

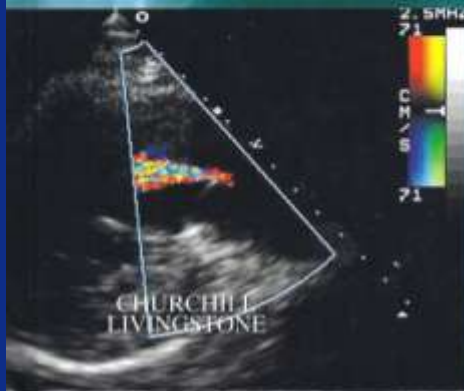
Adult Congenital Heart Disease: A Growing Problem

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CASES IN ADULT CONGENITAL HEART DISEASE



ACHD Resources in Ohio

<u>City</u>	<u>State/Prov.</u>	<u>Institution</u>	<u>Medical Director</u>	<u>Patient Vists/Yr.*</u>	<u>ACHD Oper./Yr.**</u>
<input type="checkbox"/> Akron	OH	Adult Congenital Heart Service Summa Health System, Akron General Medical Center (330) 543-8521	John R. Lane, MD	379	6
<input type="checkbox"/> Cincinnati	OH	Adult Congenital Heart Disease Clinic Cincinnati Children's Hospital Medical Center (513) 636-7593	Gary Webb, MD	550	2
<input type="checkbox"/> Cleveland	OH	Cleveland Clinic Adult Congenital Heart Program Cleveland Clinic (216) 445-7430	Richard Krasuski, MD	824	74
<input type="checkbox"/> Columbus	OH	COACH (Columbus Ohio Adult Congenital Heart) Program Nationwide Children's Hospital (614) 722-5622; (614) 293-6638	Curt J. Daniels, MD	2113	75
<input type="checkbox"/> Cleveland	OH	ADCON, Congenital Heart Center, Heart & Vascular Institute Rainbow Babies & Children's Hospital, University Hospitals (216) 844-7700	Ernest Siwik, MD	615	10

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Situations When We Might Help

- When you don't know a lot about a CHD diagnosis.
- When a CHD diagnosis includes a person's name.
 - Fontan.
 - Mustard.
 - Senning.
 - Rastelli.
 - Jatene.

Situations When We Might Help

- CHD patients who are cyanotic.
- CHD patients with pulmonary hypertension.
- CHD patients with only one ventricle.
- CHD patients needing noncardiac surgery.
- CHD patients needing special cardiac surgery.
 - David operation.
 - Ross procedure.
 - Aortic valve repair.

Situations When We Might Help

- CHD patient candidates for a heart catheterization.
- Sexually active CHD patients.
- CHD patients contemplating pregnancy.
- Pregnant CHD patients.
- CHD patients with arrhythmias.
- CHD patients with syndromes.

Consider ACHD clinics to contain resource people who have developed networks of specialists with special interests and abilities.

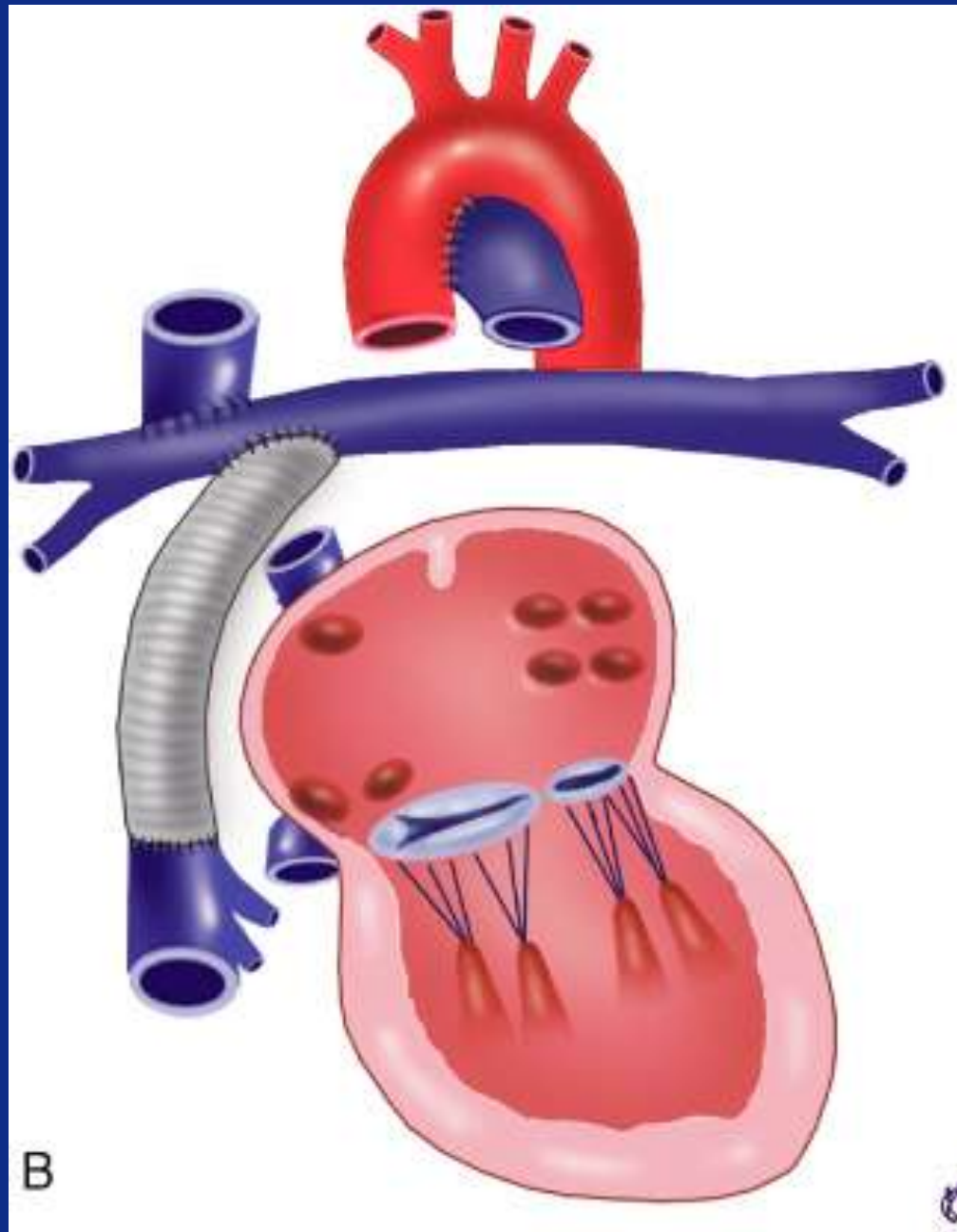


The Toronto Congenital Cardiac Centre for Adults – **founded 1959**



Patient 63 - History

- 31-year-old female supermarket clerk
- 16 weeks pregnant.
- Cyanotic at birth.
- Left BT shunt at age 11.
- Lateral tunnel Fontan procedure at age 20.
- Lost to follow-up until age 31.



Patient 63 - History

- NYHA class I.
- No functional decline recently.
- No medications.

Patient 63 “Risk Factors”

- “Don’t know much about that”.
 - Fontan.
 - Blalock Taussig.
- Man’s name.
- Pregnancy.

What is a Fontan procedure?

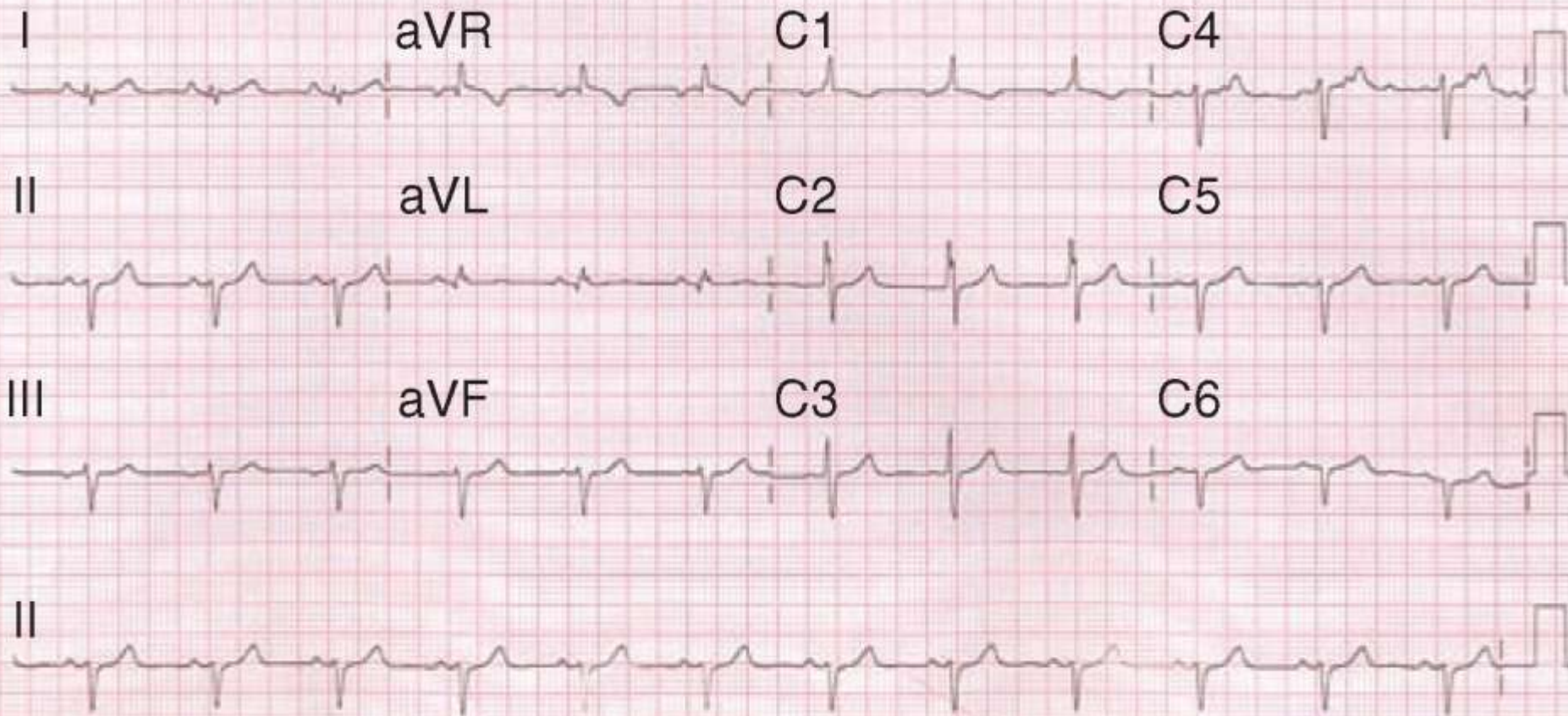
- I don't know.
- A type of transposition repair.
- A type of tetralogy repair.
- A type of single ventricle repair.

What are the pregnancy issues associated with a Fontan procedure?

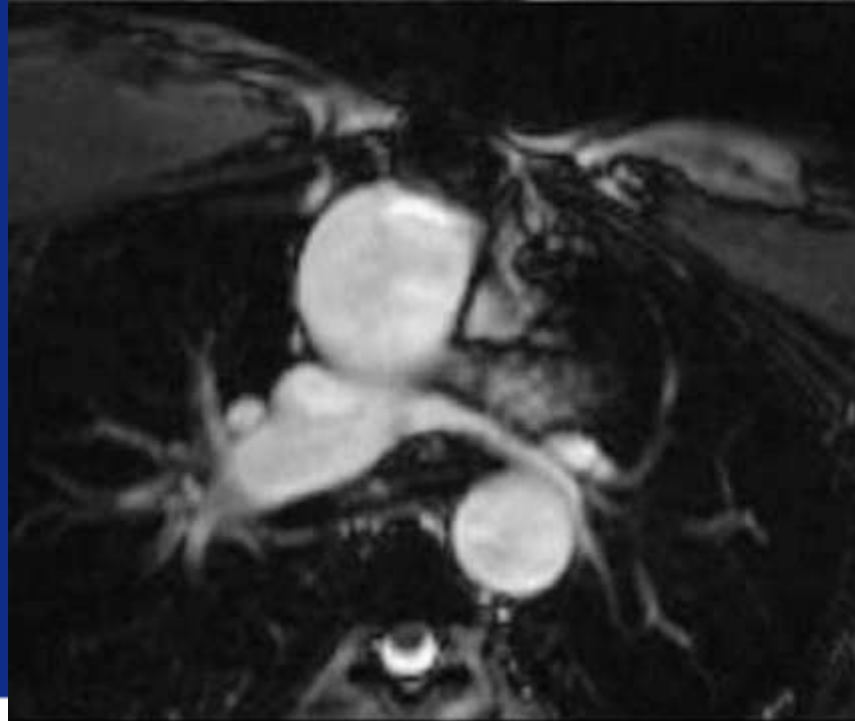
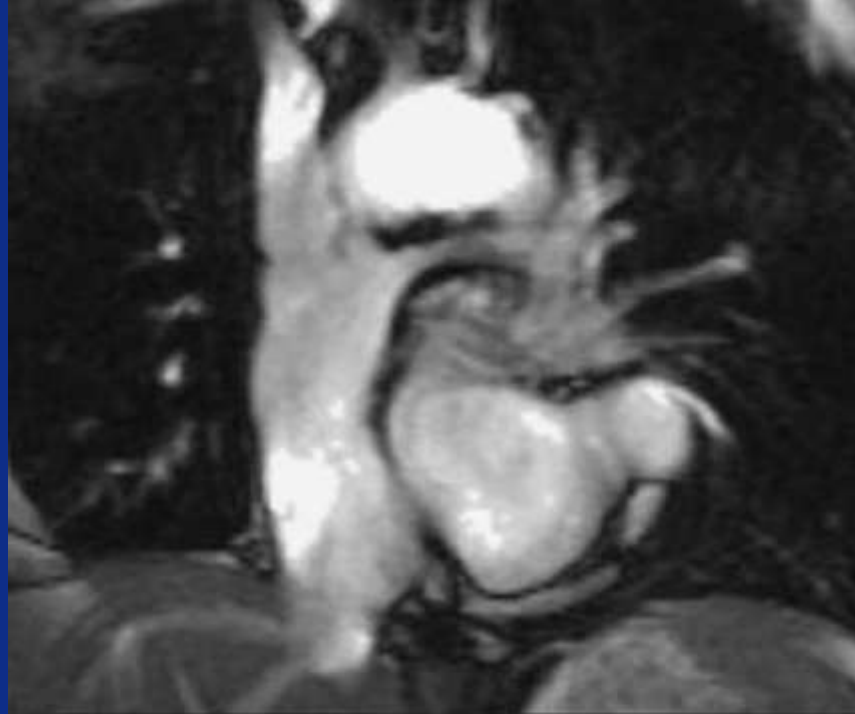
Choose as many as are applicable

- Atrial flutter.
- Thromboembolism.
- Fetal loss.
- Cyanosis.
- Maternal death.
- All of the above.

Patient 63



Patient 63



Fontan Pregnancy Management

- Close monitoring.
- High-risk pregnancy team.
- Consider anticoagulation.
- Planned vaginal delivery.
- Planned epidural.

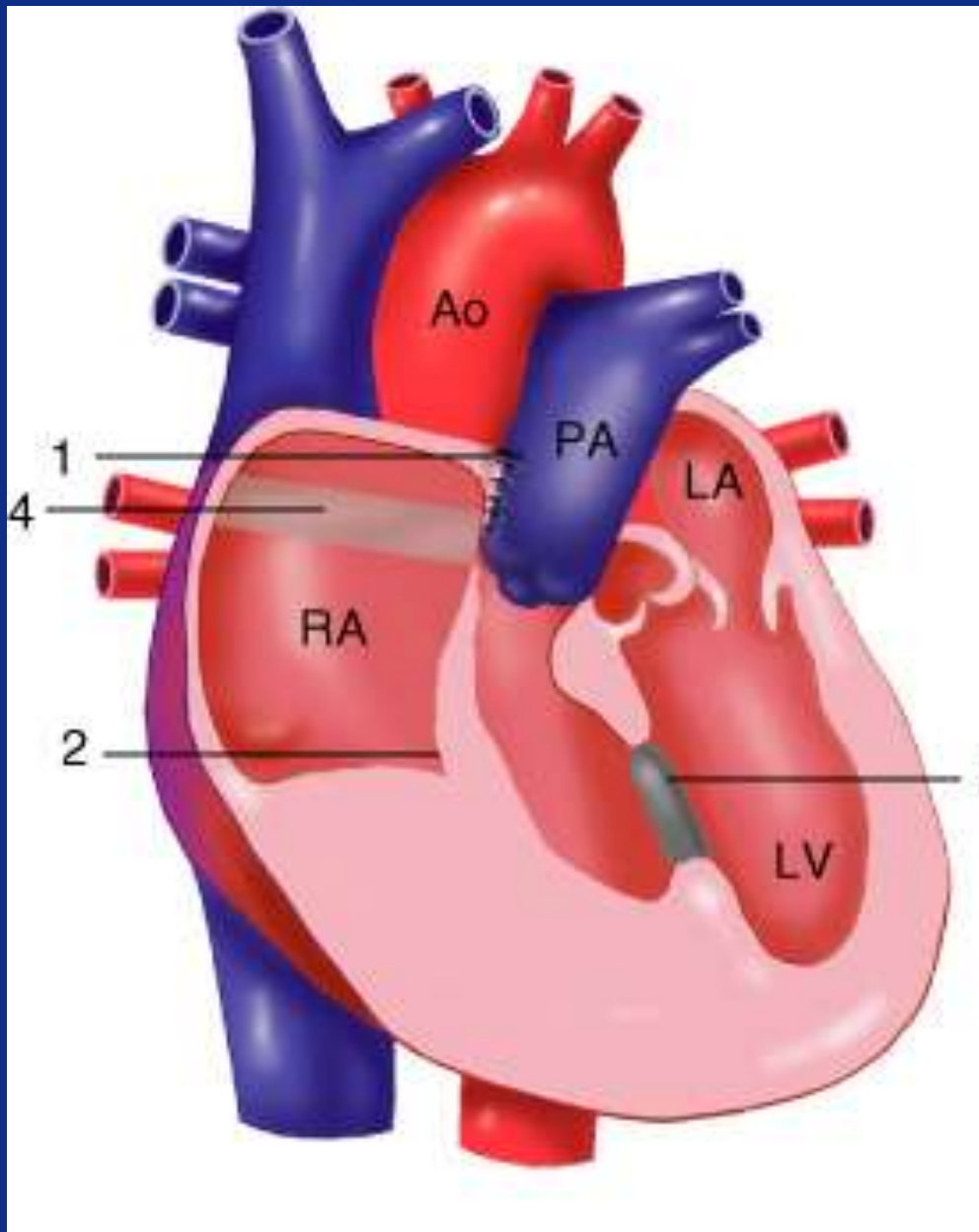
Patient 63

Patient Course and Outcome

- Induced at 39 weeks.
- Epidural anesthetic.
- Normal vaginal delivery.
- Healthy baby boy.

Patient 61 - History

- 24-year-old man, works as an engineer.
- Born with tricuspid atresia.
- Palliated with bilateral Blalock-Taussig shunts.
- "Atriopulmonary Fontan" procedure at age 6.
- Started on warfarin at age 16 for possible pulmonary embolism.



Patient 61 - History

- At age 17, developed recurrent atrial flutter.
- Treated with various antiarrhythmics, including amiodarone.
- By age 21, was in chronic atrial fibrillation.
- Was otherwise doing well (NYHA I).

Patient 61 - Physical Examination

- BP 118/72
- Pulse 70 (irregular)
- Height 68 inches
- Weight 137 pounds
- Pulse oximetry 96%

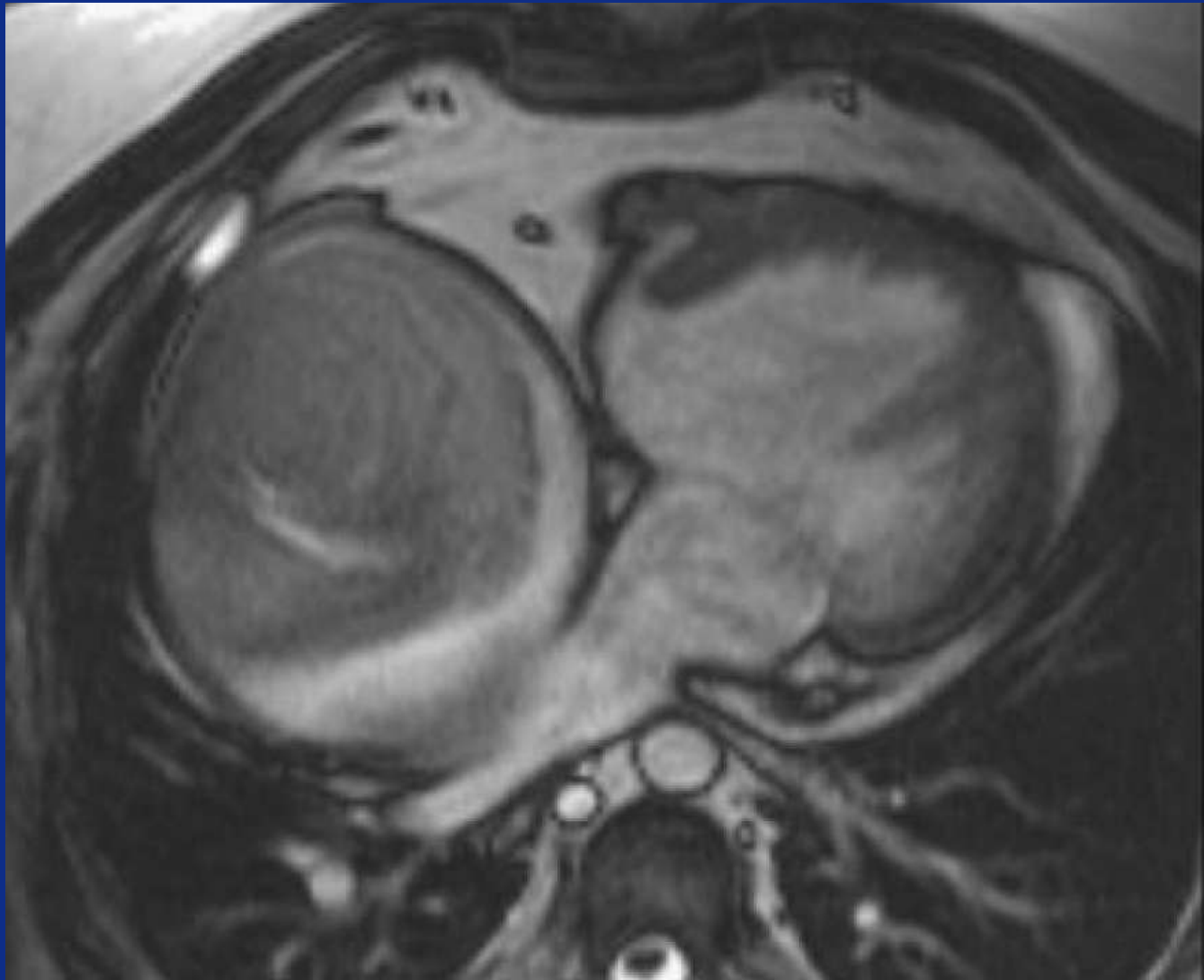
Patient 61 - Physical Examination

- Bilateral thoracotomy and midline sternotomy scars.
- JVP hard to assess.
- Chest clear.
- Normal precordial examination.
- Normal heart sounds with no murmurs.
- No hepatomegaly, ascites, or edema.

Patient 61 “Risk Factors”

- “Don’t know much about that”.
 - Fontan.
 - Blalock Taussig.
- Man’s name.
- Arrhythmia.
- One ventricle.

Patient 61



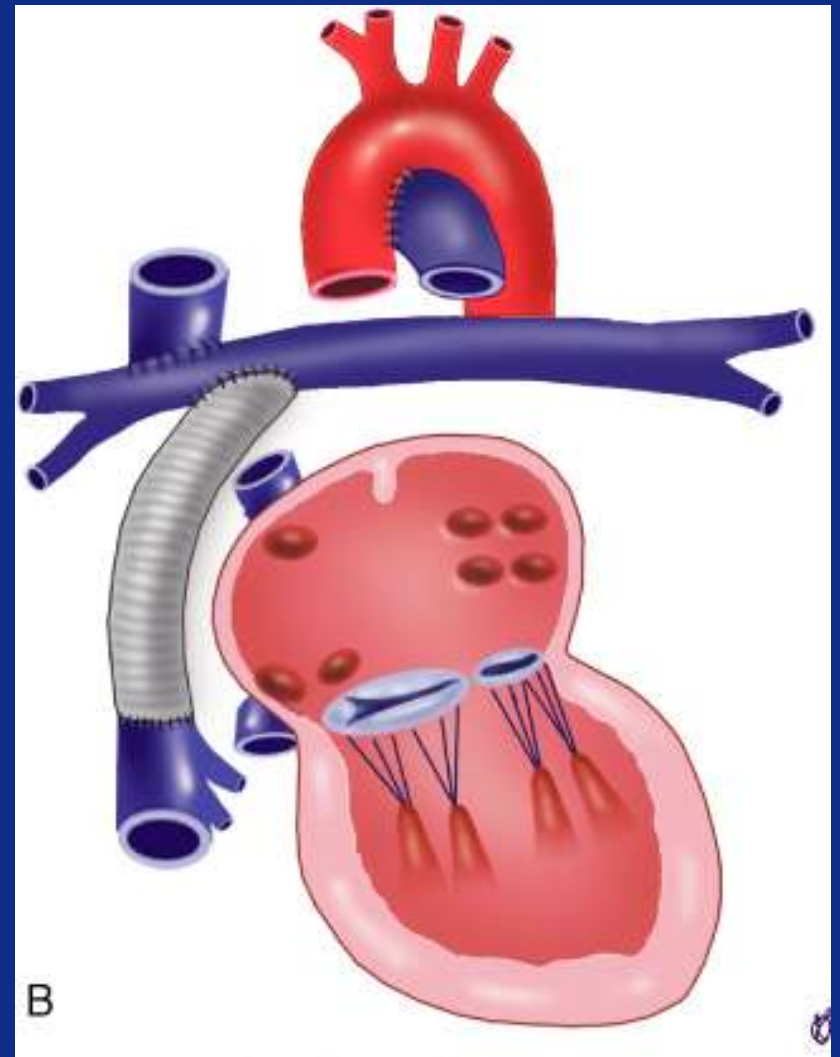
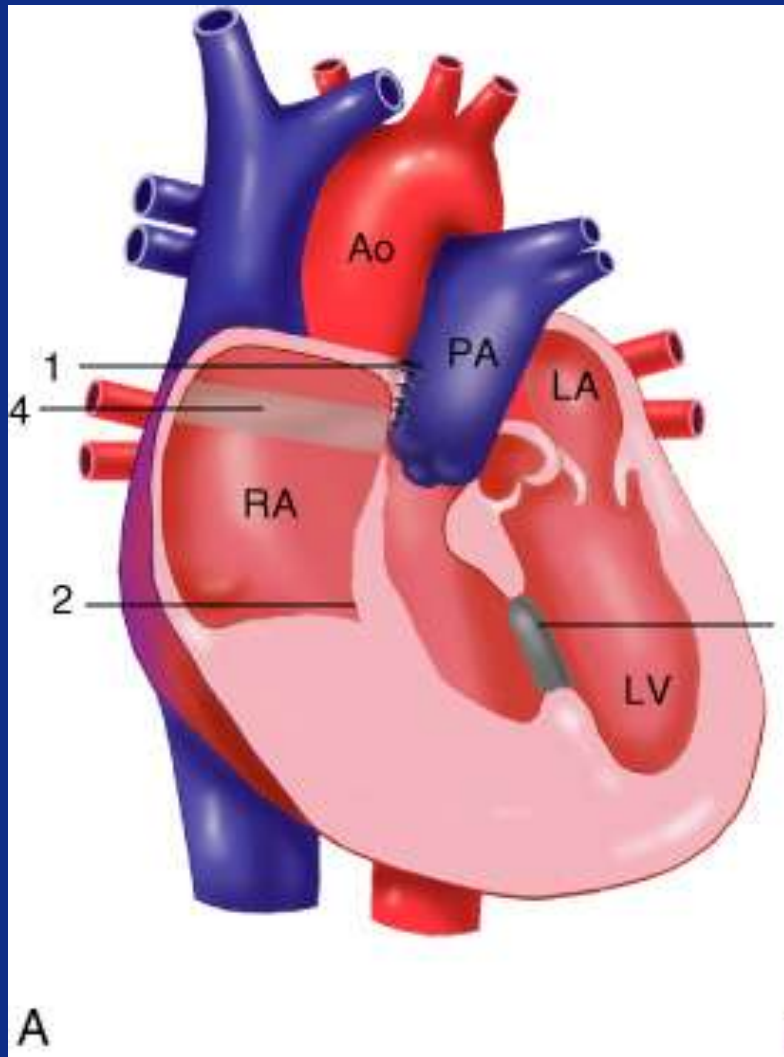
Patient 61 - What next steps?

- Optimize anticoagulation.
- Refer for immediate surgery.
- Consider thrombolysis.
- No specific treatment needed.

Patient 61

Decision and Management

- A Fontan revision was elected.
 - Right atrial thrombus removed along with excess right atrial tissue.
 - “Extracardiac Fontan” constructed.
 - Arrhythmia surgery not possible because of operative difficulties.



Patient 61

Decision and Management

- Immediately postoperatively
 - Hemodynamically unstable
 - Persistent bleeding
 - Sternal closure impossible, so skin only closure.
 - Maximum pressors.

Patient 61

Decision and Management

- Postoperative complications
 - Compartment syndrome left leg.
 - Recurrent tension pneumothorax.
 - Acute tubular necrosis
 - Tracheotomy..
 - Discharged home after four months, in atrial fibrillation, and on warfarin.

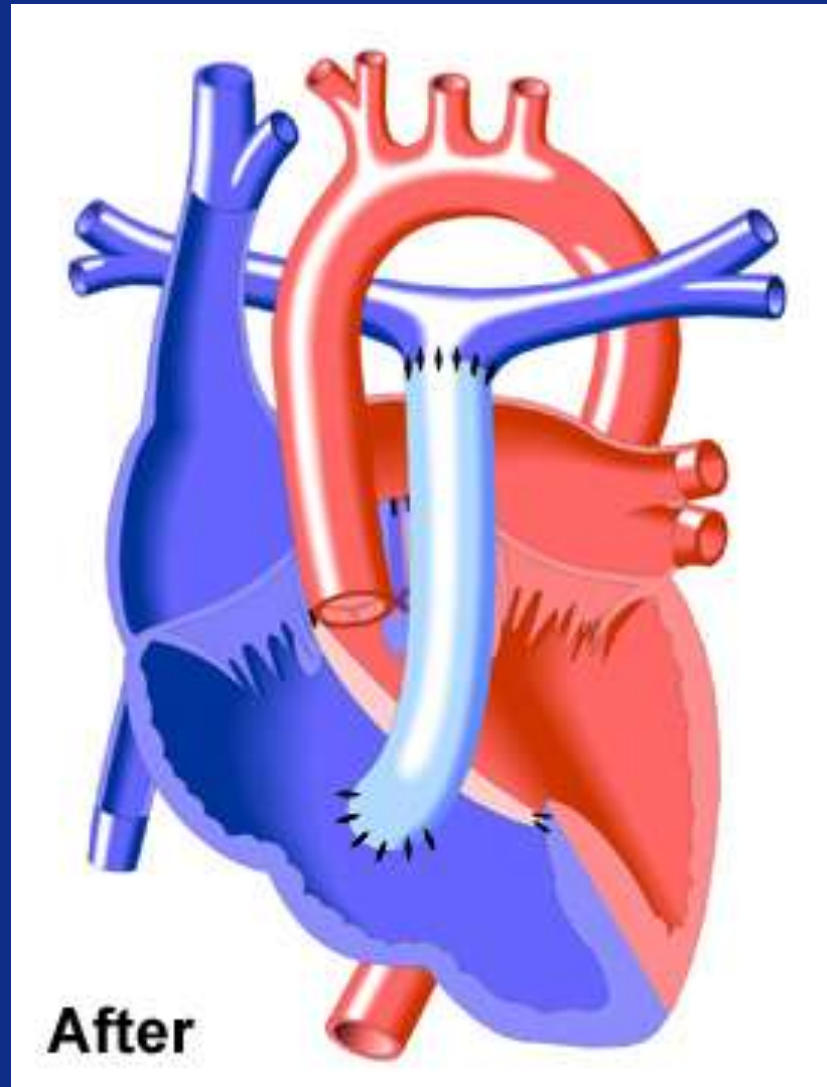
Patient 61



Patient 53 - History

- 38-year-old woman
- Original CHD diagnoses
 - TGA
 - VSD
 - PS
- Age 7, Rastelli procedure with a 20 mm Dacron conduit including a Hancock valve

Rastelli Procedure



Patient 53 - History

- Age 25, Strep sanguis endocarditis
- She was found to have severe conduit obstruction, moderate subaortic stenosis, a small residual VSD, and preserved biventricular function.
- Her conduit was replaced using a homograft, her VSD was enlarged to reduce the LVOT gradient, and a baffle was placed between LV and the aorta.

Patient 53 - History

- Age 35, heart catheterization showed a 40 mmHg gradient across the RV-PA conduit with mild LVOTO and mild AR. Sotalol was started for suspected paroxysmal atrial tachycardia.
- Age 38, NYHA class III with exertional dyspnea and fatigue. She is limited to 100 yards on the flat. She avoids steps.
- Current medications: warfarin; sotalol 80 mg BID; furosemide 20 mg daily; sertraline 100 mg daily

Patient 53 - Physical Examination

- BP 110/70
- Pulse 46
- Height 70 inches
- Weight 209 pounds
- Pulse oximetry 97%

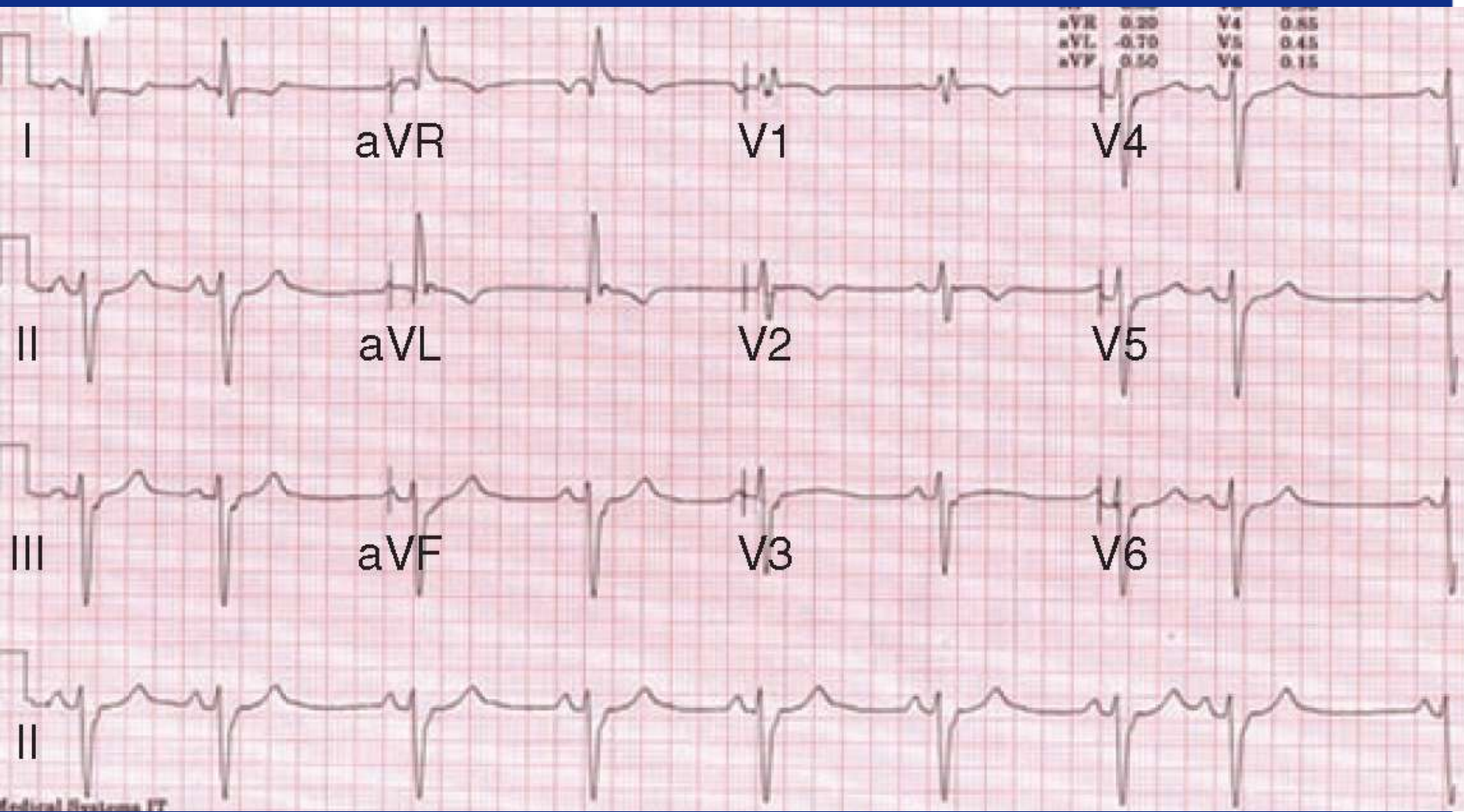
Patient 53 - Physical Examination

- JVP 8 cm ASA.
- Chest clear.
- Possible right ventricular lift.
- Second sound loud and single.
- Grade 4 long ejection systolic murmur upper left sternal border.
- No hepatomegaly or edema.

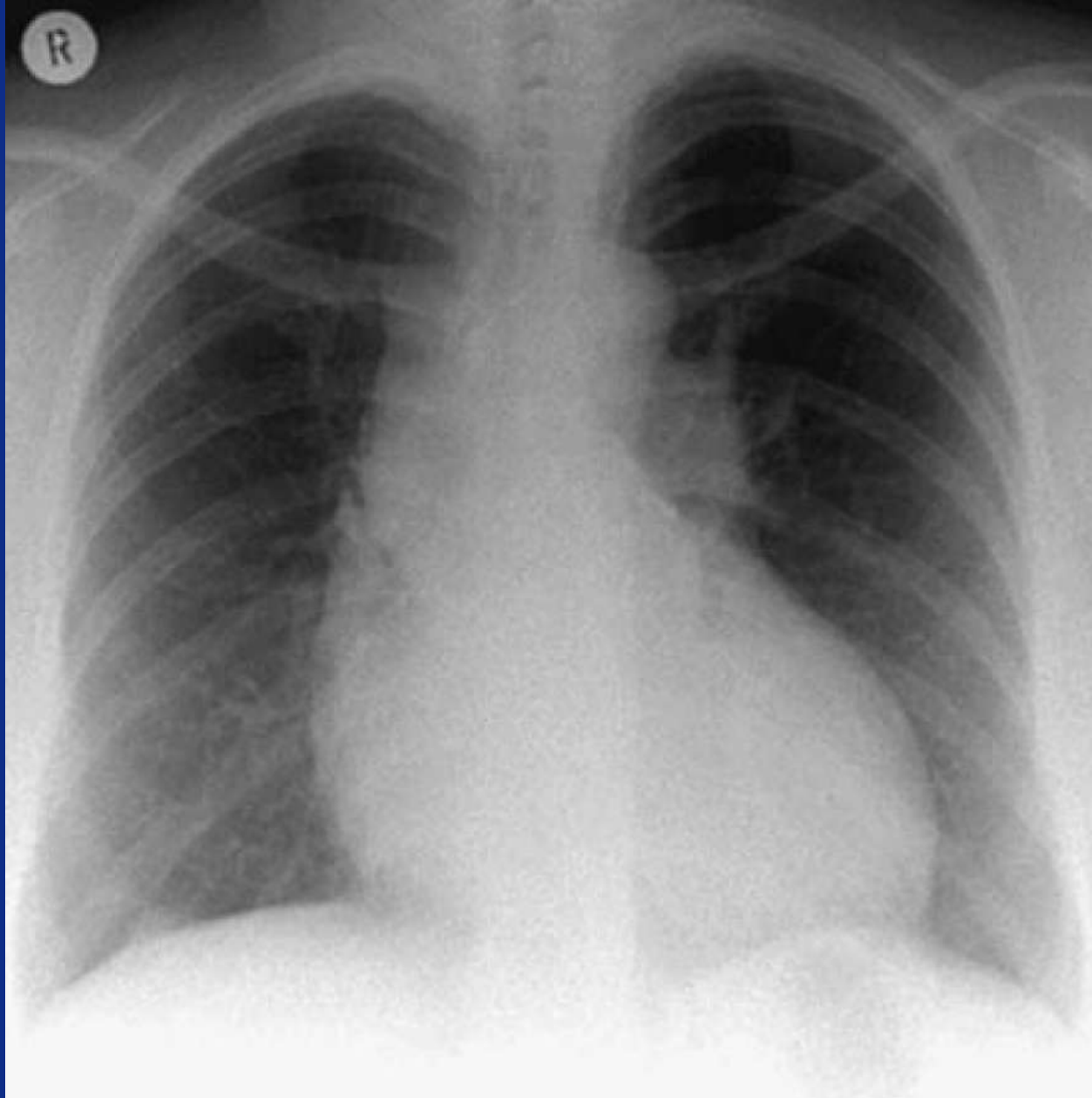
How much conduit obstruction does she have?

- Mild.
- Moderate.
- Severe.
- Don't know.

Patient 53



**Patient
53**



Patient 53 - Echo Report

- Normal LV size and function.
- Peak LVOT gradient 20 mmHg.
- RVH with mild-moderate hypokinesis.
- Mild TR.
- Valved conduit not seen.

Patient 53 – “Risk Factors”

- “Don’t know much about that”.
 - TGA.
 - VSD/PS.
- Man’s name.
- Exercise intolerance.
- Sexually active.

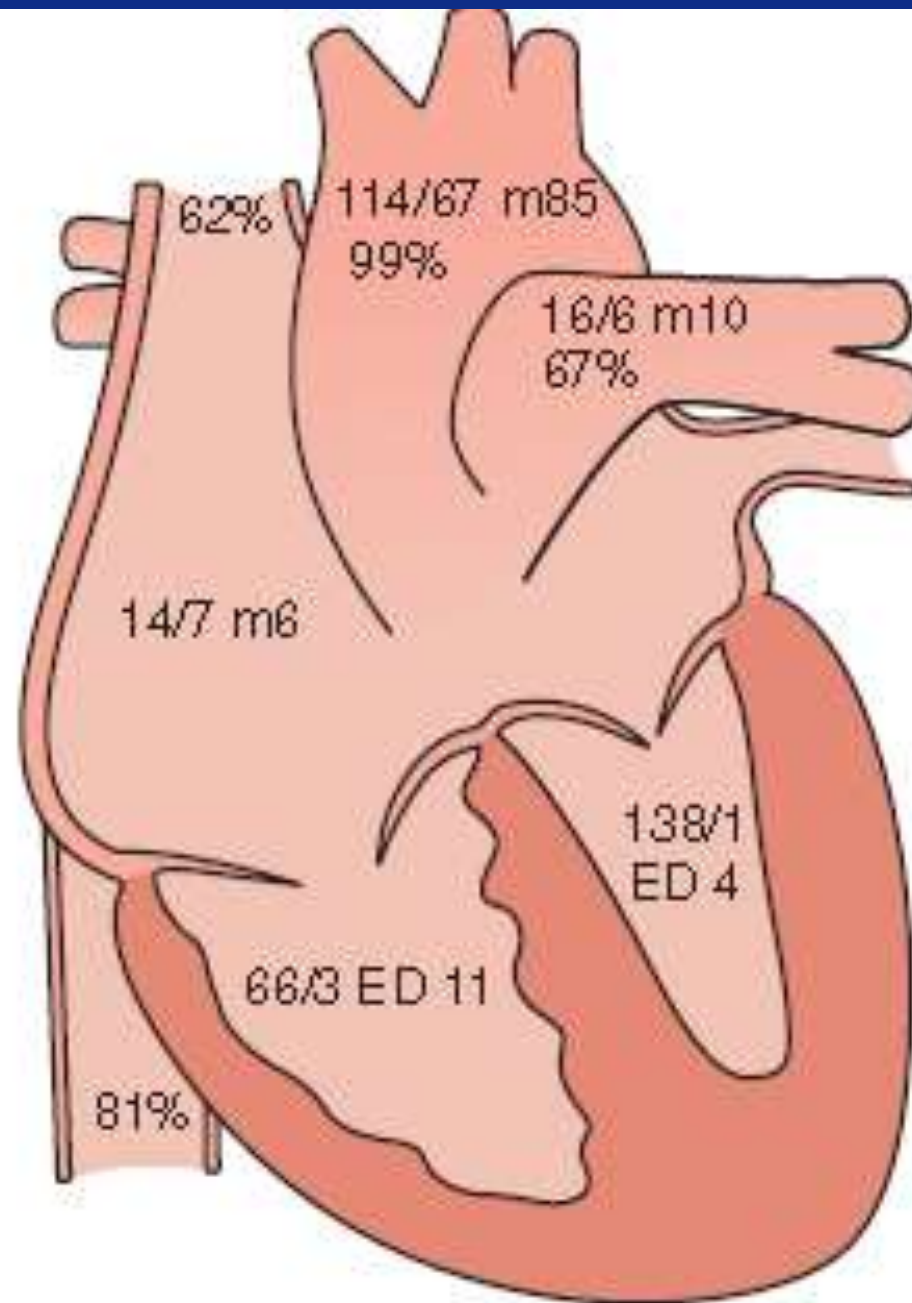
**Patient
53**



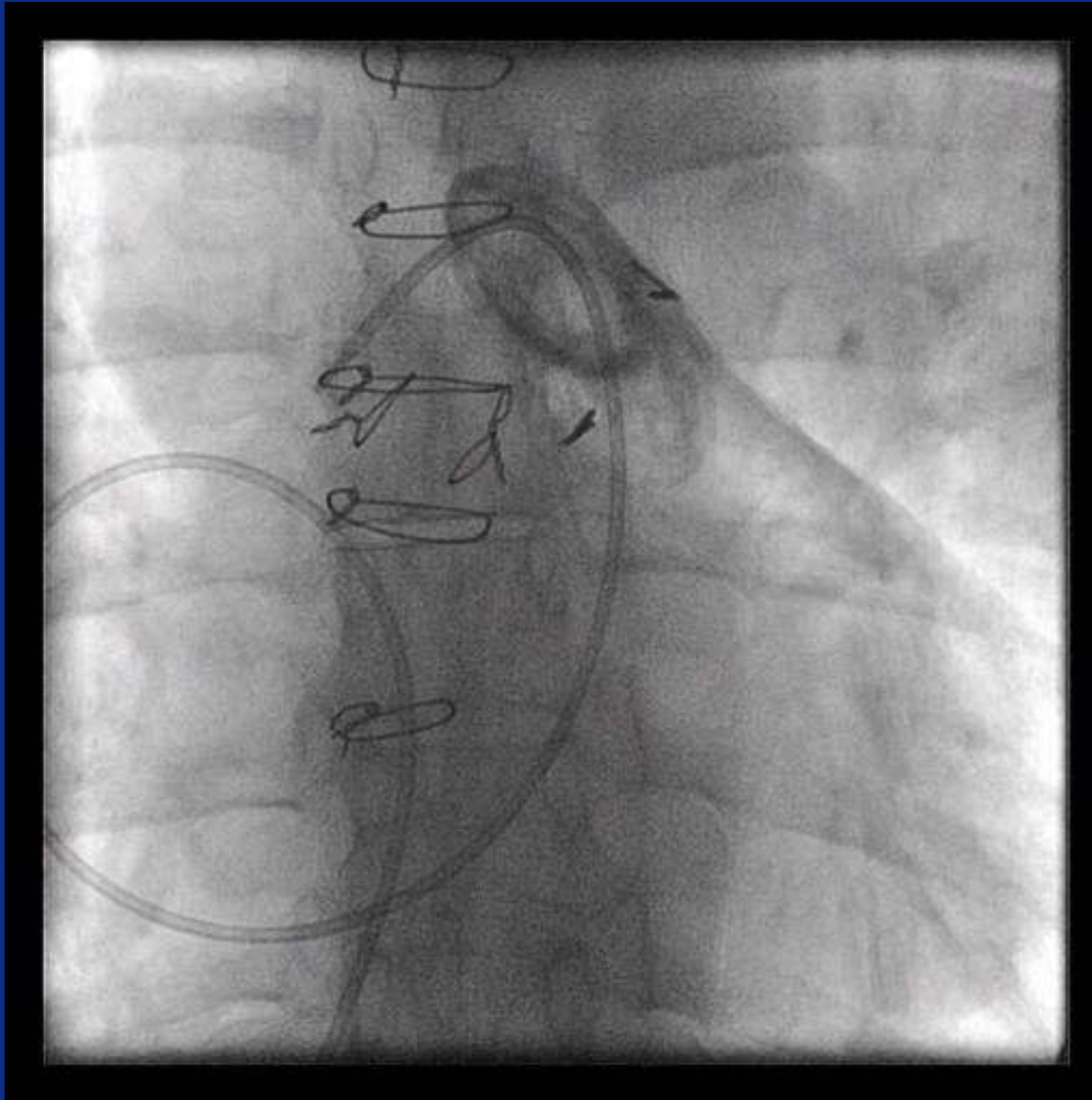
**Patient
53**



Patient 53



Patient 53



Patient 53

Why is she NYHA class III?

Choose as many as are applicable

- The stenosed conduit?
- The sotalol?
- Chronotropic incompetence?
- Anemia?
- Lung disease?

Patient 53 – Management and Outcome

- Conduit from RV to PA removed and replaced.
- Smooth postop course.
- Patient NYHA I after convalescence.

Patient 41 - History

- 31-year-old woman
- Original CHD diagnoses
 - Tetralogy of Fallot
- Age 3, transannular patch repair.
- Progressive pulmonary regurgitation, severe.
- Asymptomatic and full time work, but has “slowed down” a bit.

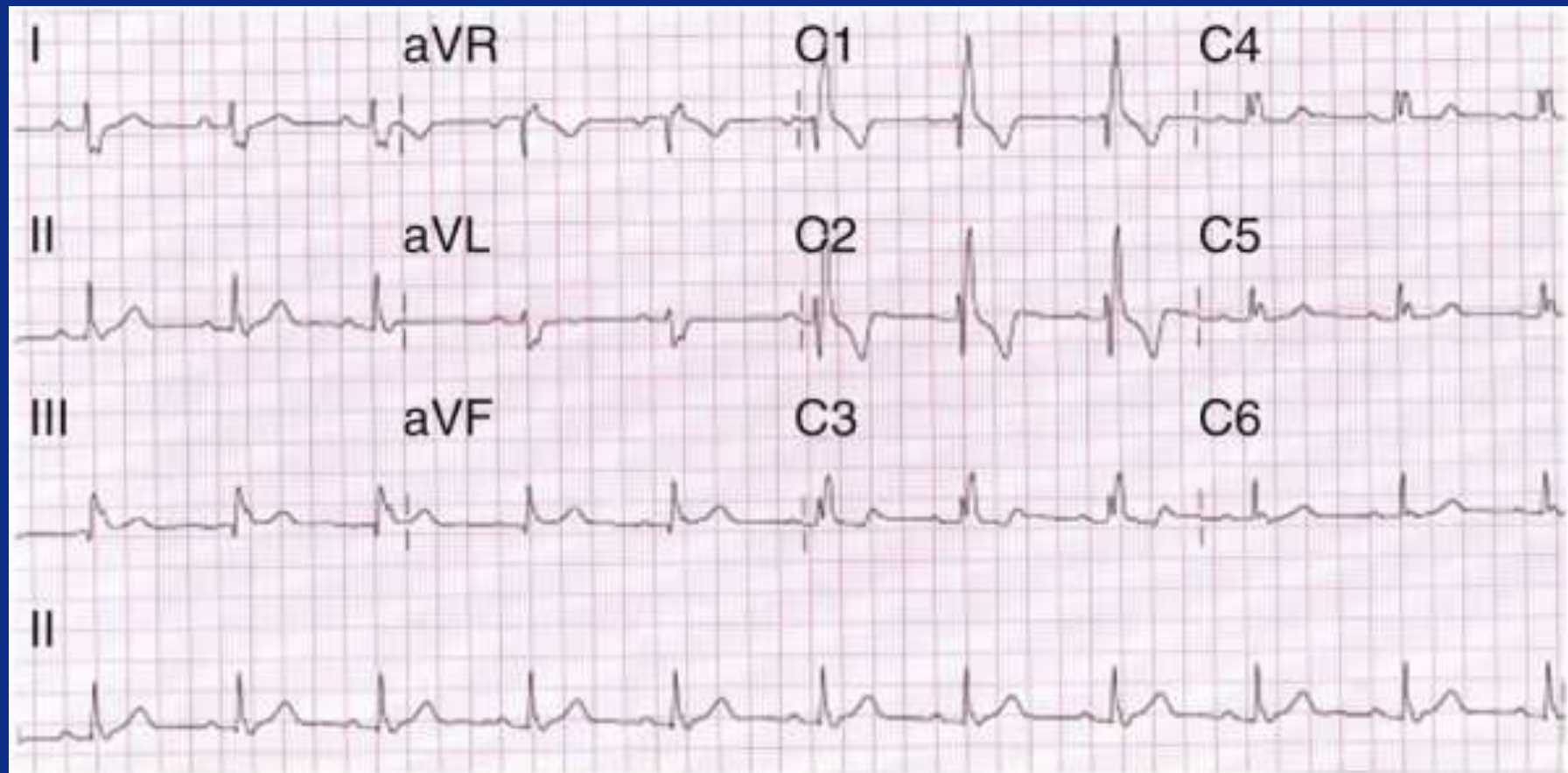
Patient 41 - Physical Examination

- BP 110/70
- Pulse 70
- Height 62 inches
- Weight 189 pounds
- Pulse oximetry 100%

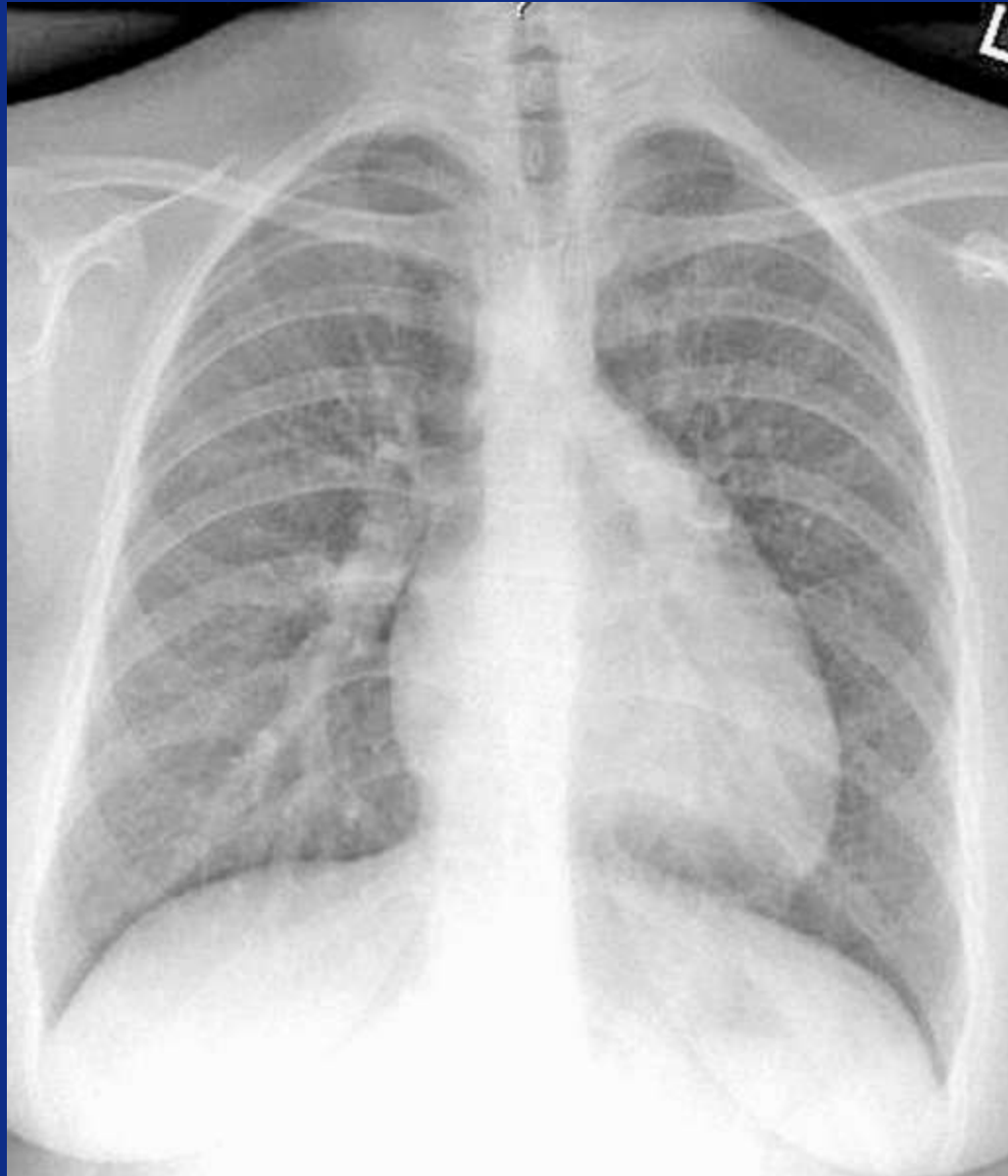
Patient 41 - Physical Examination

- Midline sternotomy scar.
- Normal JVP.
- RV heave.
- Grade 2 ejection systolic murmur.
- Grade 2 low pitched diastolic murmur.

Patient 41



Patient 41



Patient 41 - Echo Report

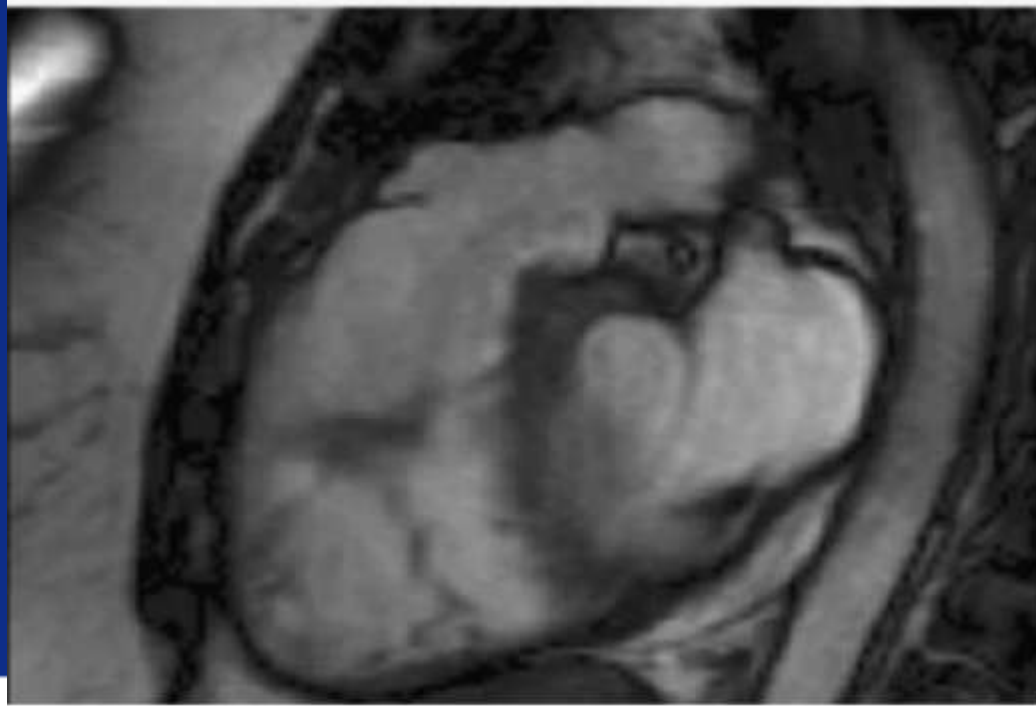
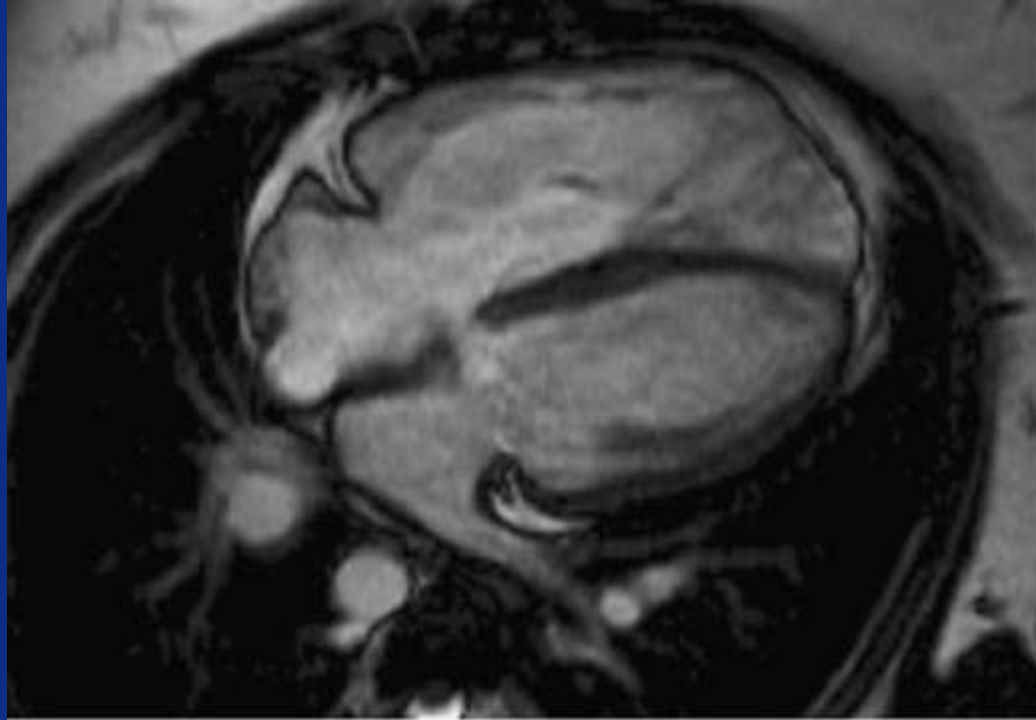
- Normal LV size and function.
- Right ventricle severely dilated.
- Laminar (severe) pulmonary regurgitation.

Patient 41 – “Risk Factors”

- “Don’t know much about that”.
 - Tetralogy.
 - Pulmonary regurgitation.
- Man’s name.
- Candidate for a heart catheterization.

Patient 41

Patient 41



Patient 41

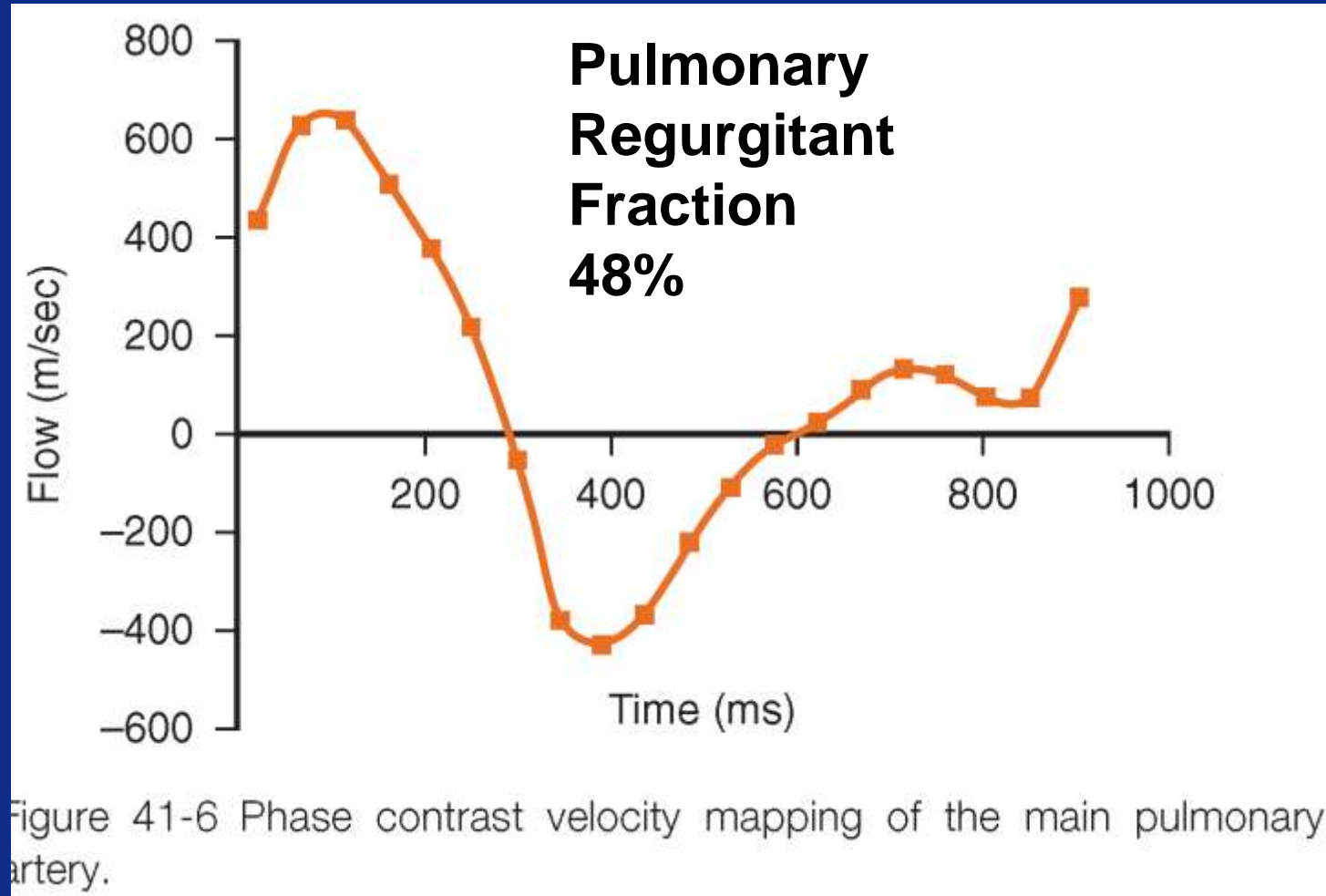
Ventricular Volume Quantification

	LV	(Normal range)	RV	(Normal range*)
EDV (mL)	116	(52–141)	244	(58–154)
ESV (mL)	32	(13–51)	88	(12–68)
SV (mL)	84	(33–97)	156	(35–98)
EF (%)	72%	(57–75)	64%	(51–75)
EDVi (mL/m ²)	60	(62–96)	126	(61–98)
ESVi (mL/m ²)	16	(17–36)	45	(17–43)
SVi (mL/m ²)	43	(40–65)	80	(38–62)
Mass index (g/m ²)	47	(47–77)	49	(20–40)

*Normal values based on gender, age, and body surface area.

Adapted from Maceira A, Prasad SK, Khan M, Pennell DJ: Normalized left ventricular systolic and diastolic function by steady state free precession cardiovascular magnetic resonance. *J Cardiovasc Mag Res* 8:417–426, 2006.

Patient 41



Patient 41

Treatment Options?

Choose as many as are applicable

- Bioprosthetic pulmonary valve replacement.
- Mechanical pulmonary valve replacement.
- Percutaneous pulmonary valve replacement.
- Continued observation.

Patient 41 – Plan and Outcome

- Pulmonary valve replacement elected.
- 23 mm homograft used (bioprosthesis).
- Previous patch excised.
- Exercise intolerance for improved after convalescence.
- RV volumes reduced by 30% on follow-up MRI.

Patient 25 - History

- 45-year-old man
- Original CHD diagnoses
 - Aortic coarctation (isolated)
- Age 3, coarctation repair (details unknown).
- Recently found to have high blood pressure.
- Treated with an ACE inhibitor and a beta blocker.
- No cardiovascular symptoms.

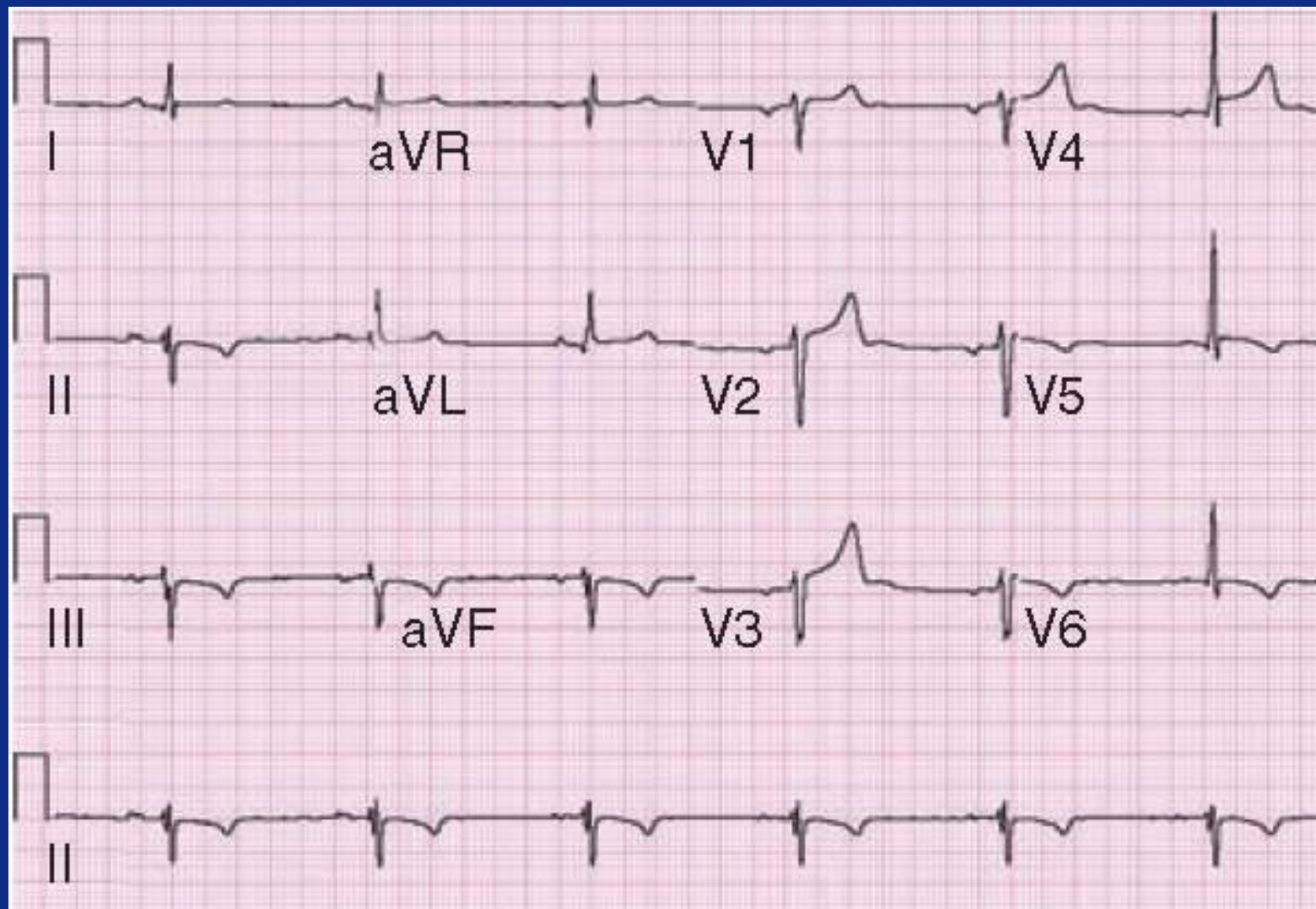
Patient 25 - Physical Examination

- BP 156/70 right arm
 115/60 left arm
 118/? right thigh
- Pulse 44
- Height 70 inches
- Weight 202 pounds
- Pulse oximetry 98%

Patient 25 - Physical Examination

- Left thoracotomy scar.
- Pulse delay right radial to right femoral.
- Very weak pedal pulses.
- Grade 2 ejection systolic murmur.
- Grade 2-3 murmur of aortic regurgitation.
- Faint continuous murmur heard over the back.

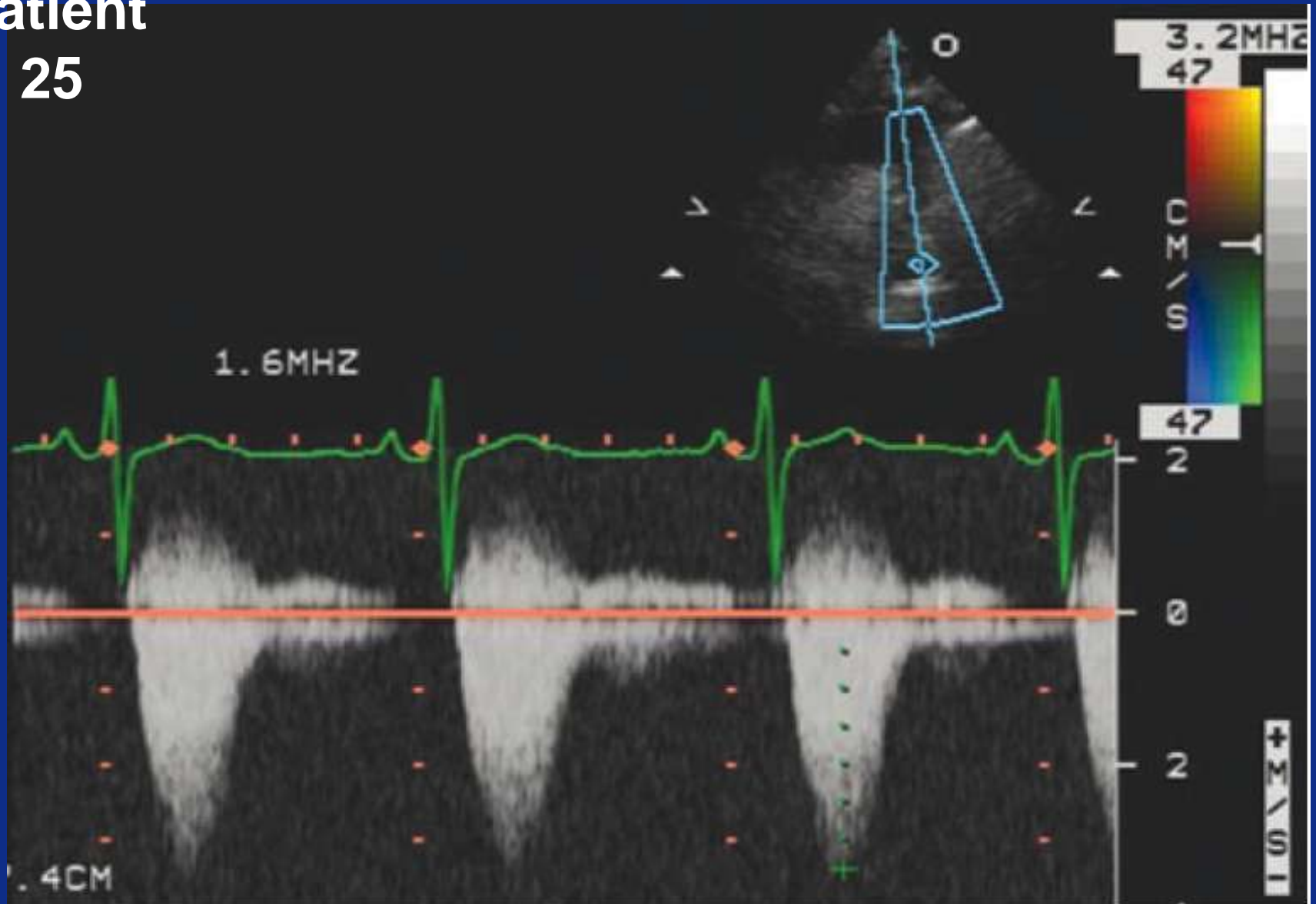
Patient 25



Patient 25



Patient 25



Patient 25 - MRI Report

- Mild dilation of the ascending aorta.
- Hypoplastic distal aortic arch with severe coarctation.
- Collateralization present.

Patient 25 - MRI



Patient 25 – “Risk Factors”

- Possible candidate for cardiovascular surgery.
- Possible candidate for a heart catheterization.

Patient 25

Treatment Options?

Choose as many as are applicable

- Surgical repair of aortic coarctation.
- Balloon dilation of aortic coarctation.
- Stent placement for aortic coarctation.
- More aggressive antihypertensive therapy.

Patient 25 – Plan and Outcome

- Stent therapy of aortic recoarctation elected.



Patient 25 – Plan and Outcome

	BP right arm	BP left arm	BP right leg
Prior to procedure	156/70	115/60	118/?
3 weeks after procedure	108/65	113/70	132/80

Patient 25 – Plan and Outcome



Case 19

Patient 19 - History

- 26-year-old woman
- Original CHD diagnoses
 - Normal heart
 - Infective endocarditis
- Age 9 and 10, aortic valve repair for aortic regurgitation.
- Age 18, Ross procedure.
- Age 26, declining exercise capacity, limited to two floors vertically.

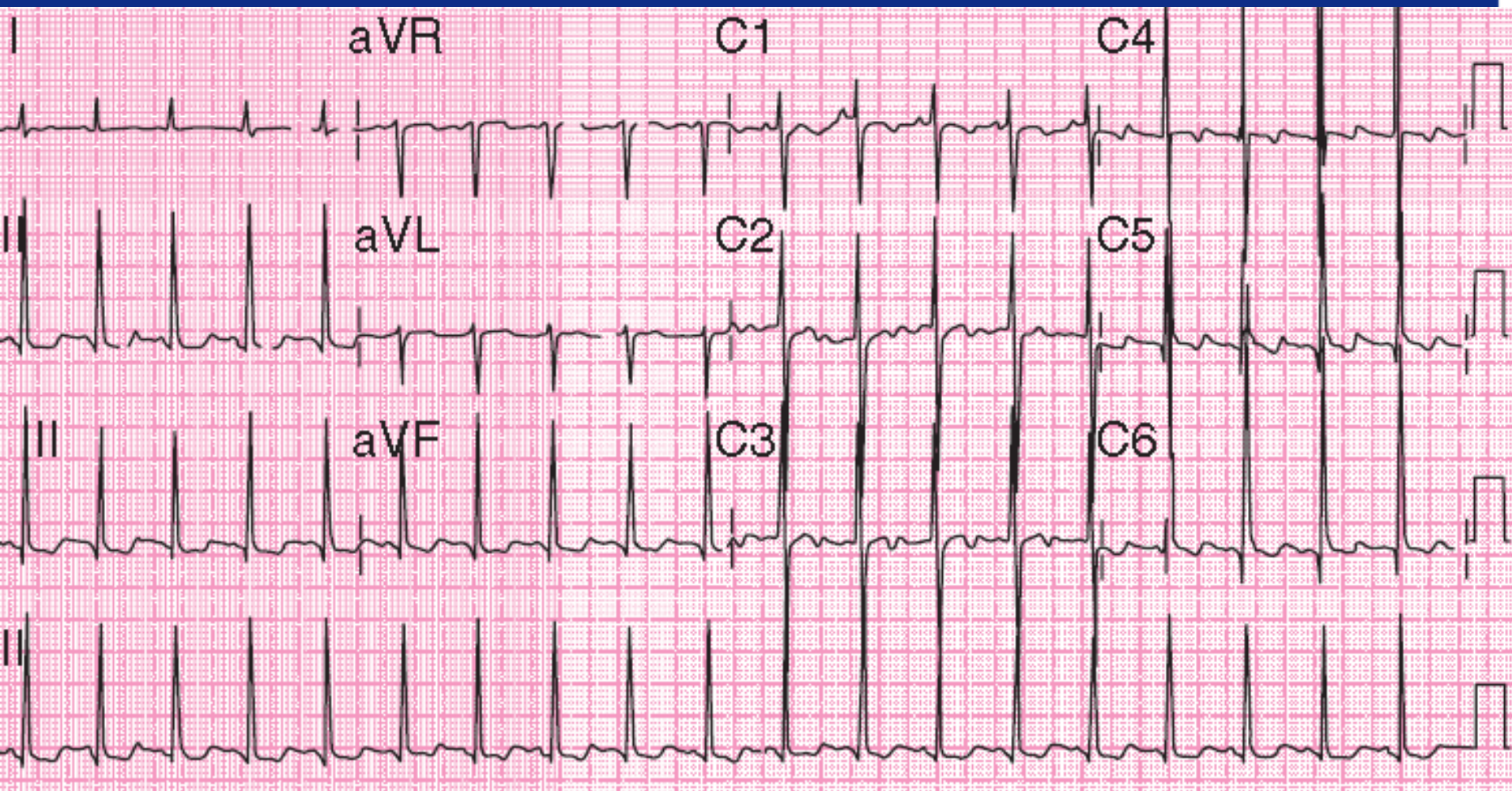
Patient 19 - Physical Examination

- BP 100/42
- Pulse 100
- Height 70 inches
- Weight 147 pounds
- Pulse oximetry 100%

Patient 19 - Physical Examination

- Bounding peripheral pulses.
- Positive Duroziez sign.
- Cardiac apex displaced and active.
- Grade 2 medium length ejection systolic murmur.
- Grade 2 high-pitched diastolic murmur LSB.

Patient 19



Patient 19



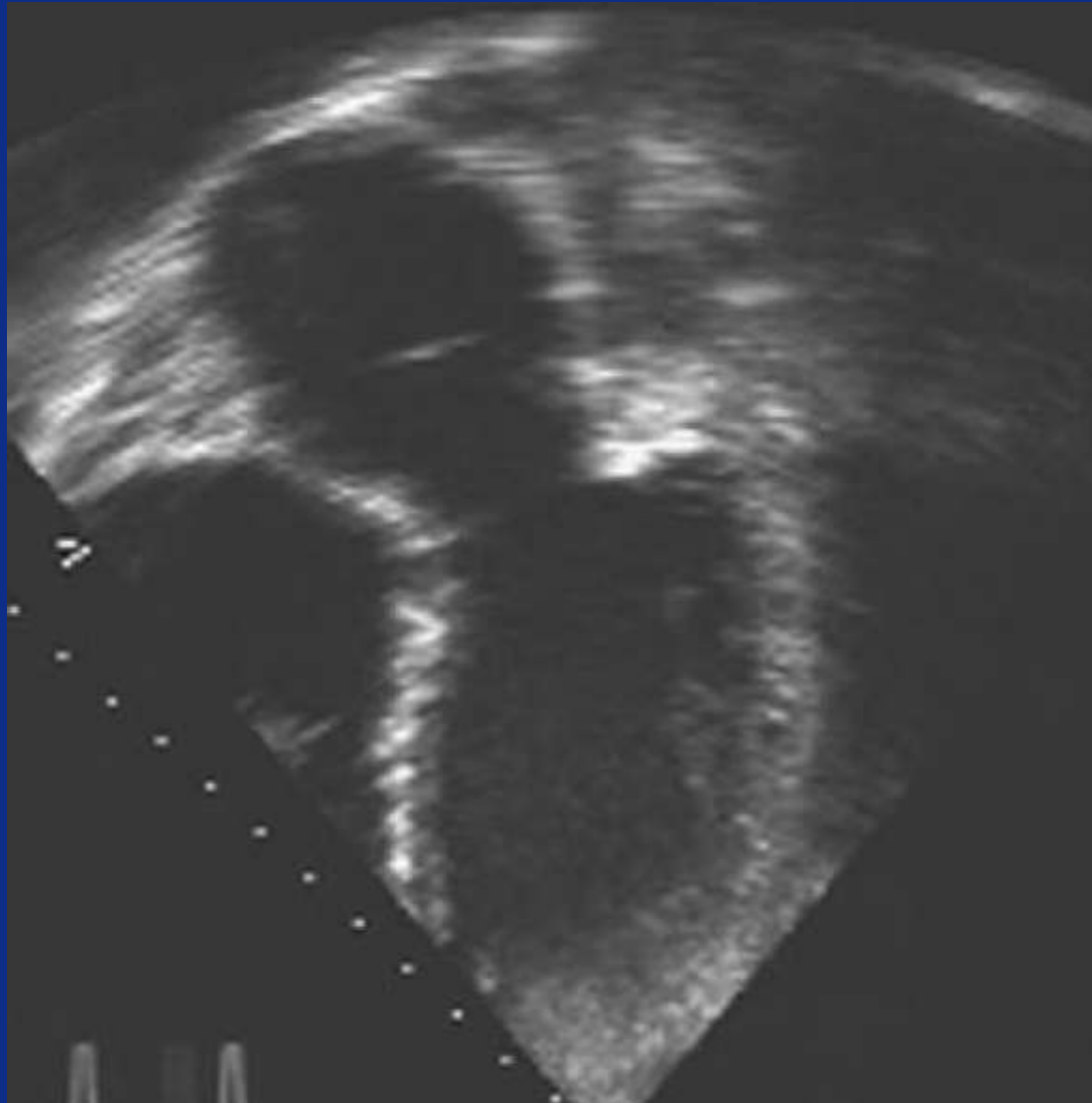
Patient 19



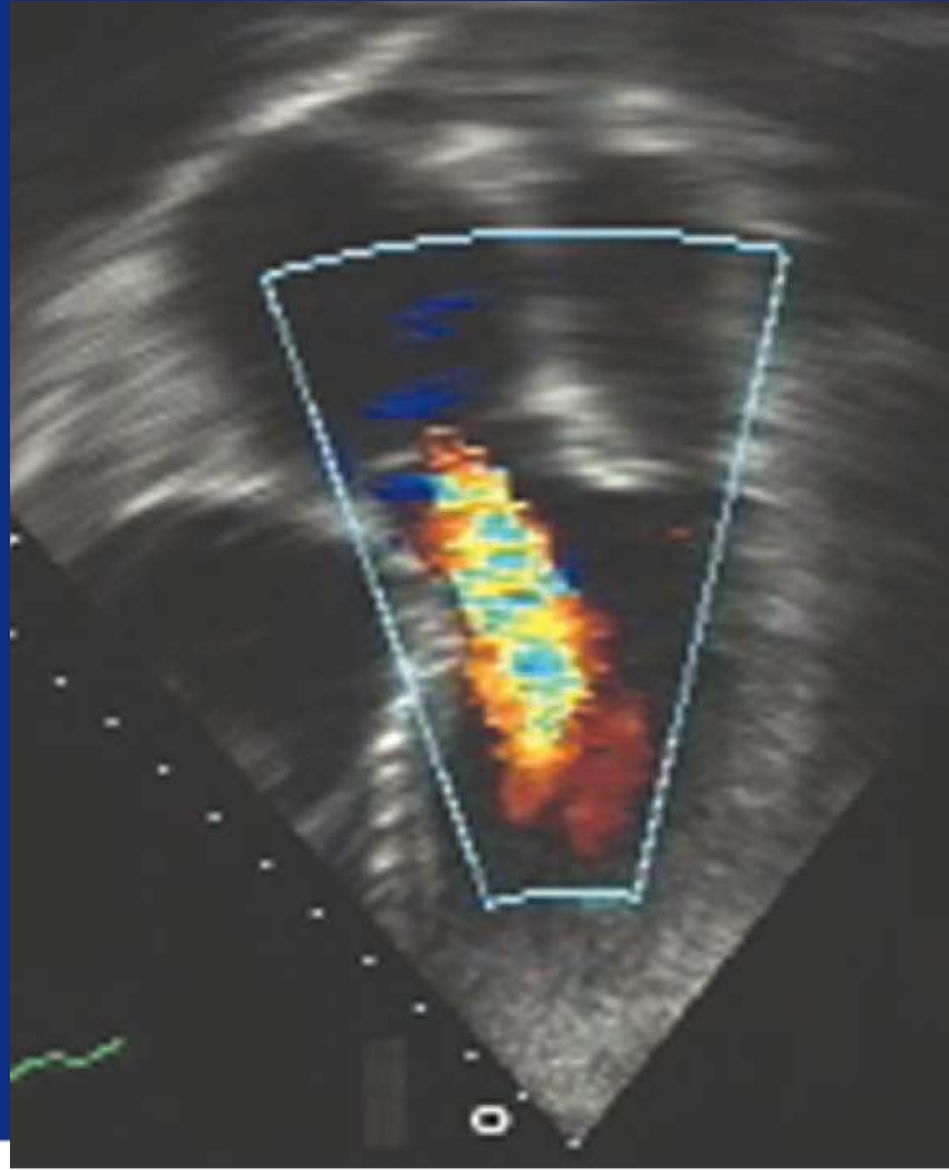
Patient 19 - Echo Report

- Left ventricle dilated to 62 mm diastolic.
- Normal LV systolic function.
- Ascending aortic dilation to 55 mm.
- Severe neoaortic valve regurgitation.
- Homograft function normal on the right side of the heart.

Patient 19 - Echo



Patient 19 - Echo



Patient 19 – “Risk Factors”

- CHD patients needing special cardiac surgery.
 - David operation.
 - Ross procedure.
 - Aortic valve repair?

Patient 19 - Which management would you consider?

Choose as many as are applicable

- Bentall procedure with mechanical aortic valve.
- Bentall procedure with bioprosthetic aortic valve.
- Valve sparing root replacement.

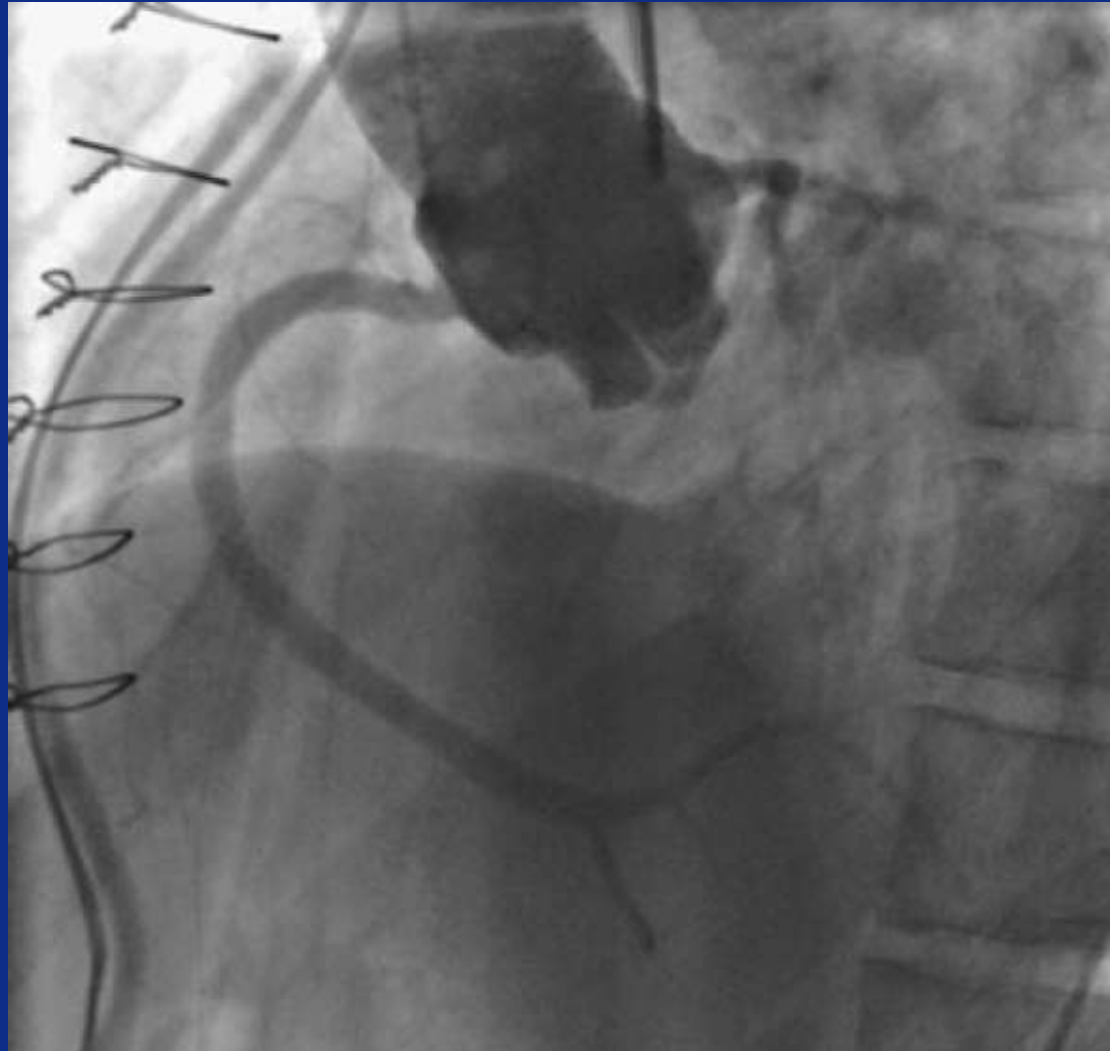
Patient 19 – Management and Outcome

- Valve sparing aortic root replacement elected.
- At surgery, the neo-aortic valve leaflets appeared normal.
- The aortic annulus was less dilated than were the sinuses.
- The aorta was replaced using a David technique, sewing a 26 mm Dacron graft with preformed sinuses down to the annulus with resuspension of the neo-aortic leaflets.
- The coronary arteries were reimplanted.

Patient 19 – Management and Outcome

- The patient did well with no residual aortic valve regurgitation.

Patient 19 – Management and Outcome



Case 6

Patient 6 - History

- 34-year-old woman
- Generally healthy childhood.
- Told that her heart was on the right side.
- Occasional episodes of hemoptysis as a teenager.
- Hemoptysis recurred after a recent successful pregnancy.
- Lifelong non-smoker.
- Very good exercise capacity.

Patient 6 - Physical Examination

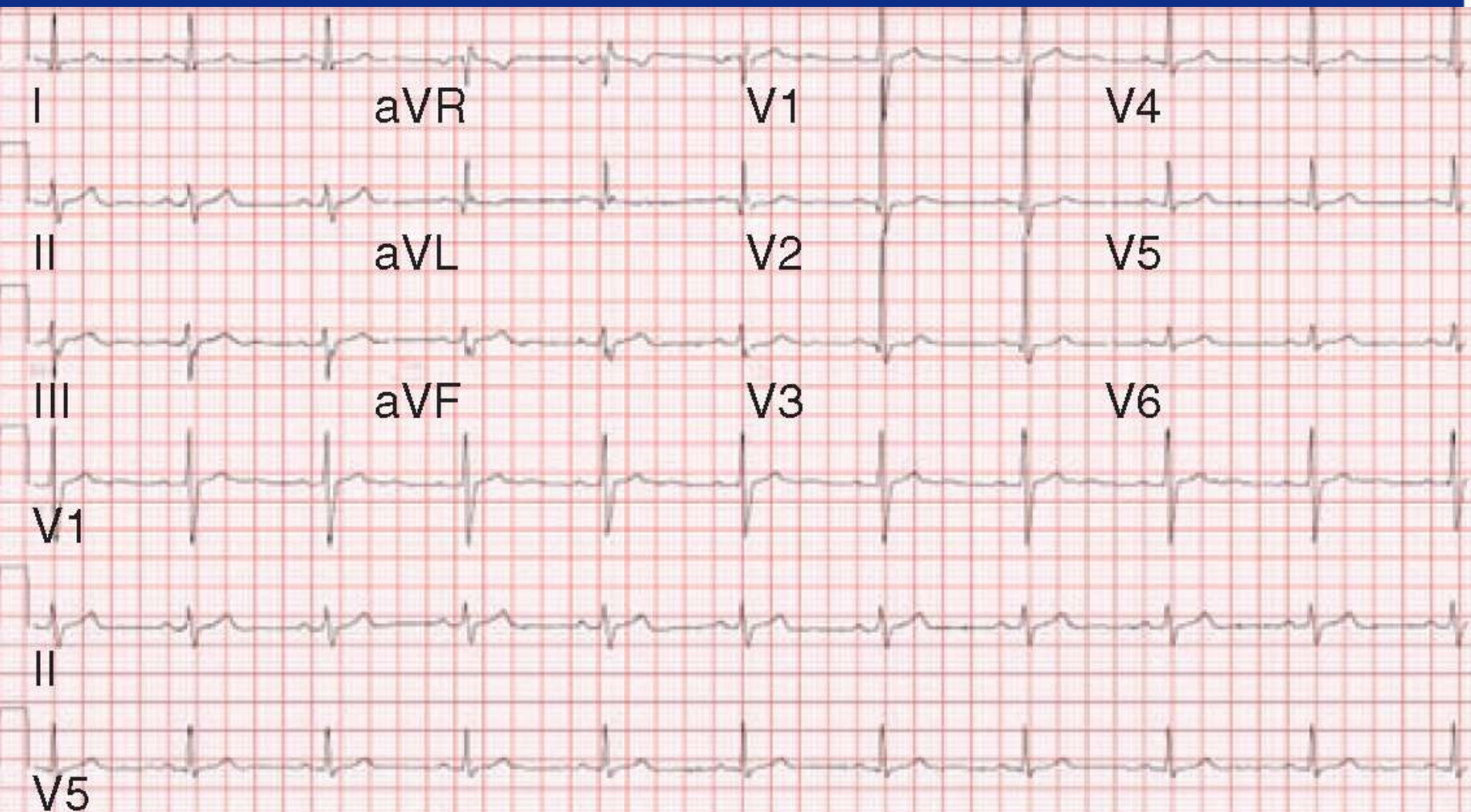
- BP 100/68
- Pulse 62
- Height 56 inches
- Weight 141 pounds
- Pulse oximetry 98%

Patient 6 - Physical Examination

- Normal JVP.
- Clear chest.
- Right-sided apical impulse.
- Heart sounds normal, no murmurs.
- Right-sided liver.

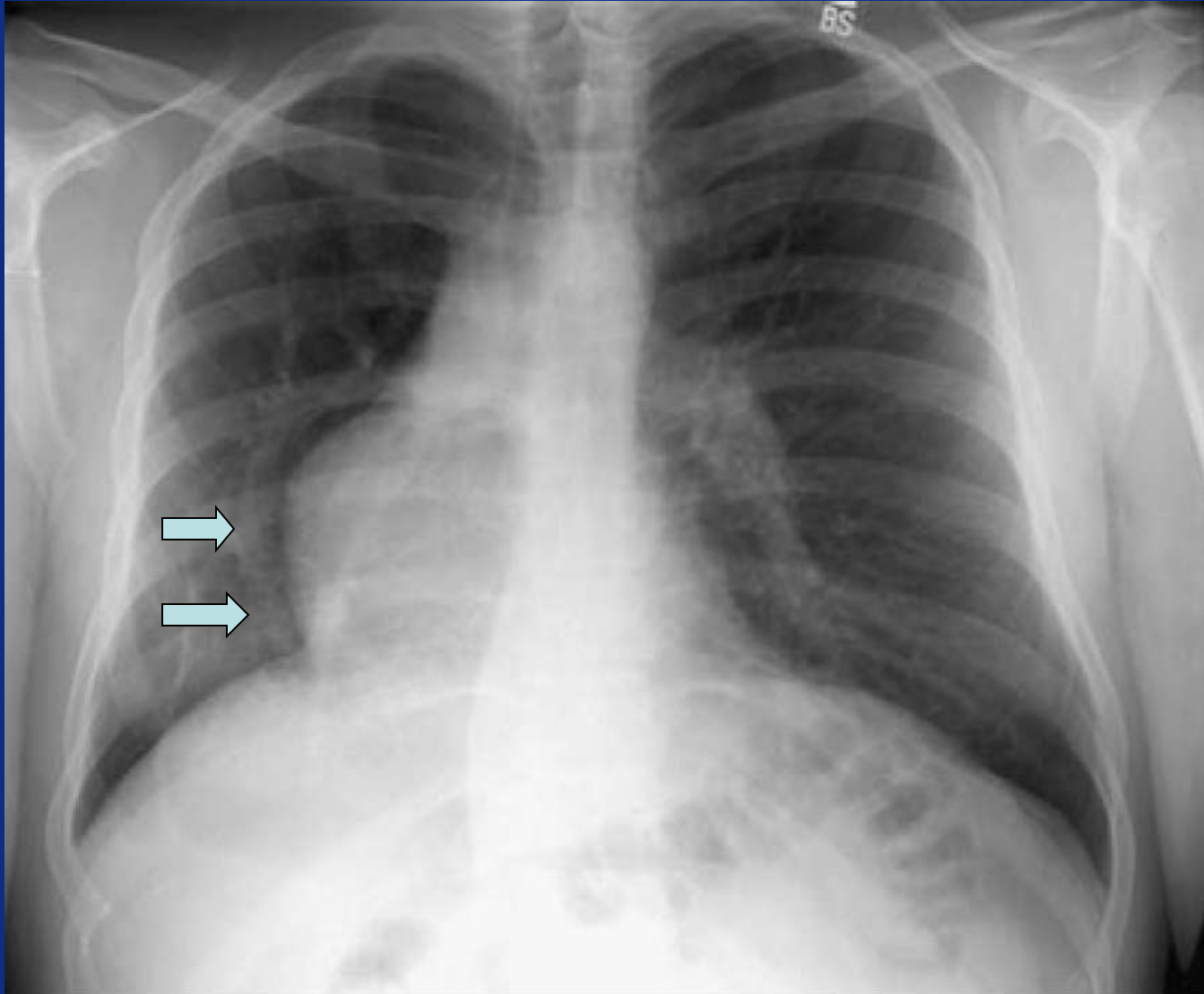
Patient

6



Patient

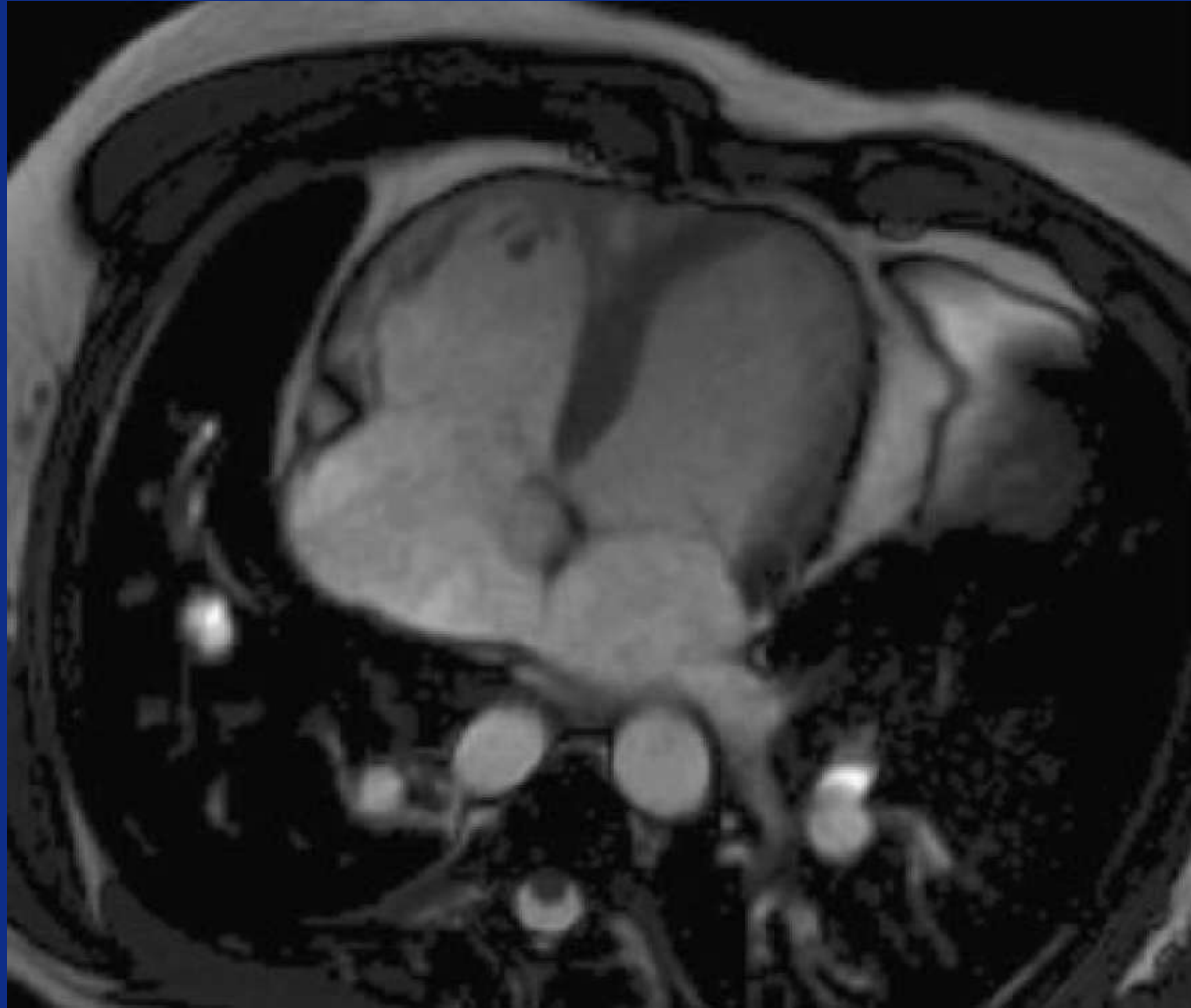
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Patient 6 - Echo Report

- Dextroposition of the heart with normal cardiac chamber anatomy.
- Mild right heart dilation.
- Normal RVSP.

Patient 6 - MRI

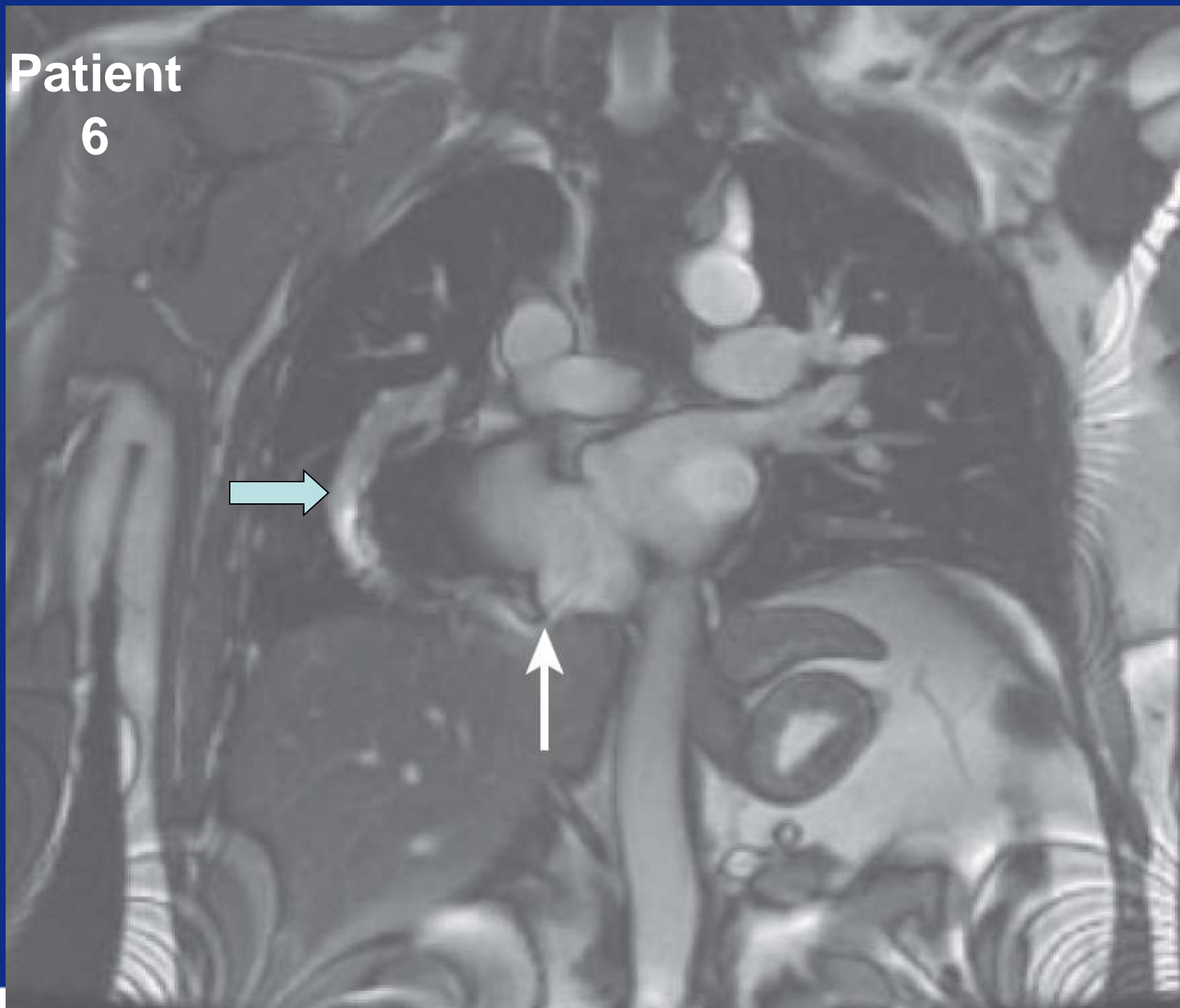


Patient 6 - MRI

Ventricular Volume Quantification

	LV	(Normal range)	RV	(Normal range*)
EDV (mL)	124	(52–141)	245	(58–154)
ESV (mL)	55	(13–51)	121	(12–68)
SV (mL)	69	(33–97)	124	(35–98)
EF (%)	56%	(57–75)	51%	(51–75)
EDVi (mL/m ²)	78	(62–96)	154	(61–98)
ESVi (mL/m ²)	35	(17–36)	76	(17–43)
SVi (mL/m ²)	43	(40–65)	78	(20–40)
Mass index (g/m ²)	59	(47–77)	46	(20–40)

**Patient
6**



Patient 6 - CT





**Patient
6**

10.00 mm/div

Patient 6 – “Risk Factors”

- “Don’t know much about that”.
 - Anomalous pulmonary venous connection.
 - Scimitar syndrome.

Patient 6 - What management would you consider?

Choose as many as are applicable

- Observation.
- Surgical repair of scimitar circulation.
- Embolization of anomalous arterial supply.
- Surgical resection of abnormal lung segment.

Patient 6 – Management and Outcome

- Observation was elected.
- Many patients with scimitar syndrome have a shunt less than 2/1 and do not need redirection of their venous return.
- She did not have lung sequestration on CT scanning. Her lung parenchyma was normal.
- Over an 8-year follow-up, the patient has done well.