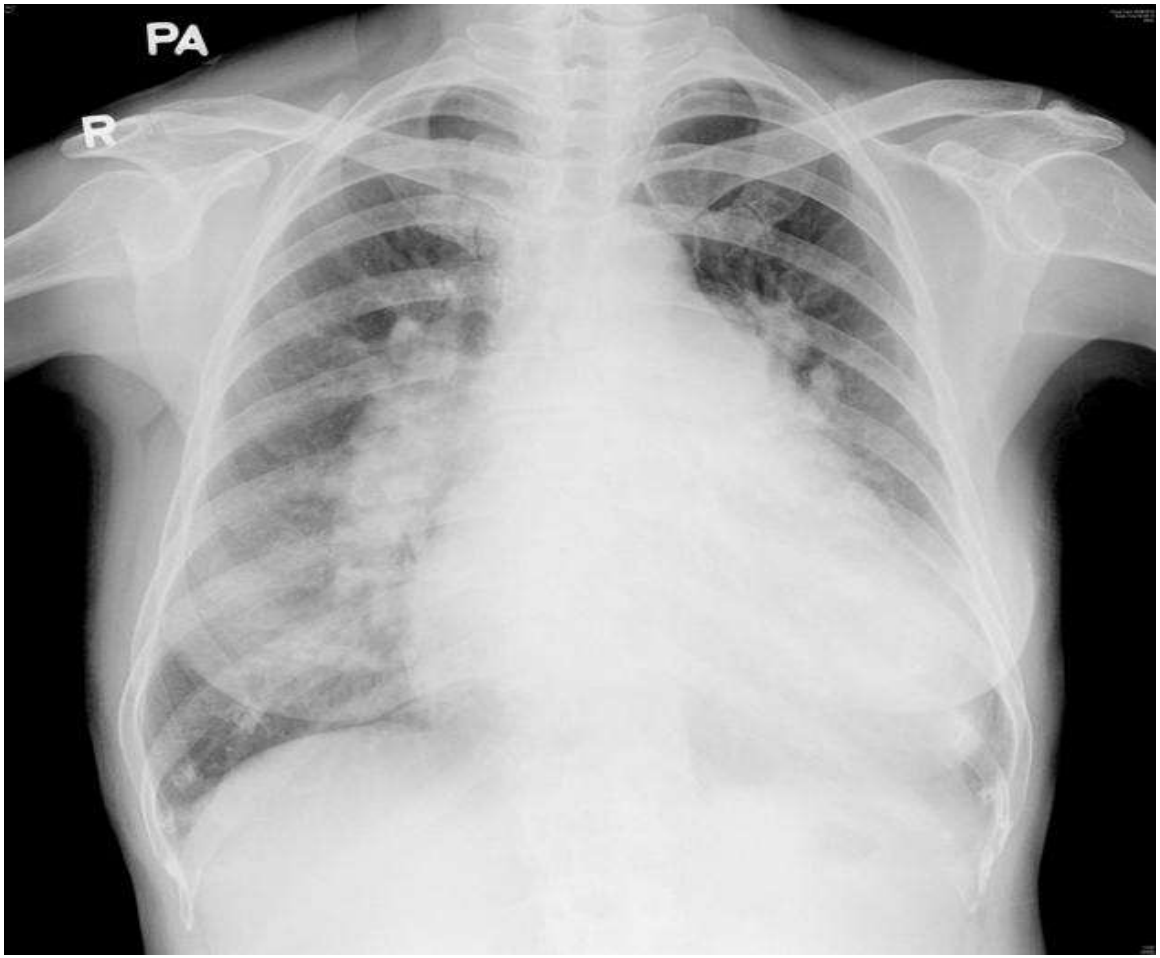

BU525
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
RHYTHM STRIP: II
25 mm/sec; 1 cm/mV
00000-0000
40
00160

ECG

- Sinus rhythm.
- Borderline PR interval.
- Suspicious for right atrial overload (lead 2).
- Right axis deviation to +115^{o}.
- qR complex lead V_1 suggesting suprasystemic RV pressures or RV dysfunction.
- R V_1 of 14 mm suggests RV pressure overload.



CXR



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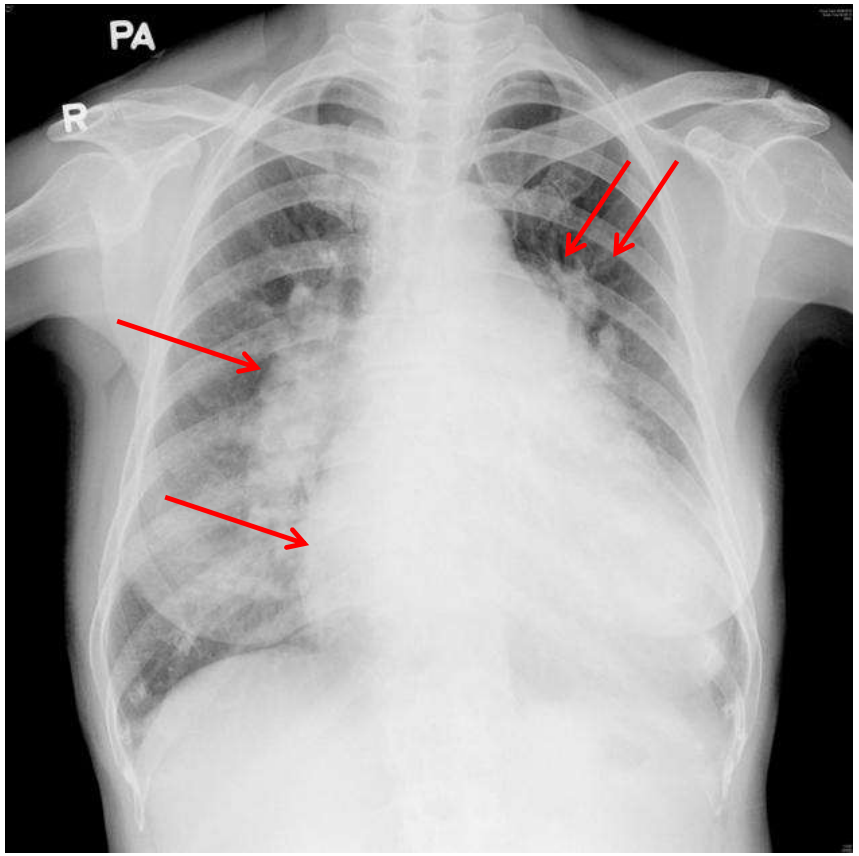


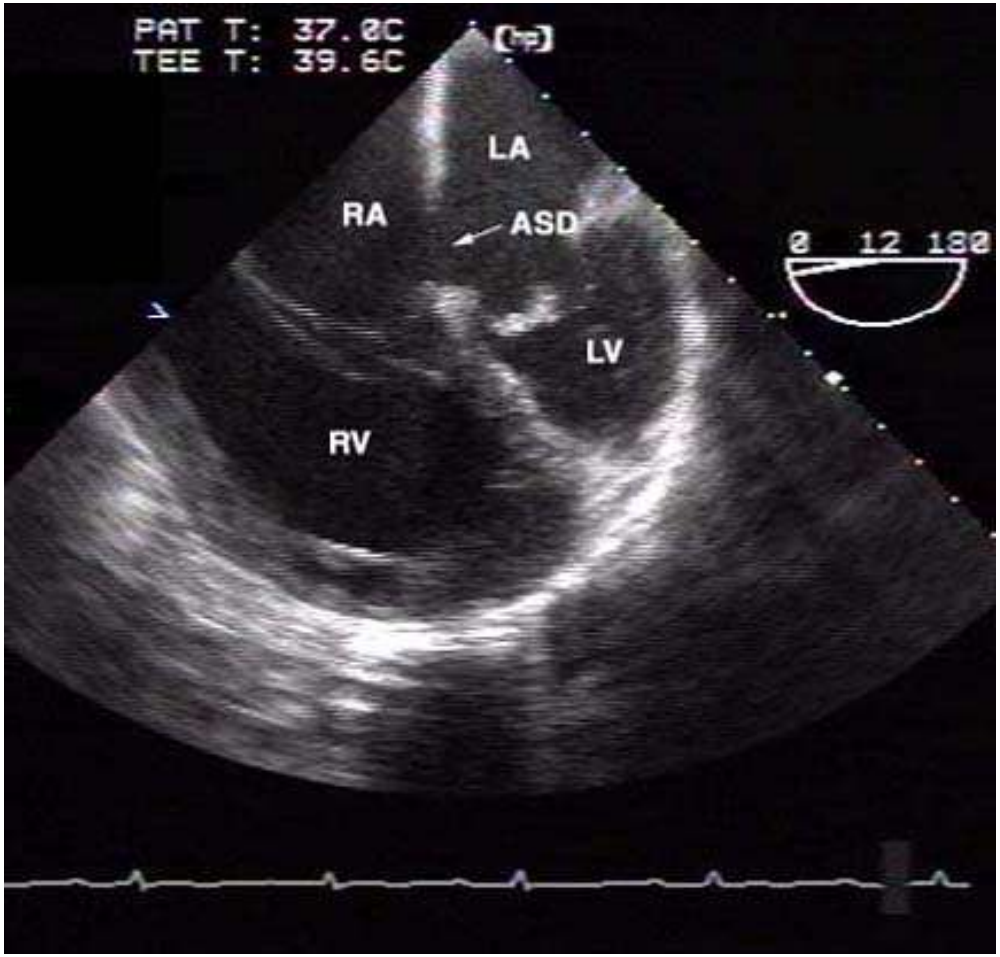
What are the positive findings so far?

- Dyspnea on mild exertion
- Easy fatigability
- Effort intolerance
- Cyanoses
- First degree clubbing
- Pulmonary hypertension
- Oxygen desaturation
- Relative polycysthmia
- ECG: RVH – P pulmonale
- CXR: Prunning

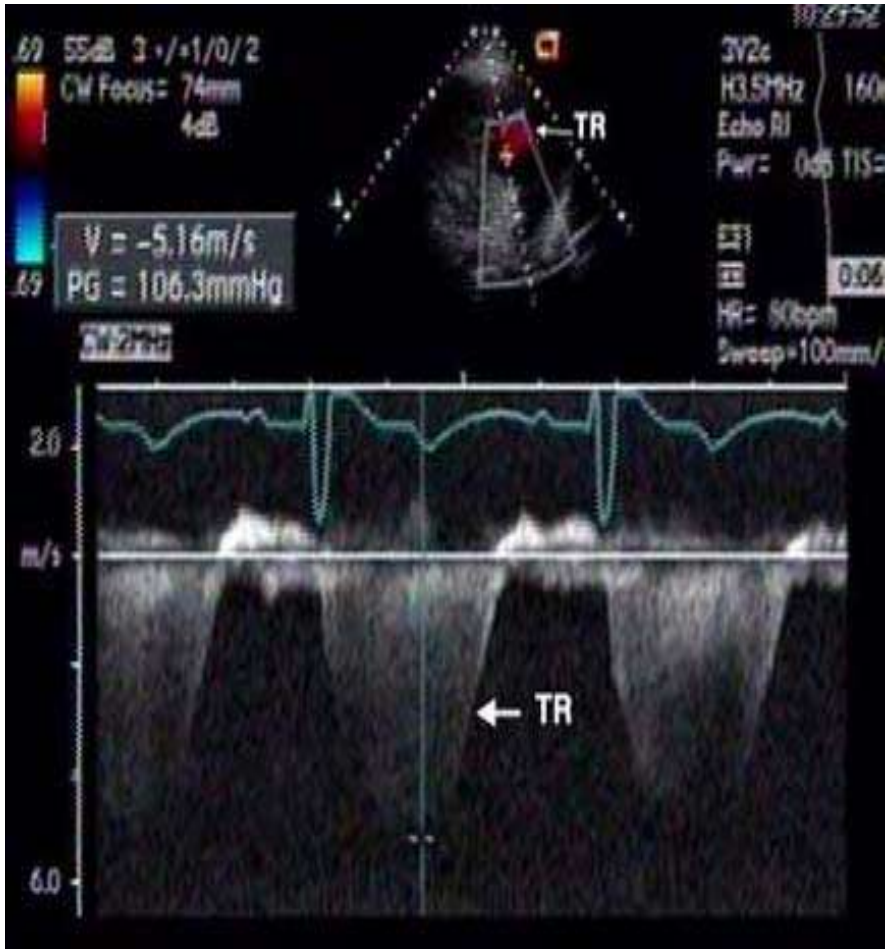
What is your probable diagnosis?

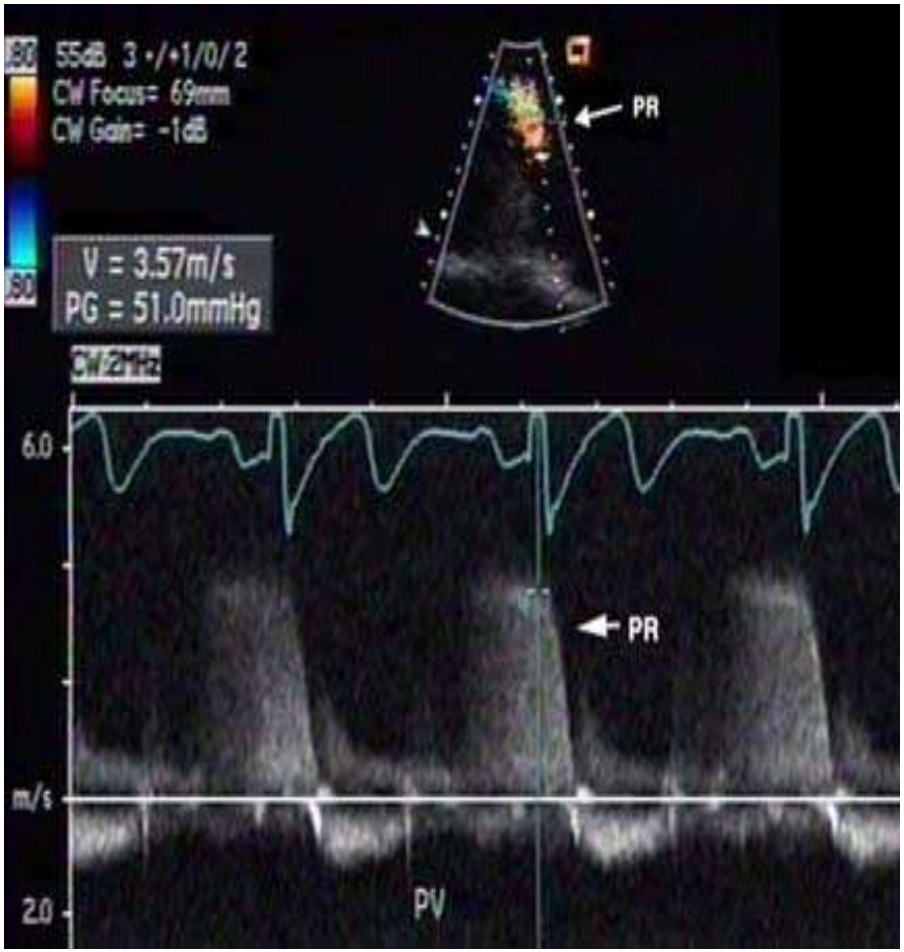
What is your next step?

Echocardiogram



Echocardiogram





What about cardiac catheterization?

Role of cardiac catheterization

- The main purpose in cardiac catheterization of a patient with ES is to determine vascular physiology and to assess pulmonary vascular reactivity to vaso-active substances such as oxygen, nitric oxide and prostacyclin.
- Based on these measurements therapeutic decisions on treatment with vaso-active substances, catheter interventions, or surgery can be made.

Role of cardiac catheterization

- An acute reduction of the mean pulmonary arterial pressure of >10 mm Hg with a resultant mean pulmonary arterial pressure of 40mmHg or less without a fall in cardiac output is considered a positive vaso-reactivity response.

Role of cardiac catheterization

- In patients with evidence of vasoreactivity during acute testing but high PVR and bidirectional shunting, balloon test occlusion may provide additional information on the suitability for closure and the possible post-procedural outcome.

- A drop in cardiac output and/or an increase in right ventricular filling pressures with test balloon occlusion would suggest a low likelihood to benefit from permanent closure, as well as a higher perioperative risk

Eisenmenger's syndrome

- Is defined as obstructive pulmonary vascular disease that develops as a consequence of a large pre-existing left-to-right shunt causing pulmonary artery pressures to increase and approach systemic levels, such that the direction of blood flow then becomes bi-directional or right-to-left.

Epidemiology

- The frequency of pulmonary hypertension and the development of reversed shunting vary depending on the specific heart defect and operative interventions.
- 50% of infants with a large, non-restrictive ventricular septal defect (VSD) or patent ductus arteriosus (PDA) develop pulmonary hypertension by early childhood.
- 10% of patients with a large secundum atrial septal defect (ASD) progress to pulmonary hypertension but usually not until after the third decade of life.

Clinical picture

Symptoms:

- Breathlessness
- Fatigue
- Lethargy
- Severely reduced exercise tolerance
- Presyncope, syncope
- Others: bleeding tendency, symptoms of erythrocytoses as myalgia, lethargy, parathesia, headache, tinnitus, blurred vision

Signs:

- Central cyanosis
- Clubbing
- Jugular venous pulse A-wave dominant, and, in the presence of a significant TR, the V wave may be prominent.
- Right ventricular heave +/- palpable S_2.
- Loud P_2
- High-pitched early diastolic murmur of PR
- Right-sided S_4
- Pulmonary ejection click
- Single S_2

Complications

- Haemoptysis
- Bleeding disorders
- Hyperuricaemia, gout, nephrolithiasis
- Polycythaemia, hyperviscosity syndrome
- Angina pectoris
- Arrhythmias (atrial fibrillation/flutter)
- Syncope
- Sudden death, stroke, transient ischaemic attacks
- Paradoxical emboli
- Infective endocarditis
- Pulmonary arterial aneurysm/calcification
- Brain abscess
- Progressive valvar stenosis or regurgitation
- Renal dysfunction (especially proteinuria)
- Hyperbilirubinaemia may cause gallstones
- Hypertrophic osteoarthropathy

Investigations

Blood tests:

- CBC
- ABG
- Pulse oximetry
- Uric acid
- Renal function
- Iron study
- BNP

Imaging:

- ECG
- CXR
- Echocardiogram
- Transoesophogeal echo
- MRI
- Cardiac catheterization

Differential diagnoses

Updated Clinical Classification of Pulmonary Hypertension (Dana Point, 2008)

1. Pulmonary arterial hypertension (PAH)
1.1. Idiopathic PAH
1.2. **Heritable**
1.2.1. **BMPR2**
1.2.2. **ALK1, endoglin (with or without hereditary hemorrhagic telangiectasia)**
1.2.3. **Unknown**
1.3. Drug- and toxin-induced
1.4. Associated with
1.4.1. Connective tissue diseases
1.4.2. HIV infection
1.4.3. Portal hypertension
1.4.4. Congenital heart diseases
1.4.5. **Schistosomiasis**
1.4.6. **Chronic hemolytic anemia**
1.5 Persistent pulmonary hypertension of the newborn

1'. **Pulmonary veno-occlusive disease (PVOD) and/or pulmonary capillary hemangiomatosis (PCH)**

2. Pulmonary hypertension owing to left heart disease
2.1. **Systolic dysfunction**
2.2. **Diastolic dysfunction**
2.3. Valvular disease

3. Pulmonary hypertension owing to lung diseases and/or hypoxia
3.1. Chronic obstructive pulmonary disease
3.2. Interstitial lung disease
3.3. **Other pulmonary diseases with mixed restrictive and obstructive pattern**
3.4. Sleep-disordered breathing
3.5. Alveolar hypoventilation disorders
3.6. Chronic exposure to high altitude
3.7. Developmental abnormalities

4. **Chronic thromboembolic pulmonary hypertension (CTEPH)**

5. **Pulmonary hypertension with unclear multifactorial mechanisms**
5.1. **Hematologic disorders: myeloproliferative disorders, splenectomy**
5.2. **Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis:** lymphangioleiomyomatosis, neurofibromatosis, vasculitis
5.3. **Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders**
5.4. **Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure on dialysis**

Treatment

- Oxygen therapy ??
- Pulmonary vasodilators:
  - Prostacyclines
  - Nitric oxide
  - Endothelin receptor antagonist
  - Phosphodiestrase inhibitors
- Endocarditis prophylaxis
- Management of eryhrocytosis
- Thromboses ??

Medications

- The medical treatment of Eisenmenger syndrome is directed toward the improvement of symptoms related to heart failure and pulmonary hypertension and the prevention and management of complications related to cyanotic congenital heart disease.

- A partial list of medications used in the management of Eisenmenger syndrome includes aspirin, to prevent thrombotic complications; allopurinol, for gout; iron supplementation, for microcytosis; and digitalis and diuretics, for symptoms of heart failure.

Specific pulmonary vasodilators

- Endothelin antagonists:

These agents competitively bind to endothelin-1 (ET-1) receptors ET_A and ET_B in endothelium and vascular smooth muscle, inhibiting vessel constriction and elevation of blood pressure.

- **Bosentan (Tracleer)**
- **Ambrisentan (Letairis)**

- Phosphodiesterase-5 Enzyme Inhibitors

These agents act synergistically with nitric oxide to promote smooth muscle relaxation through its anti proliferative effect.

- **Sildenafil**
- **Tadalafil**

Specific pulmonary vasodilators

- Prostaglandins

These drugs can be effective in reversing reactive pulmonary vasoconstriction and can, therefore, lower pulmonary vascular resistance, decrease afterload, reduce the right ventricle, and reduce right-to-left shunting. In some patients, chronic prostacyclin analogue therapy (epoprostenol) can be of benefit, particularly as a bridge to heart-lung transplantation.

- **Epoprostenol (Flolan, Veletri)**
- **Iloprost (Ventavis)**

Pregnancy and contraception

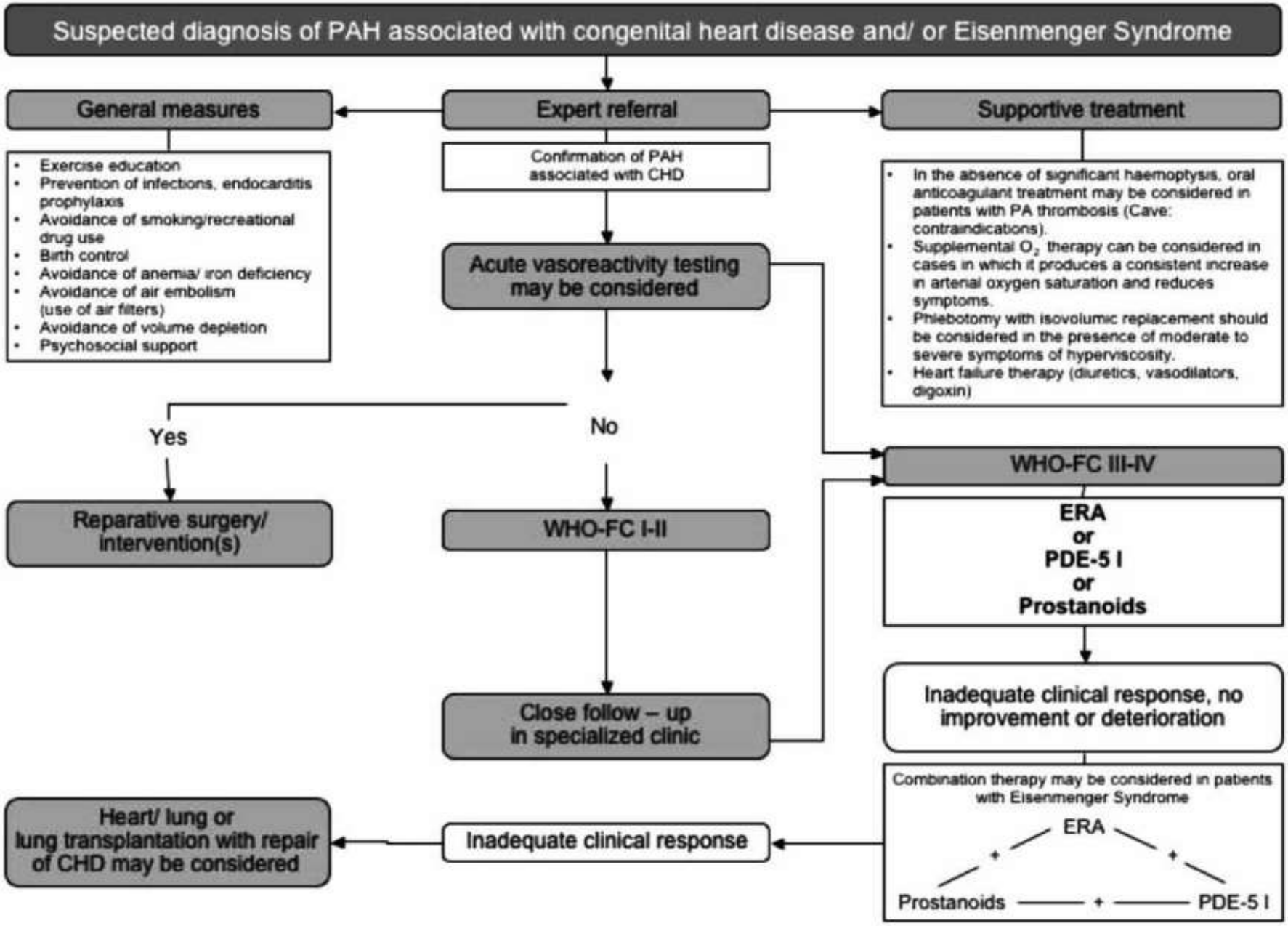
- Pregnancy should be avoided. If it occurs, early termination is advised. If pregnancy is continued, maternal mortality approaches 50% with each pregnancy and fetal loss is similar.
- Tubal ligation is recommended for contraception. Intrauterine devices should be avoided as they may cause significant menorrhagia and increase the risk of endocarditis.
- Combined oral contraceptive pills should be avoided.
- The risk of congenital heart defects in offspring is approximately 10% but depends on the initial cardiac defect.

Other risks

- General anaesthesia
- Dehydration
- Haemorrhage
- Non-cardiac surgery and cardiac surgery
- Certain drugs, eg vasodilators, diuretics, combined oral contraceptive pills, danazol
- Anaemia (prevention of iron deficiency is important)
- Cardiac catheterization
- Intravenous lines (risk of paradoxical air embolism and infection)
- Altitude exposure
- Lung infections

Prognosis

- Most patients with Eisenmenger's syndrome survive to adulthood, with a reported 77% and 42% survival rate at 15 and 25 years of age.
- The most common causes of death are sudden death, congestive heart failure and haemoptysis.
- Pregnancy, perioperative mortality following non-cardiac surgery, and infectious causes (brain abscesses and endocarditis) account for most of the other deaths.
- Reduced systemic blood flow and elevated right atrial pressure are associated with high mortality rates in adults with Eisenmenger's syndrome


Suspected diagnosis of PAH associated with congenital heart disease and/ or Eisenmenger Syndrome
General measures
Expert referral
Supportive treatment
Exercise education
Prevention of infections, endocarditis prophylaxis
Avoidance of smoking/recreational drug use
Birth control
Avoidance of anemia/ iron deficiency
Avoidance of air embolism (use of air filters)
Avoidance of volume depletion
Psychosocial support
Confirmation of PAH associated with CHD
In the absence of significant haemoptysis, oral anticoagulant treatment may be considered in patients with PA thrombosis (Cave: contraindications).
Supplemental O_2 therapy can be considered in cases in which it produces a consistent increase in arterial oxygen saturation and reduces symptoms.
Phlebotomy with isovolumic replacement should be considered in the presence of moderate to severe symptoms of hyperviscosity.
Heart failure therapy (diuretics, vasodilators, digoxin)
Acute vasoreactivity testing may be considered
Yes
No
WHO-FC III-IV
Reparative surgery/ intervention(s)
WHO-FC I-II
ERA
or
PDE-5 I
or
Prostanoids
Close follow – up in specialized clinic
Inadequate clinical response, no improvement or deterioration
Combination therapy may be considered in patients with Eisenmenger Syndrome
ERA
Prostanoids
PDE-5 I
Heart/ lung or lung transplantation with repair of CHD may be considered
Inadequate clinical response

Other possible presentations

TABLE 30. Imaging Findings and Late Complications in Adult Congenital Heart Disease

Lesion	ECG and CXR Findings	Late Complications
Small VSD	Normal	Endocarditis
Large VSD	ECG: RV or RV/LV hypertrophy CXR: LA and LV enlargement, increased pulmonary vascular markings; with PAH: prominent central pulmonary arteries, reduced peripheral pulmonary vascular markings	Left heart enlargement, PAH
Small PDA	Normal	Endocarditis
Large PDA	ECG: LA enlargement, LV hypertrophy; with PAH: RV hypertrophy CXR: Cardiomegaly, increased pulmonary vascular markings; calcification of PDA (occasional); with PAH: prominent central pulmonary arteries, reduced peripheral pulmonary vascular markings	Endocarditis, heart failure
Pulmonary valve stenosis	ECG: Normal when RV systolic pressure <60 mm Hg; if RV systolic pressure >60 mm Hg: RA enlargement, right axis deviation, RV hypertrophy CXR: Pulmonary artery dilatation, calcification of pulmonary valve (rare); RA enlargement may be noted	Risk of severe pulmonary valve regurgitation after pulmonary valvuloplasty
Repaired tetralogy of Fallot	ECG: RBBB, increased QRS duration (QRS duration reflects degree of RV dilatation) CXR: Cardiomegaly with pulmonary or tricuspid valve regurgitation; right aortic arch in 25% of cases	Post repair: Increased atrial and ventricular arrhythmia risk QRS >180 msec increases risk of ventricular tachycardia and sudden death

Thank you