Case Report#3

PH and congenital anomaly

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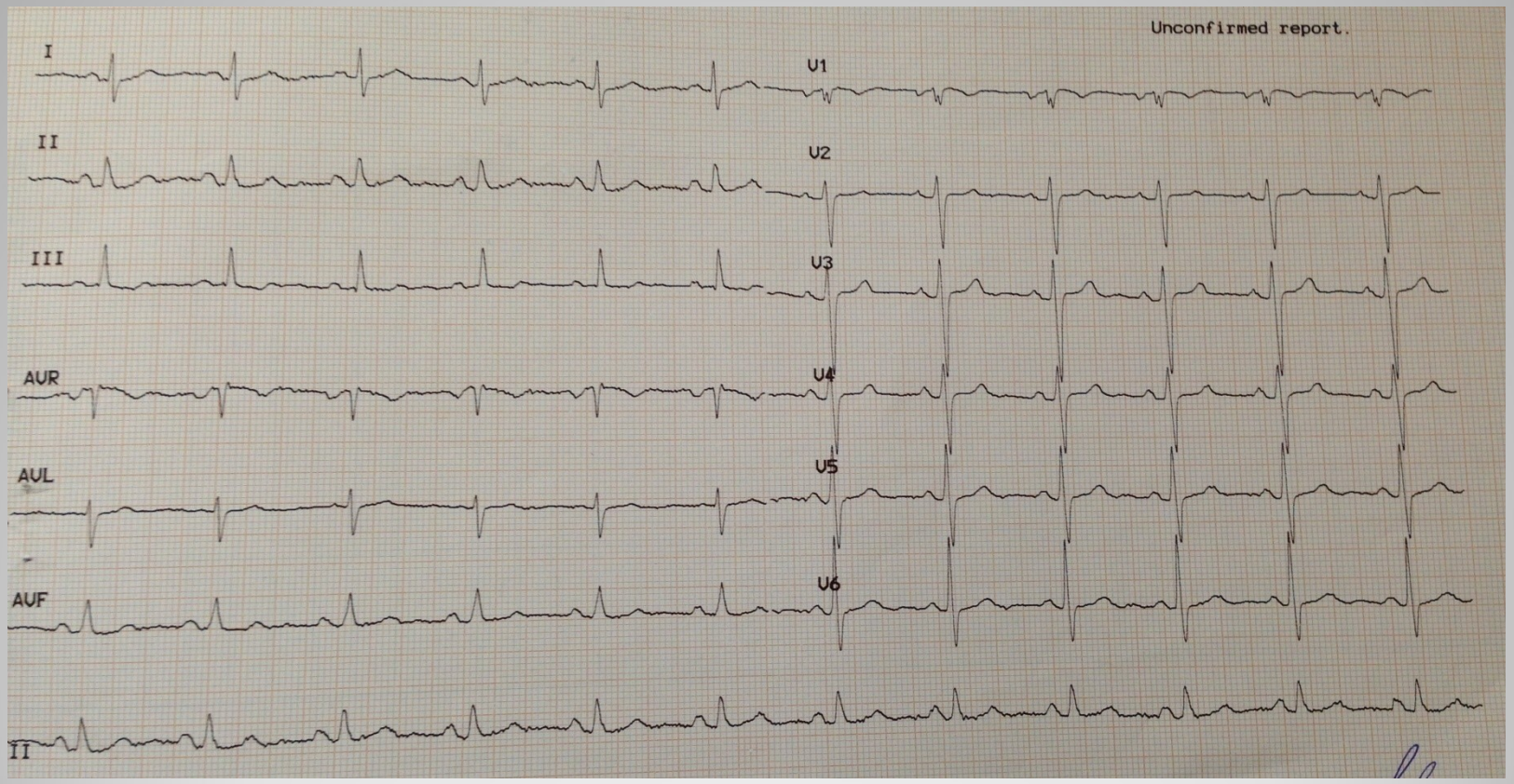
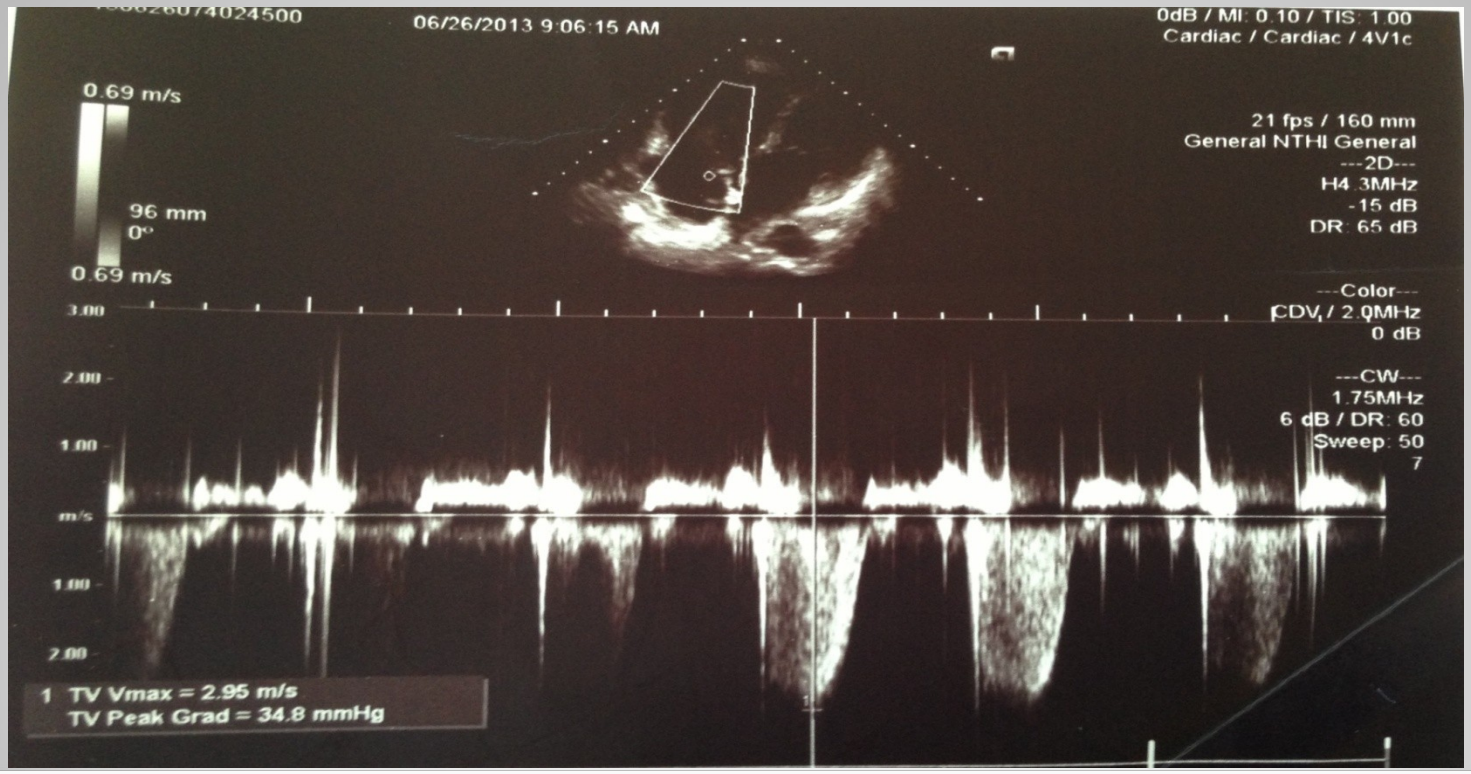
- 59- year – old female;
- Fatigue, dyspnea, chest discomfort on exertion;
- Co-morbidities- Arterial hypertension; Hypothyroidism;
- Referred for PAH work up; referring diagnosos: Idiopatic PAH

- RR- 130/80 mmHg;
- HR- 74/min;
- Split S2, systolic murmur with p. max over the pulmonary valve;

- ECG:

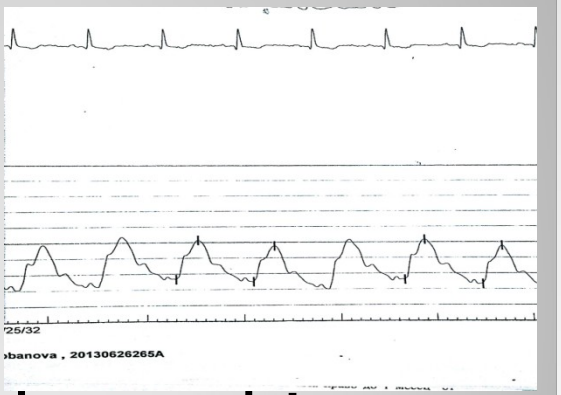


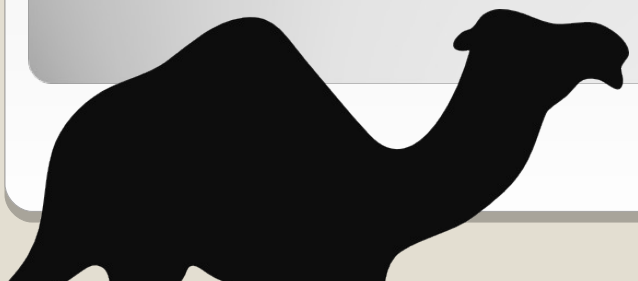
- TTE: Dilated RV, TAPSE- 15mm,
- LV- normal volumes; EF- 56%, LA – “slightly dilated”

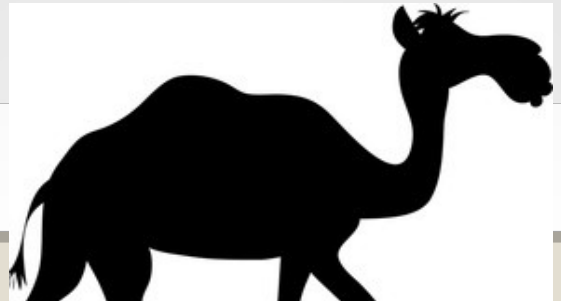


TEE with agitated solution: turbulent flow in RA

- **Heart Catheterization**:
- Left heart catheterization- no stenoses of the coronary arteries;
  LV -134/0/16mmHg; PCWP – not measured
- Right heart catheterization:
  PAP – 48/25/32 mmHg;
  RA – 14/12/12
  Sat O2 PA – 84%
  Thermodulution curve – double peaking





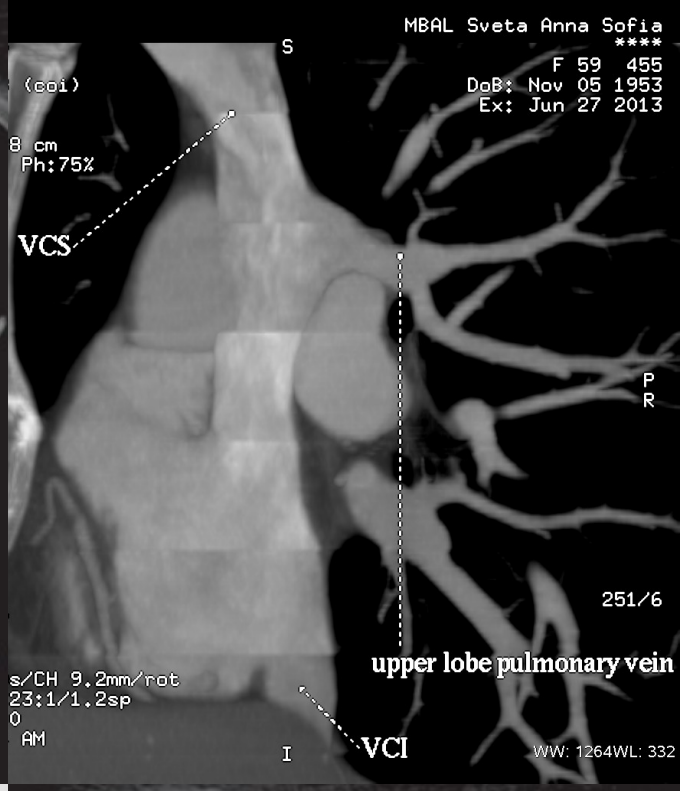


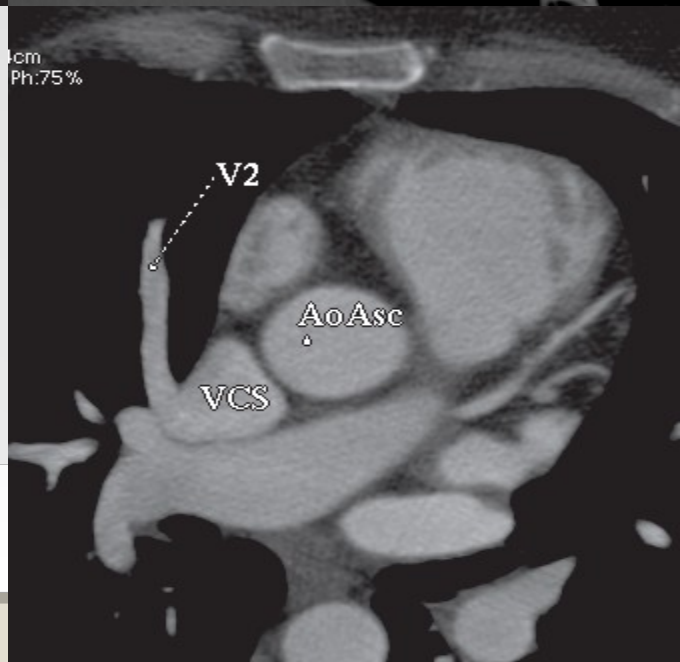
Full run oxymetry

- Sat. O2 VCS- 88 %;
- VCI- 74.7 %;
- RA- 78.6 %;
- PA- 84.2 %;
- Ao- 91.6 %;
- Qp/Qs= 1.5/1

MSCT :

- Intact atrial septum;
- Partial anomalous venous drainage of the right-sided upper and middle lobe pulmonary veins in VCS;
- Dilated RV and truncus pulmonalis;





- **CONTRAST CT SCAN**:
- Intact atrial septum;
- Partial anomalous venous drainage of the right-sided upper and middle lobe pulmonary veins in VCS;
- Dilated RV and truncus pulmonalis

1. Surgery
2. PAH specific therapy
3. Conventional meds
4. Something else

What to do?

PAVD in 59 yo woman

- **Cardiosurgery correction:**

- Redirection the flow from upper and middle lobe pulmonary veins into LA;

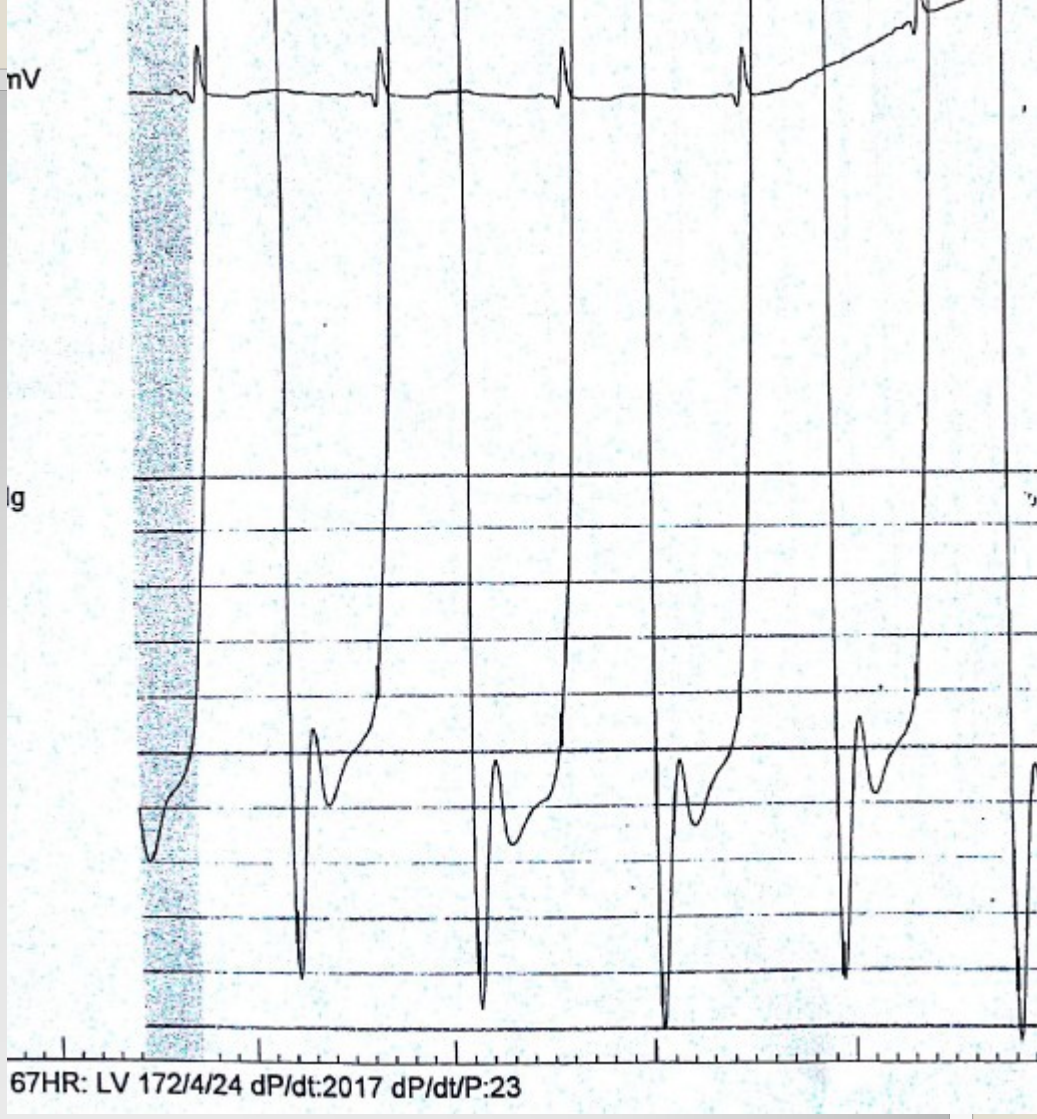
- Plastic operation of the tricuspid valve with ring;

- Uneventful recovery;

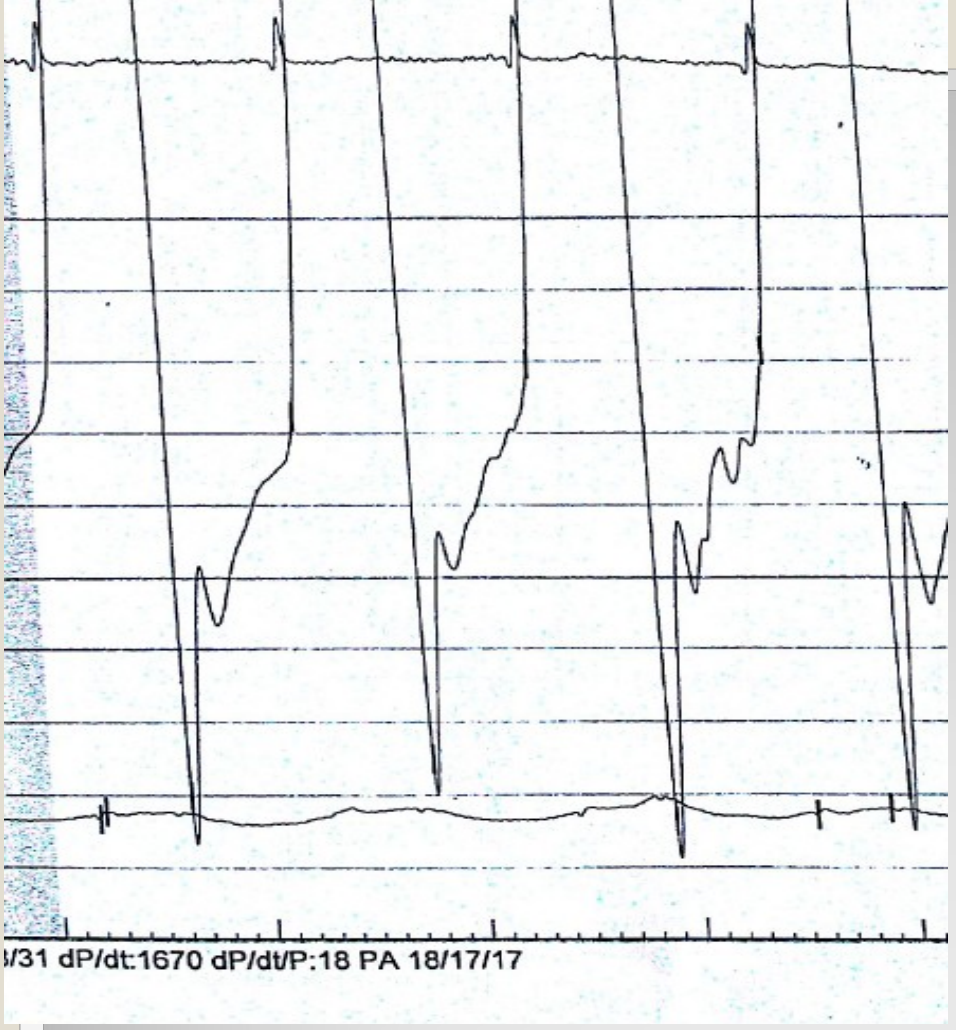
- Patient complains that “nothing has change and even getting worse”
- Unremarkable biochemistry
- Echo estimated SPAP- 60mmHg
- New cath was appointed….

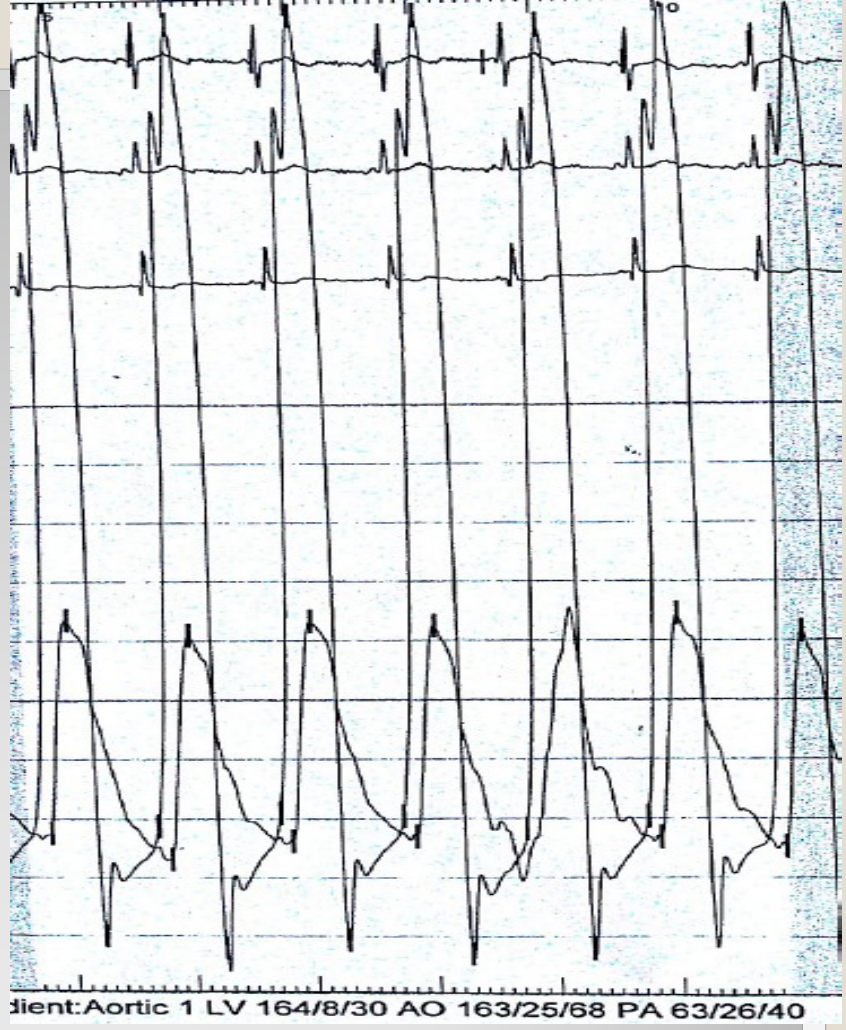
2 mo later….

- PA 60/22/40
- RA 14/11/12
- LV 170/4/23
- Sat PA – 66%
- CI – 2.3 l/min/m2



Presures





300 cc of Nacl 0.9%

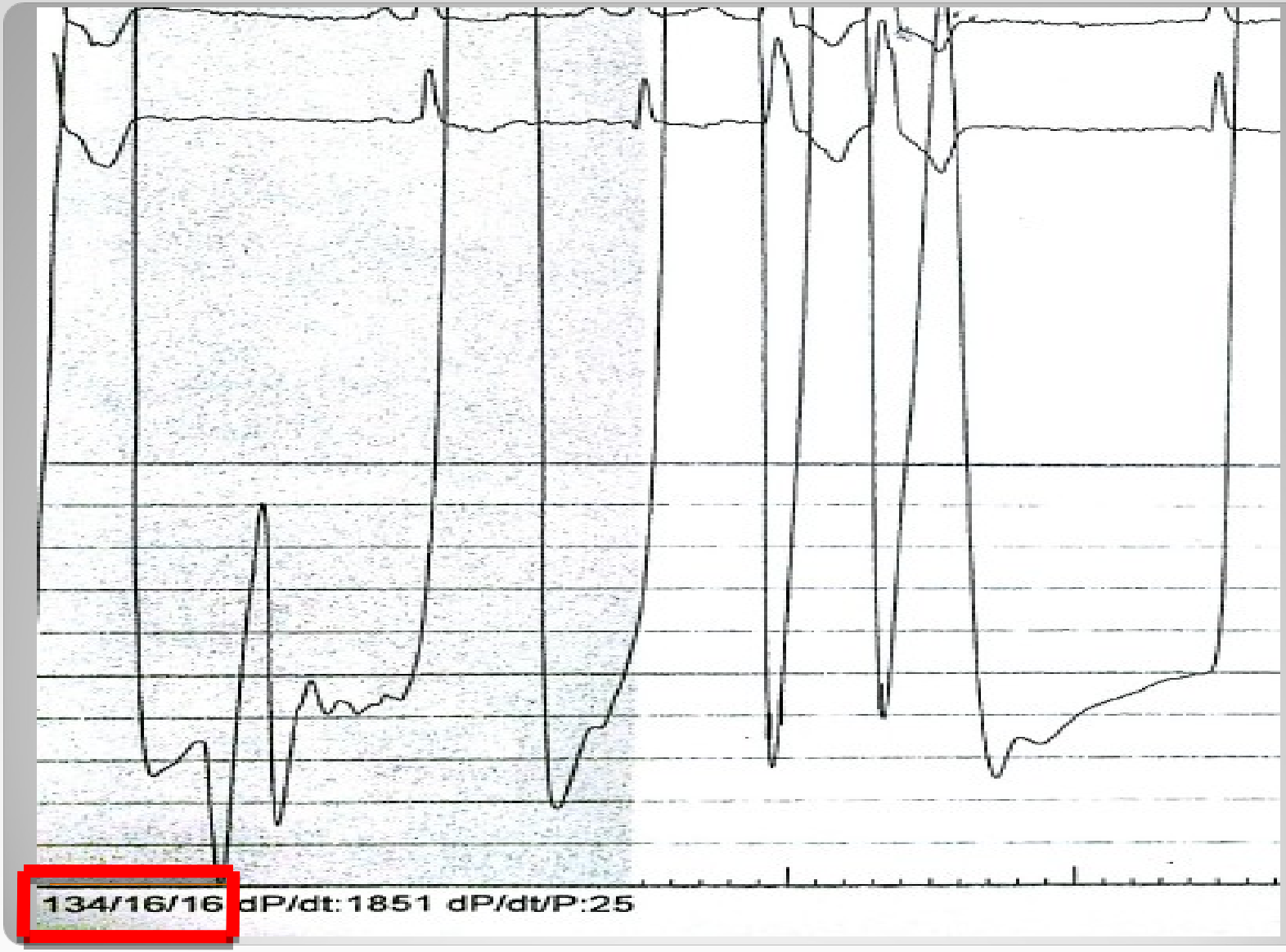
- LV diastolic dysfunction in adult with corrected partial anomalous venous return
- Postcapilary PH with normal TPG

Started on ACEI, BB, Diuretics with no effect on FC

Final diagnosis

Take home messages:

- PH is a syndrome and can be multifactorial
- Not all shunts need to be closed
- Redirecting the flow to the stiff LV can exacerbate diastolic dysfunction
- Always pay meticulous attention to your curves to exclude LV diastolic dysfunction



LV pressures from the 1st cath