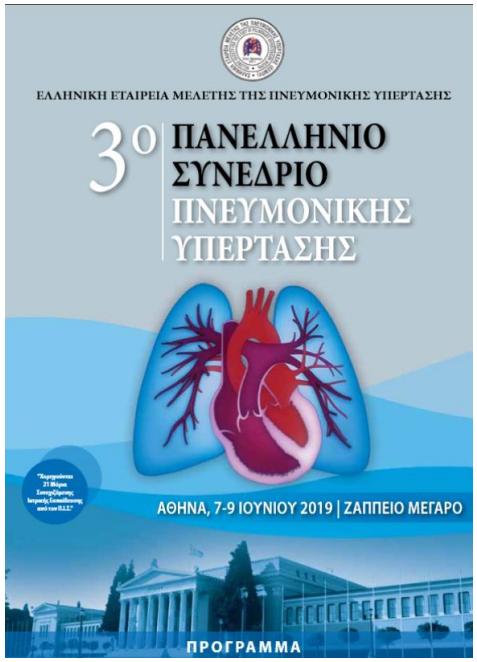
Ενδιαφέροντα περιστατικά με ΠΥ

Μαρία Δρακοπούλου

Α' Πανεπιστημιακή Καρδιολογική Κλινική

Ιπποκράτειο Νοσοκομείο



Background

DoB:

- 20th August 1990 (27y F)

Cardiac Diagnoses:

- Situs solitus, levocardia, AV and VA concordance
- Perimembranous VSD, PFO

Presented at RBH at the age of 12 years old

Diagnostic cardiac catheterization (12/7/2002)

Haemodynamics: In air

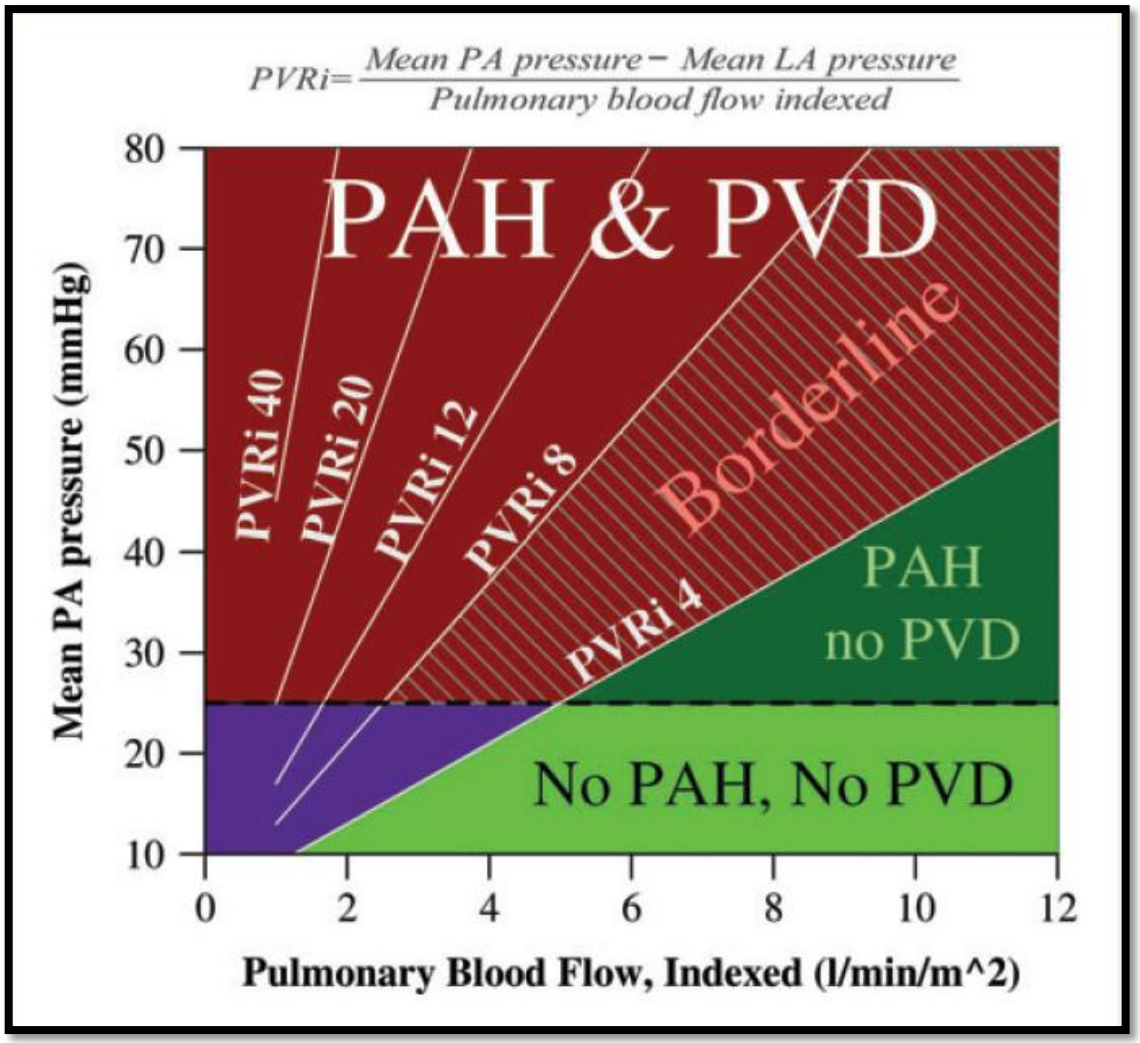
Site	SaO_2 (%)	Pressure (mm Hg)
IVC	85.6	
SVC	74.4	
RA	82	
RV	82.6	66/-4/4
PA	85	60/27/44
Desc Ao	96.5	99/50/69

Findings: 1. Qp:Qs Air 1

2. PVRI

Air: VO2 150ml/min 6.3 units.m2

Calculating PVR in patients with CHD



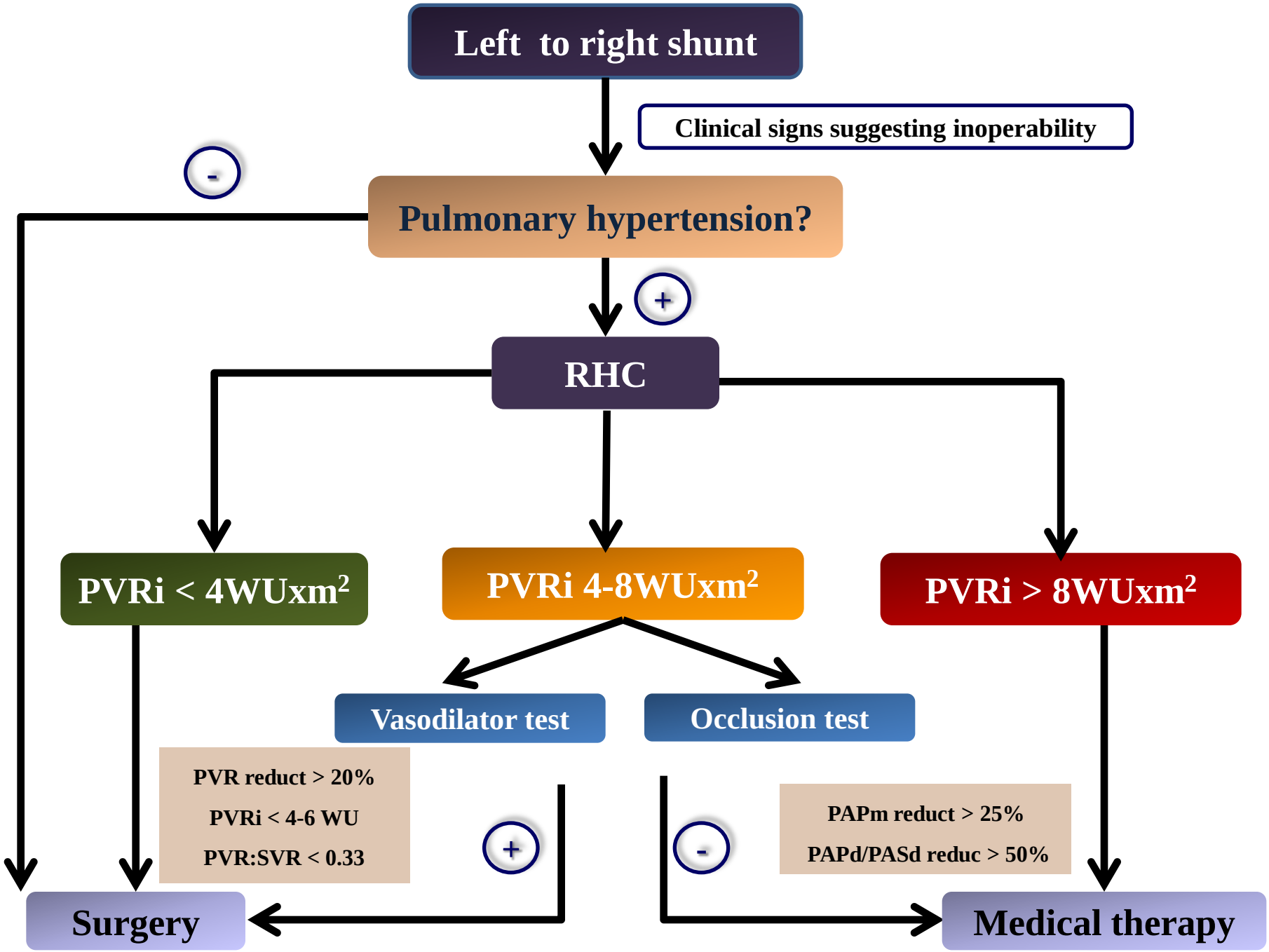
K. Dimopoulos, S.J. Wort, M.A. Gatzoulis, European Heart Journal (2014) 35, 691–700

What is the role of Vasoreactivity Testing in CHD?

AVT in the evaluation of children with PVR

Recommendations	COR	LOE
Cardiac catheterisation is indicated in all paediatric patients with pulmonary hypertension (PH) to confirm diagnosis, to evaluate the severity and when PH-specific drug therapy is considered.	I	C
Initial cardiac catheterisation should include right as well as left heart catheterisation to establish the diagnosis (not only RHC), if there is no contraindication.	I	C
Cardiac catheterisation may be omitted in acutely presenting, critically ill patients requiring immediate initiation of therapy.	I	B
Cardiac catheterisation for the diagnosis of PAH should include acute vasoreactivity testing (AVT; see main text), unless there is a reasonable contraindication, such as PH associated with left heart disease (PH Group II).	I	C
AVT to assess prognosis and indication for specific PH therapy: The haemodynamic change that defines a positive response to AVT in PH without shunt (Qp:Qs=1:1) (see main text) for children should be considered as a >20% fall in mean pulmonary arterial pressure (mPAP) and PVRi/SVRi ratio without a decrease in cardiac output.	IIa	C
Haemodynamic indicators of PH severity are PVRi/SVRi ratio and PVRi, rather than per cent fall in mPAP during AVT. Severe PH with high PVRi/SVRi ratio and high PVRi requires advanced and/or combination therapy.	IIa	C
AVT to assess operability of APAH-CHD: The haemodynamic change that defines a positive response to AVT in PH with shunt (Qp:Qs >1.5:1) (see main text) for children should be considered as a >20% fall in PVRi and PVRi/SVRi with respective final values <6 iWU and <0.3	IIa	C

Apitz C, et al. Heart 2016;102:ii23–ii29


Left to right shunt
Clinical signs suggesting inoperability
-
Pulmonary hypertension?
+
RHC
PVRi < 4WUxm2
PVRi 4-8WUxm2
PVRi > 8WUxm2
Vasodilator test
Occlusion test
PVR reduct > 20%
PVRi < 4-6 WU
PVR:SVR < 0.33
+
-
PAPm reduct > 25%
PAPd/PASd reduc > 50%
Surgery
Medical therapy

Diagnostic cardiac catheterization (12/7/2002)

Haemodynamics: In 100% oxygen and 20ppm NO

Site	SaO_2 (%)	Pressure (mm Hg)
IVC	95	
SVC	71.6	
RA	89.6	2/2/1
LA	98.5	7/10/6
RV		60/-2/6
PA	96	60/26/44
LV		87/-1/7
Desc Ao	98.5	94/60/77

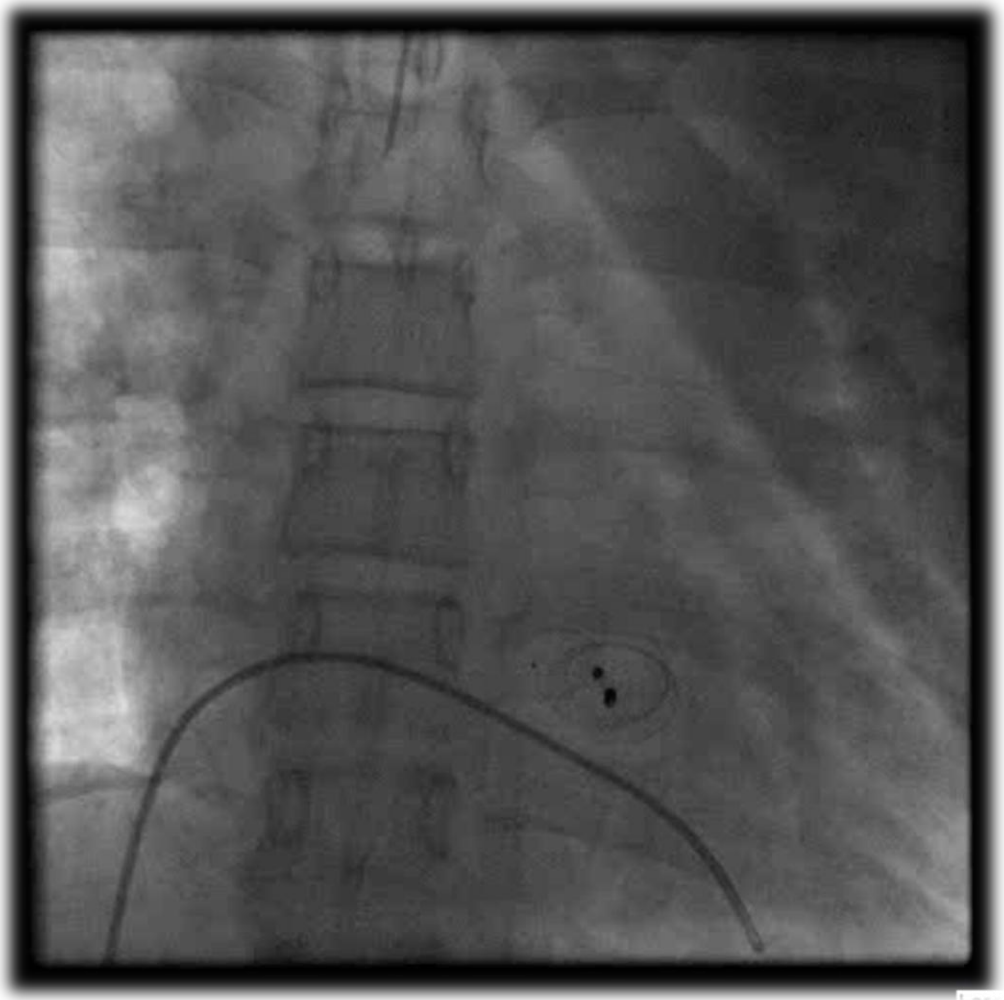
Findings: 1. Qp:Qs 100% oxygen 20ppm NO 4.77

2. PVRI

100% VO2 150ml/min 4.7 units.m2

VSD closure (23/10/2003)

14mm Amplatzer perimembranous VSD occluder



Diagnostic cardiac catheterization (31/5/2005)

Haemodynamics:

Cond 1 (baseline, FiO2 0.21)		
Site	SaO_2 (%)	Pressure (mm Hg)
SVC	77	
RA		6/7/5
RV		68/8
MPA	73	62/34 – 49
LA	92	10/9/6
LV		84/6
LV-Ao pullback		no gradient
Ao	92	78/51 - 65

Cond 2 (FiO2 1.0)		
Site	SaO_2 (%)	Pressure (mm Hg)
SVC	79	
RA		9/7/6
MPA	79	56/11 – 34
LA	99	9/8/6
Ao	98	77/50 – 63

Cond 3 (FiO2 1.0, NO 80 ppm)		
Site	SaO_2 (%)	Pressure (mm Hg)
SVC	81	
RA		9/8/7
MPA	82	57/21 – 38
LA	98	11/10 – 8
Ao	98	75/50 – 63

Cond 4 (washout, FiO2 0.21)		
Site	SaO_2 (%)	Pressure (mm Hg)
SVC	70	
RA		9/9/7
MPA	70	60/27 – 43
LA	90	11/12/9
Ao	90	76/47 – 60

Cond 5 (FiO2 0.21, NO 80 ppm)		
Site	SaO_2 (%)	Pressure (mm Hg)
SVC	69	
RA		8/7/6
MPA	70	59/23 – 40
LA	92	10/9/6
Ao	91	83/48 – 67

Hb 13.3, HR 68-72, VO2 for 14 yrs/HR 70 is 120 ml/min

PVR and shunt calculations:

Qp/Qs in baseline condition: 0.8 – 1

Condition 1(baseline, FiO2 0.21)	*Condition 2 (FiO2 1.0)*	*Condition 3 (FiO2 1.0, NO 80 ppm)*	*Condition 4 (washout, FiO2 0.21)*	*Condition 5 (FiO2 0.21, NO 80 ppm)*
12.5 W.u. m2	**9.4 W.u. m2**	**8.3 W.u. m2**	**10.3 W.u. m2**	**11.3 W.u. m2**

Clinical Follow-up

2005

June 2011

Clinical Status:

Deteriorated in her exercise capacity

Walks for approximately 5 minutes on the flat

WHO class III

Clinical Examination:

Height Weight

BP 110/63, HR 73bpm, SpO2 98% on air

JVP not raised

Heart Sounds:

S1 normal, increased S2

Normal lung sounds

No hepatomegaly, no peripheral oedema

⬇

Started Ambrisentan 5mg

December 2014

Clinical Status:

Improved exercise capacity,

WHO Class II

Manages 10mins on the flat and ~ 2 flights of stairs

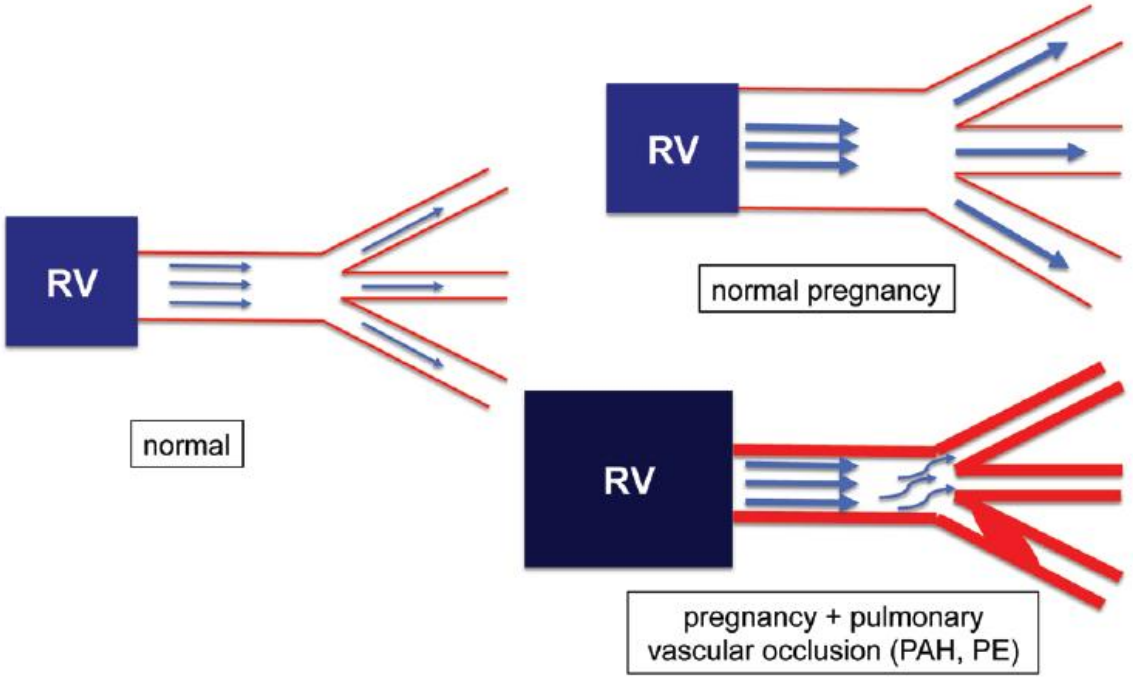
⬇

Wants to attempt pregnancy

Heart rate increases
Circulatory volumes increase
Cardiac output increases
Increased preload
Systemic vasodilatation
Endothelial dysfunction
Reduced colloid osmotic pressure
Coagulopathy
Increased oxygen consumption

Prolonged Valsalva
IVC decompression volume
Bleeding & autotransfusion
Pain & distress
Increased shunting
IATROGENIC !

Why such an issue in PAH?



- Increase in HR & pro-arrhythmic
- Increase in 0_2 consumption (20%)
- Increased tidal volume & minute ventilation
- Elevated diaphragm - 4cm
- Hypercoagulability

Recommendations for pregnancy and PAH

COR	LOE	Recommendations
I	C-LD	1. Women with CHD should receive prepregnancy counseling with input from an ACHD cardiologist to determine maternal cardiac, obstetrical and fetal risks, and potential long-term risks to the mother.[S3.13.1-1–S3.13.1-4]
I	C-LD	2. An individualized plan of care that addresses expectations and contingencies should be developed for and with women with CHD who are pregnant or who may become pregnant and shared with the patient and all caregivers.[S3.13.1-2,S3.13.1-3]
I	C-EO	5. In collaboration with an ACHD cardiologist to ensure accurate assessment of pregnancy risk, patients at high risk of maternal morbidity or mortality, including women with pulmonary arterial hypertension (PAH), Eisenmenger syndrome, severe systemic ventricular dysfunction, severe left-sided obstructive lesions, and/or ACHD AP classification ID, IID, IIID* should be counseled against becoming pregnant or be given the option of terminating pregnancy.
I	B-NR	6. Men and women of childbearing age with CHD should be counseled on the risk of CHD recurrence in offspring.[S3.13.1-9]

2018 AHA/ACC Guidelines. Circulation. 2019;139:e698–e800

Pregnancy Assessment

Pregnant PAH Patient

↓

Counsel on Pregnancy Risks

Discuss Therapeutic Abortion

(Recommended especially in high risk patients)

Indicators of PH patients at high risk of poor outcomes in pregnancy:

1) early clinical deterioration
2) severe RV dysfunction
3) BNP elevation
4) FC III or IV symptoms

1st trimester →

Stop ETRA & warfarin/coumadin
Refer to high risk obstetrician
Monthly clinic visits
Monthly echocardiogram, BNP & 6MWT
Optimize PAH therapy
Start LMWH – if bed rest or inpatient
Recommend therapeutic abortion if right heart failure develops

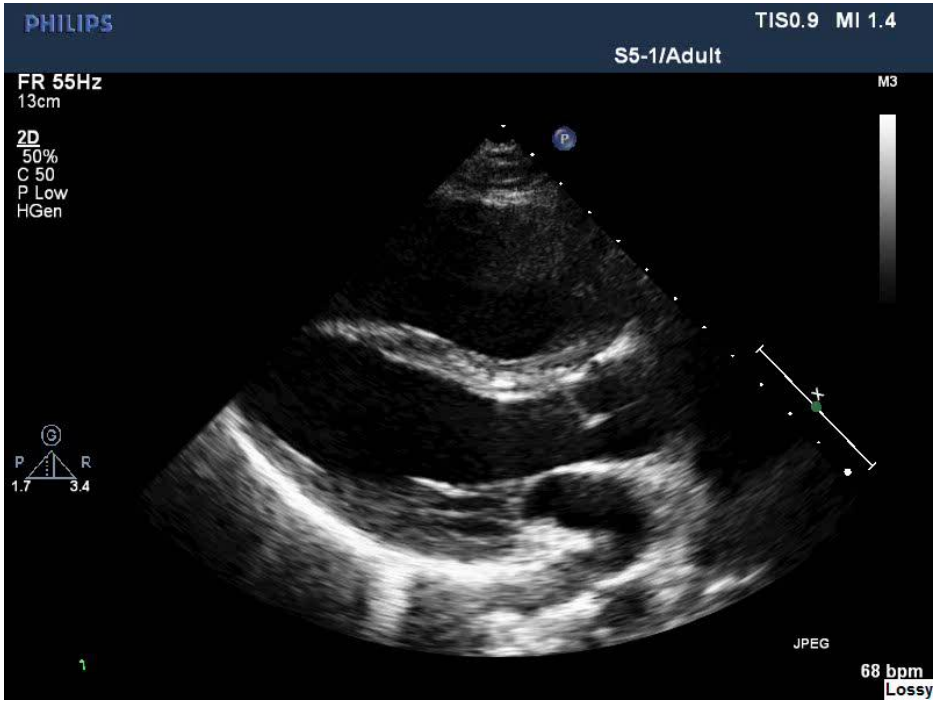
2nd trimester →

Multidisciplinary approach with high risk obstetrician, PAH physician & anesthesiologist
Monthly clinic visits
Monthly echocardiogram, BNP & 6MWT
Start LMWH – if bed rest or inpatients
Optimize PAH therapy

3rd trimester →

Multidisciplinary approach with high risk obstetrician, PAH physician & anesthesiologist
Weekly clinic visits
Weekly echocardiogram, BNP & 6MWT
Optimize PAH therapy
Start LMWH – if bed rest or inpatient
Elective cesarean section at week 34
Close post-operative ICU monitoring

TTE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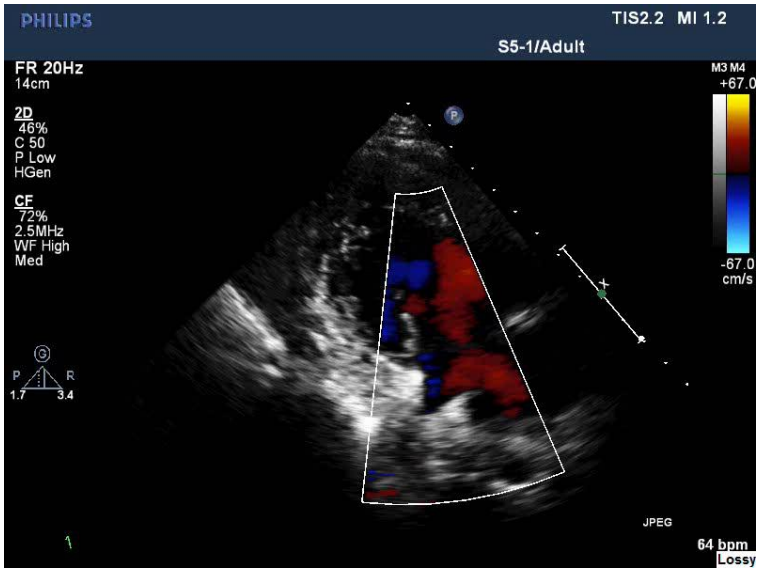
C 50
P Low
HGen
M3
P R
1.7 3.4
JPEG
68 bpm
Lossy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cm/s
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1.7 3.4
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64 bpm
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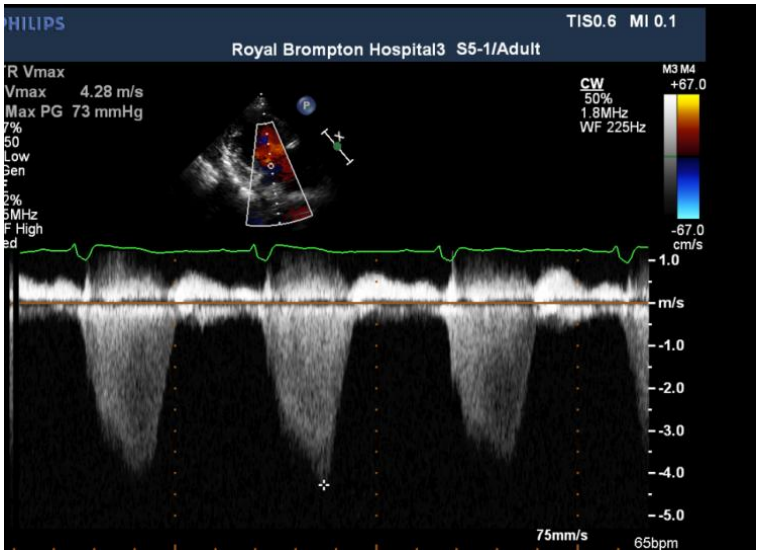
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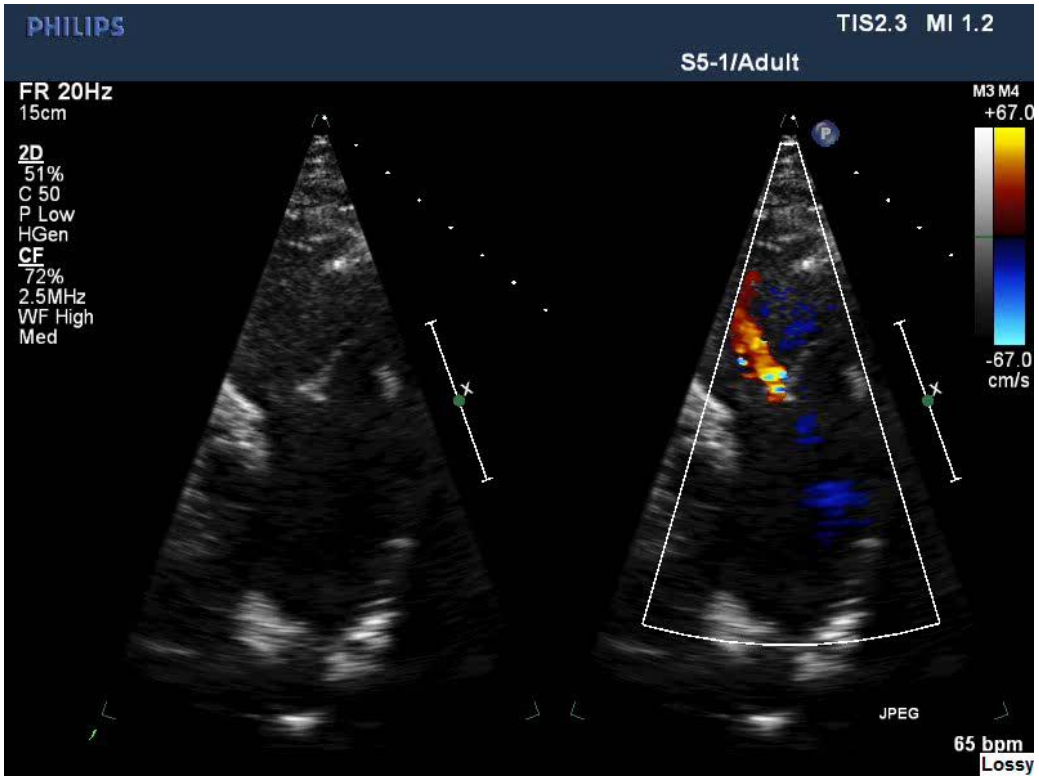
R Vmax
Vmax 4.28 m/s
Max PG 73 mmHg
CW
50%
1.8MHz
WF 225Hz
M3 M4
+67.0
-67.0
cm/s
1.0
m/s
-1.0
-2.0
-3.0
-4.0
-5.0
75mm/s
65bpm

TTE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FR 20Hz
15cm
2D
51%
C 50
P Low
HGen
CF
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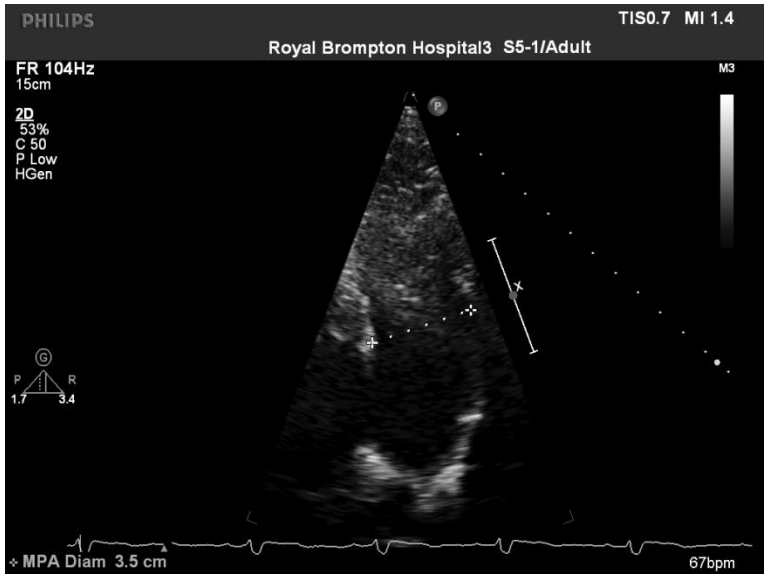
2.5MHz
WF High
Med
M3 M4
+67.0
-67.0
cm/s
JPEG
65 bpm
Lossy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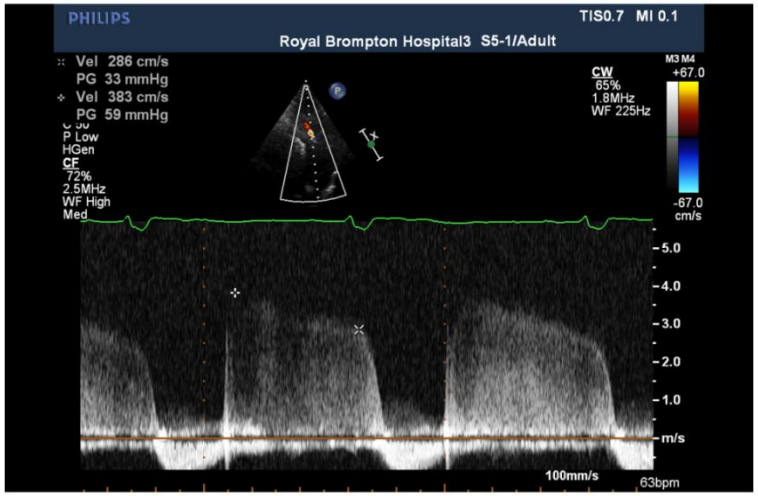
15cm
2D
53%
C 50
P Low
HGen
M3
MPA Diam 3.5 cm
67bpm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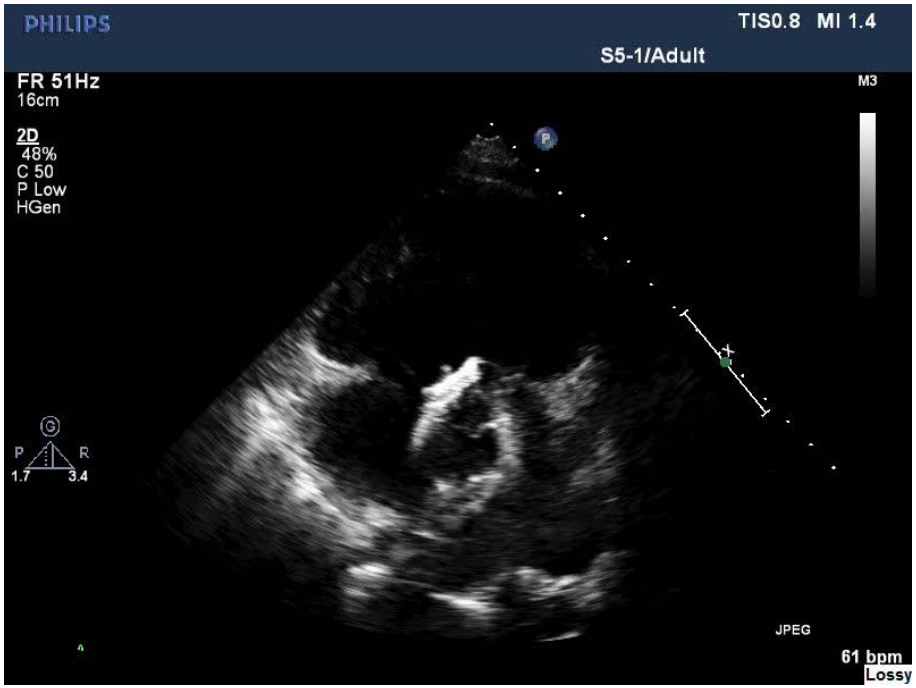
Royal Brompton Hospital3 S5-1/Adult
Vel 286 cm/s
PG 33 mmHg
Vel 383 cm/s
PG 59 mmHg
CW
65%
1.8MHz
WF 225Hz
M3 M4
+67.0
-67.0
cm/s
P Low
HGen
CF
72%
2.5MHz
WF High
Med
5.0
4.0
3.0
2.0
1.0
m/s
100mm/s
63bpm

TTE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R
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JPEG
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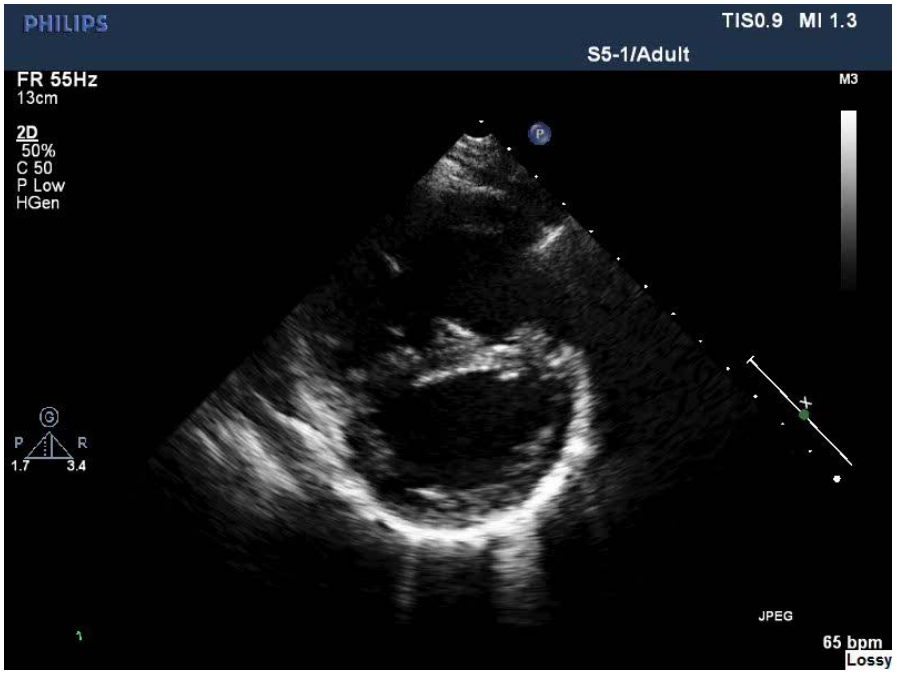
Lossy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C 50
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HGen
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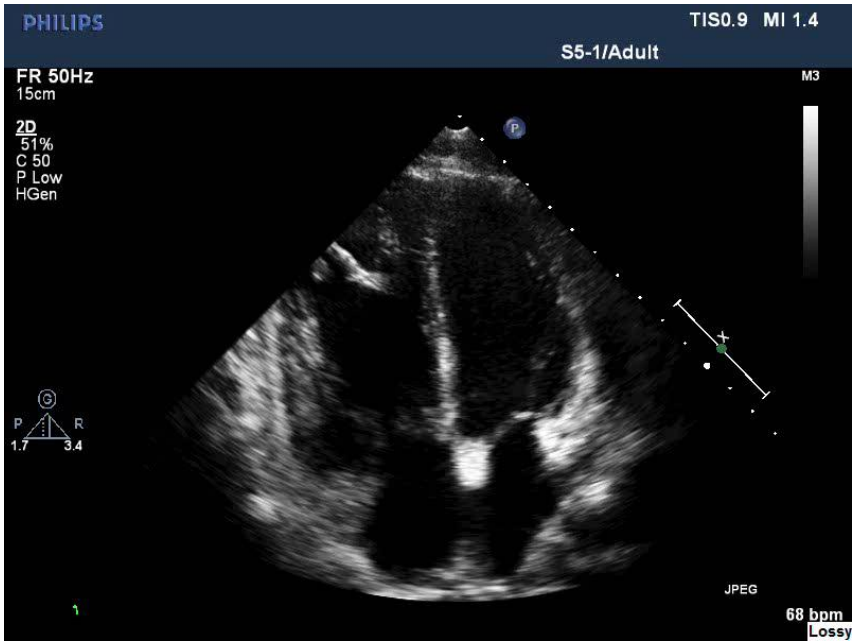
3.4
JPEG
65 bpm
Lossy

TTE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S5-1/Adult
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C 50
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HGen
M3
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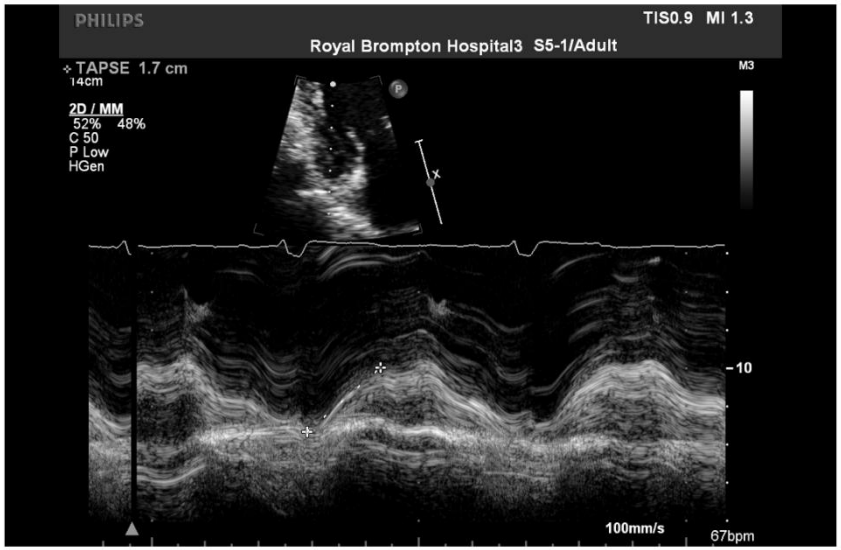
R
1.7
3.4
JPEG
68 bpm
Lossy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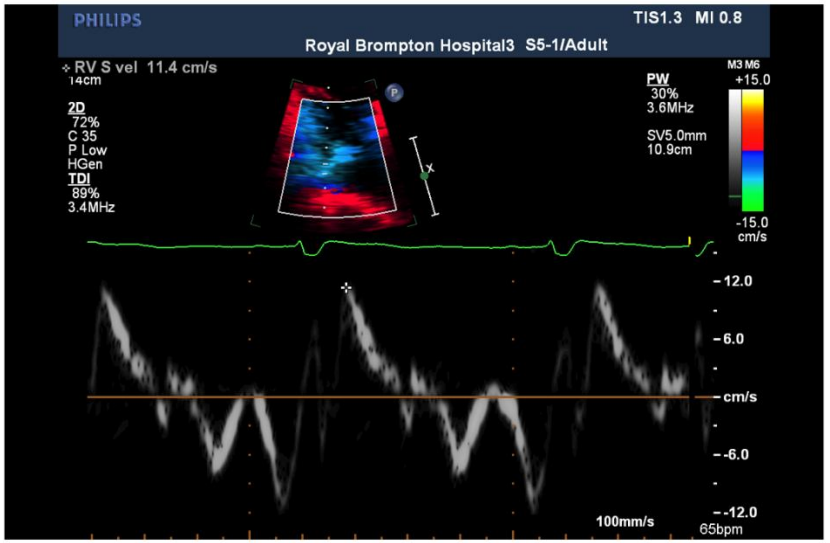
TIS0.9 MI 1.3
Royal Brompton Hospital3 S5-1/Adult
TAPSE 1.7 cm
14cm
2D / MM
52% 48%
C 50
P Low
HGen
M3
10
100mm/s
67bpm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Royal Brompton Hospital3 S5-1/Adult
RV S vel 11.4 cm/s
14cm
2D
72%
C 35
P Low
HGen
TDI
89%
3.4MHz
PW
30%
3.6MHz
SV5.0mm
10.9cm
M3 M6
+15.0
-15.0
cm/s
12.0
6.0
cm/s
-6.0
-12.0
100mm/s
65bpm

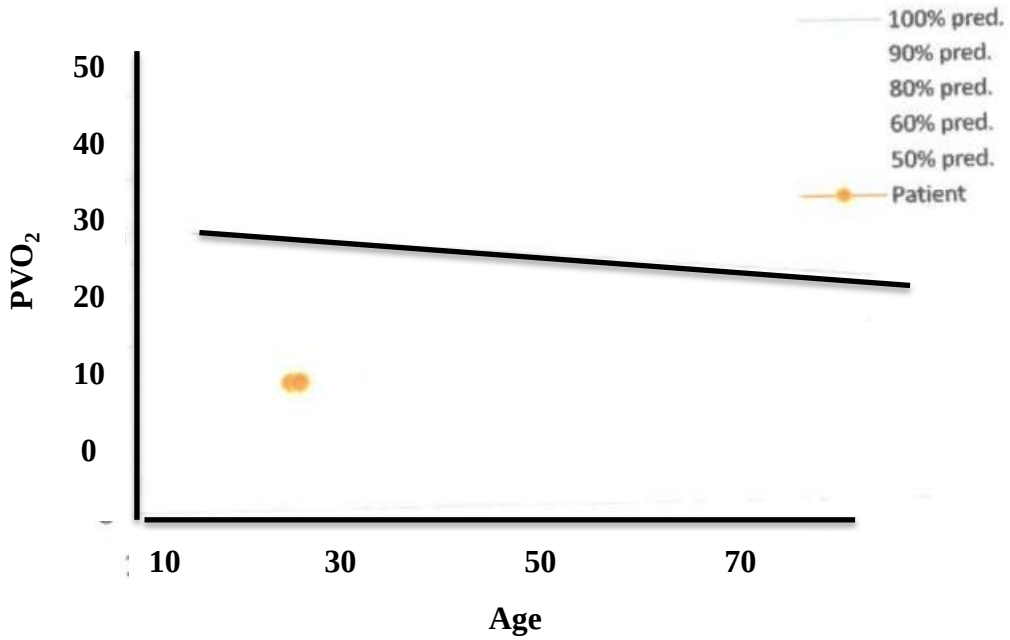
CMR

Normal female ranges (see graphs for BSA indexed LV and RV values)

	EDV (mL)	ESV (mL)	SV (mL)	EF (%)	Mass index (g/m^2)
LV	116	42	74	64	39
RV	174	76	98	56	

Cardiopulmonary Exercise Test with MVO2(June 2014)

Date	01/04/2014	3/12/2014
Age	23.6	24.3
RER	1.15	1.02
Max HR	179	160
PVO_2	15.2	15.1
PVO_2 % predicted	42%	42%
VE/VCO_2	63.0	57.8
Time	6m, 42s	9m, 25s



Rhythm	SR+RBBB			
Resting Pulse	64	Resting BP	90	62
Peak Exercise Pulse	160	Peak Exercise BP	98	62

Right Heart Catherization

	Pressures		Oxygen saturation (%)		
	Baseline	Condition 1	Baseline	Condition 1	Condition 2
Breathing gas	air	NO 40ppm/air	air	NO 40ppm/air	-
High superior caval vein	-	-	70.7	68.1	-
Low superior caval vein	-	-	66.9	66.3	-
Inferior caval vein	-	-	70.3	73.3	-
Right atrium (A/V/mean)	8/6/4	-	70.6	71.7	-
Right ventricle (Sys/EDP)	82/6	79/7	70.6	-	-
Pulmonary artery (Sys/dia/mean)	82/36/56	74/38/54	69.2	71.8	-
PA wedge pressure (A/V/mean)	13/13/8	8/9/6	-	-	-
Left ventricle (Sys/EDP)	-	-	-	-	-
Arterial parameters (non-invasive)	90/50/59	91/51/61	97	97	-
Arterial parameters (invasive)	-	-	-	-	-
Arterial P02	-	-	-	-	-
Left upper pulmonary vein	-	-	-	-	-
Left atrium	-	-	-	-	-
Free text	-	-	-	-	-

Haemoglobin (g/dL)	14
Heart rate	65
Heart rhythm	Sinus rhythm

	Baseline	Condition 1
Pulmonary gradient (mm Hg)	48	48
Pulmonary resistance (WU)	14.6	13.2
Systemic gradient (mm Hg)	55	57
Systemic resistance (WU)	16.7	15.7
Qp/Qs	1.00	1.00

Recommendations for pregnancy and PAH

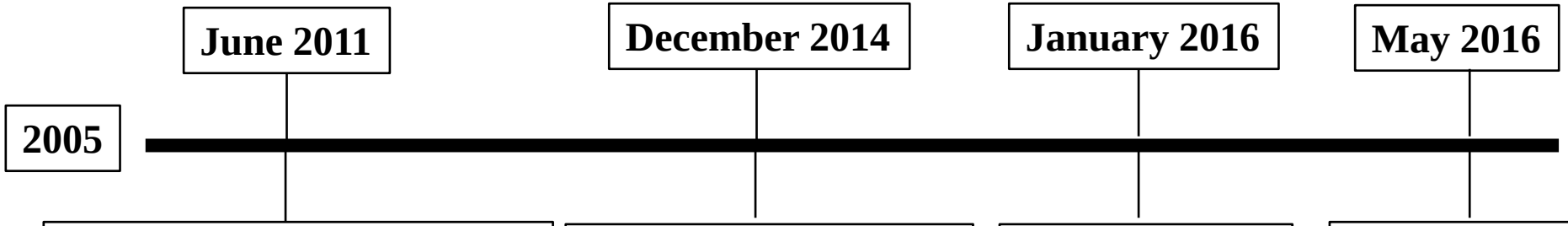
Recommendations	Class[a]	Level[b]
Right heart catheterization is recommended to confirm the diagnosis of PAH (group 1). This can be performed during pregnancy but with very strict indications.[10]	I	C
Treatment dose LMWH is recommended in pregnant patients with chronic thrombo-embolic pulmonary hypertension.	I	C
If a PAH patient conceives on targeted PH therapies, consideration should be given to withdrawing embryotoxic drugs, taking into account the risks of withdrawal.	IIa	C
In treatment-naive pregnant PAH patients, initiating treatment should be considered.[23]	IIa	C
Pregnancy is not recommended in patients with PAH.[119]	III	B

ESC Guidelines, European Heart Journal (2018) 39, 3165–3241

MDT Meeting (December 2014)

- **Preconception counseling:**
  - Patient advised that there is a **high mortality risk** associated with pregnancy
- **Plan:**
  - To be followed at the **antenatal clinic**
  - **Ambrisentan was stopped and Sildenafil 20mg td** was commenced

Clinical Follow-up



Clinical Status:

Deteriorated in her exercise capacity

Walks for approximately 5 minutes on the flat

WHO class III

Clinical Examination:

Height Weight

BP 110/63, HR 73bpm, SpO2 98% on air

JVP not raised

Heart Sounds:

S1 normal, increased S2

Normal lung sounds

No hepatomegaly, no peripheral oedema

Started Ambrisentan 5mg

Clinical Status:

Improved exercise capacity,

WHO Class II

Manages 5-10mins on the flat and ~ 2 flights of stairs

Wants to attempt Pregnancy

Miscarriage

Pregnancy

Optimization PAH therapy

Sildenafil 50mg td

Start anticoagulation

Clexane 40mg od

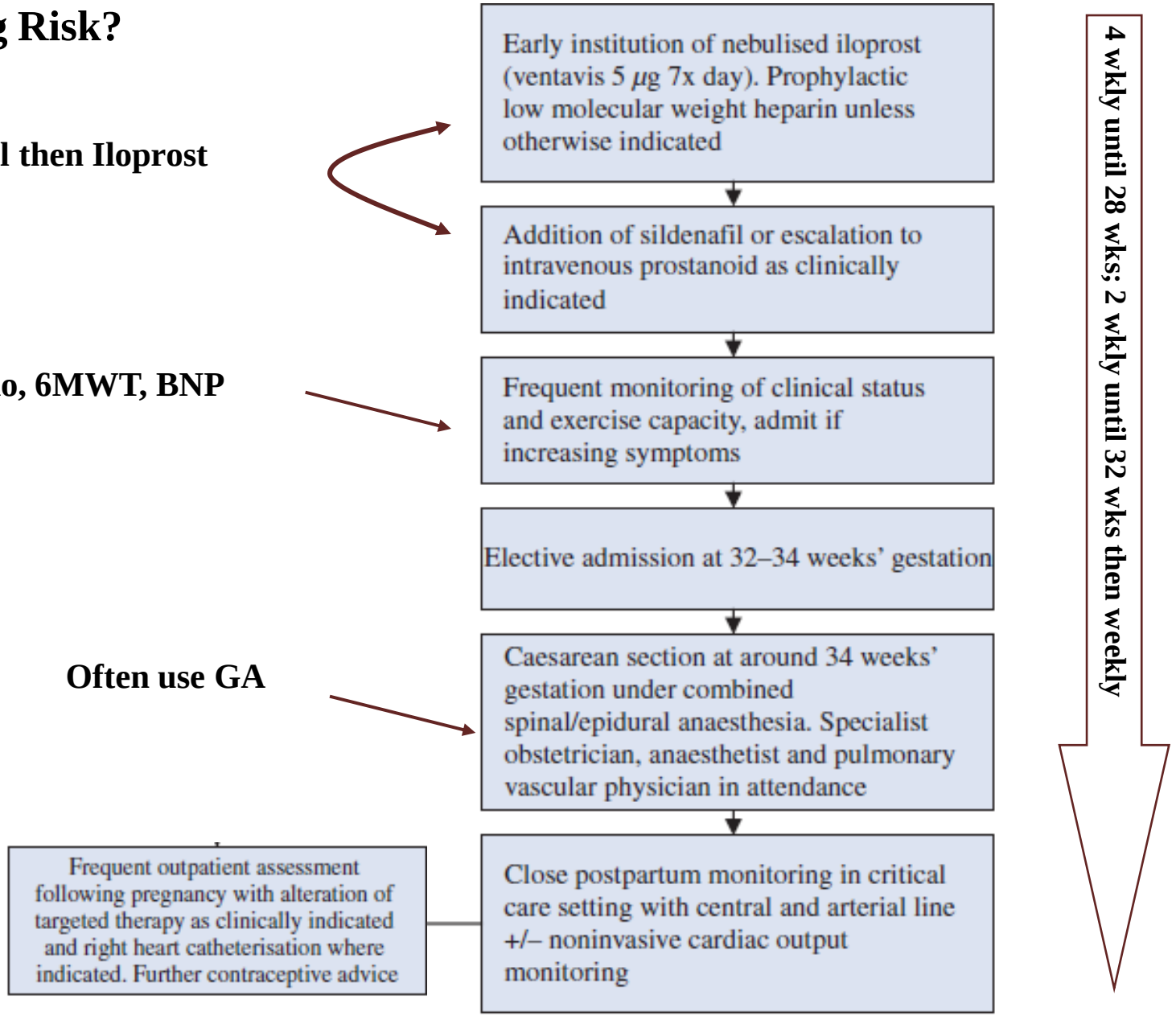
Multidisciplinary approach with monthly visits

Minimising Risk?

Sildenafil then Iloprost

Echo, 6MWT, BNP

Often use GA



Kiely DG et al, BJOG 2009

Admission

23+6 weeks

20.11.2016

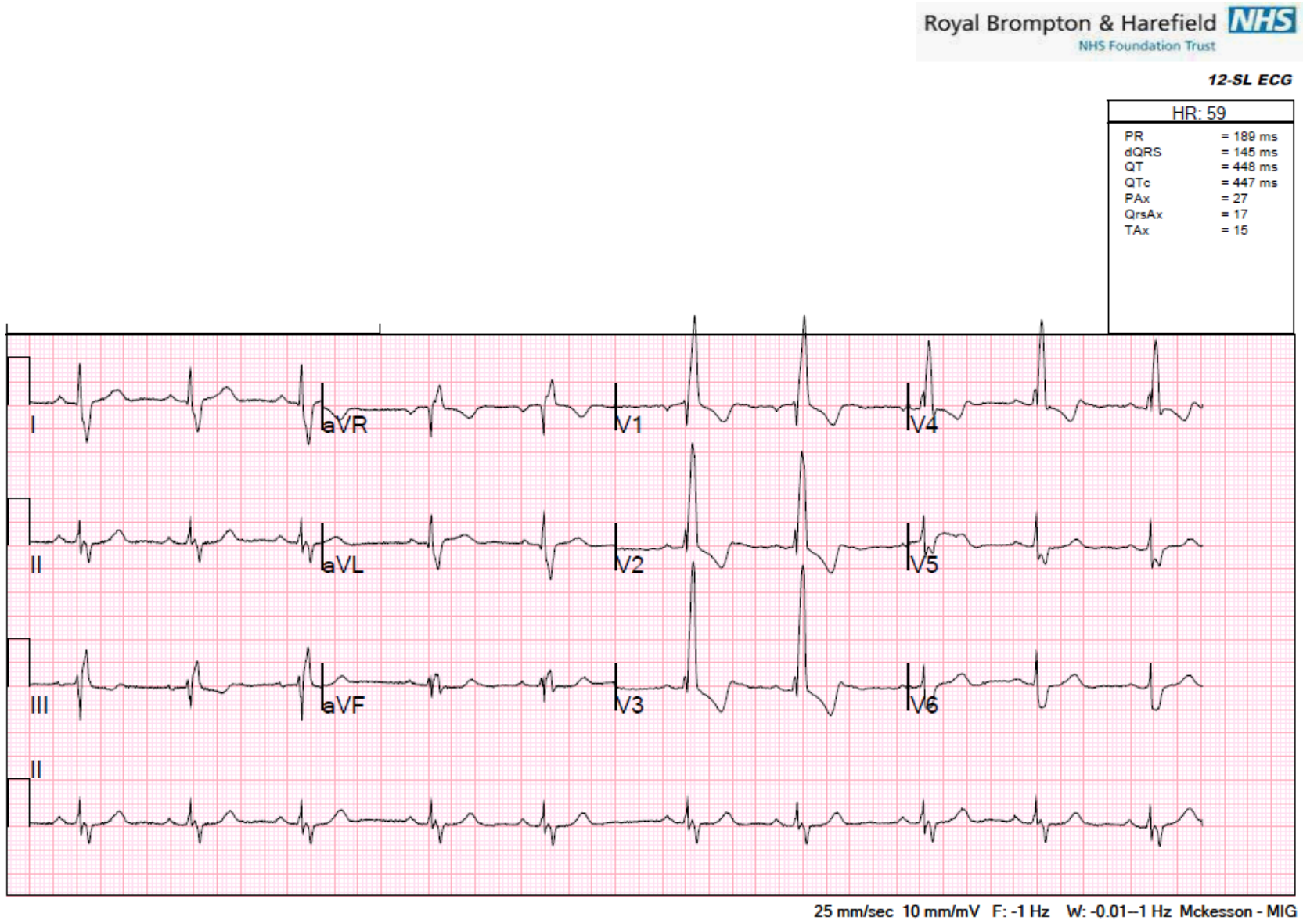
November 2016

-Small volume hemoptysis potentially related to an **upper respiratory infection**

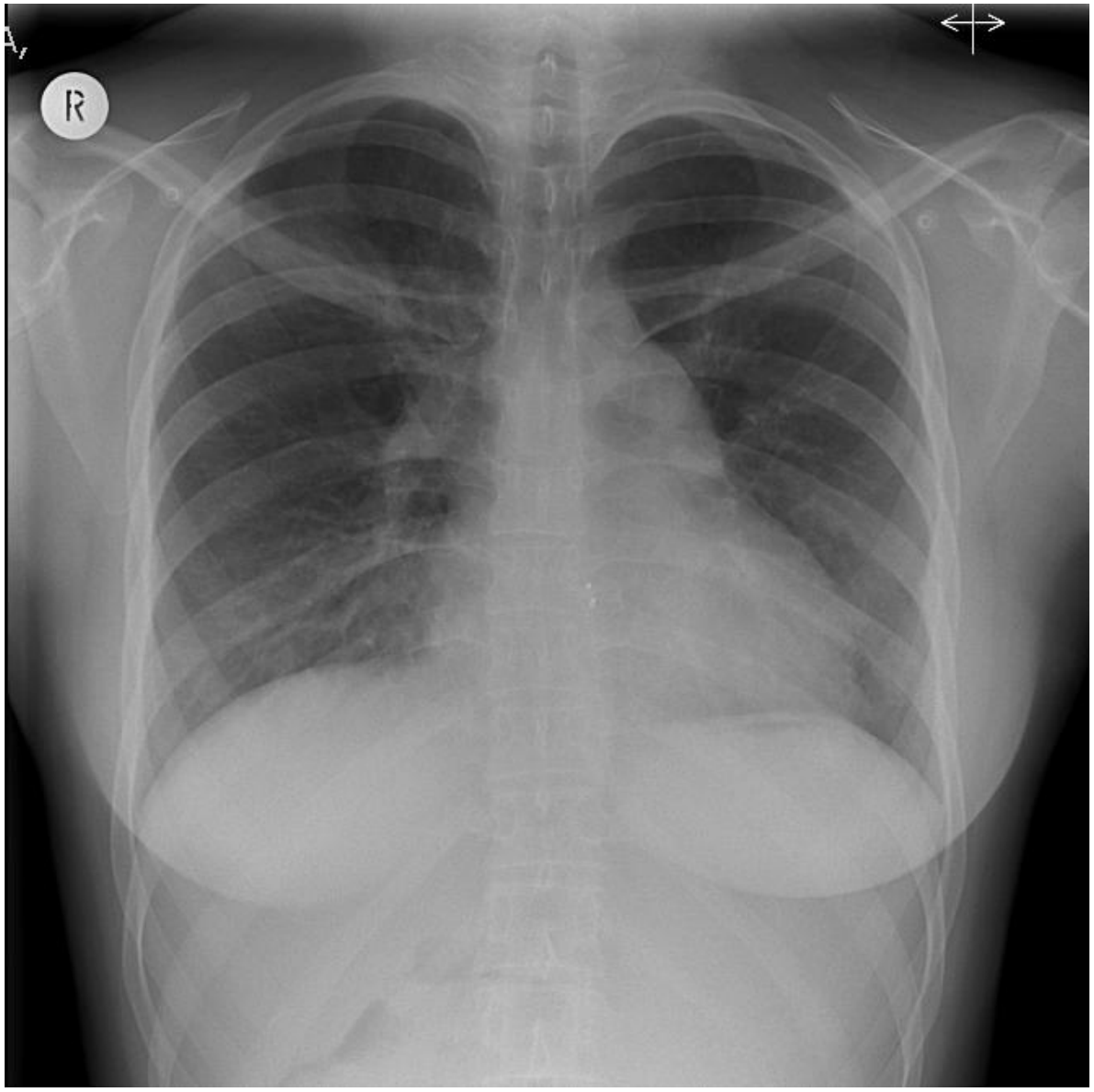
-Increasing SOB and presyncope on effort which represented a change in symptoms from her last review in clinic 3 weeks earlier.

Medication	**19/11/2016**
Sildenafil 50mg td	√
Clexane 40mg od	√

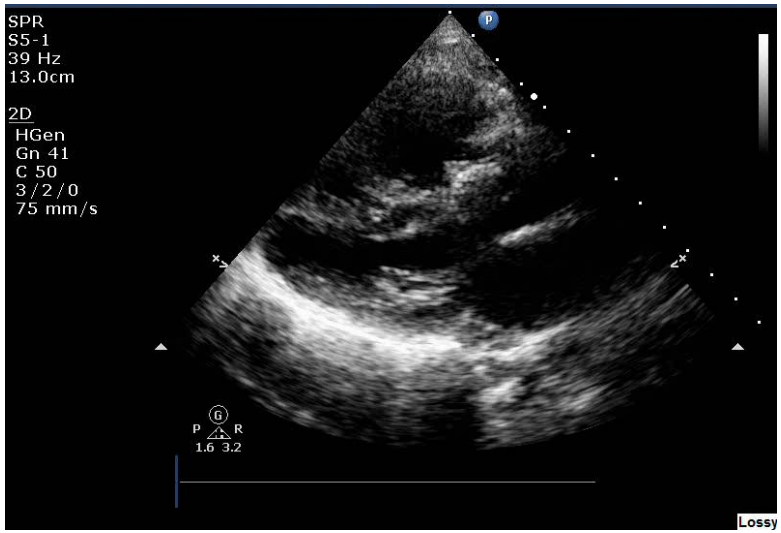
ECG (November 2016)

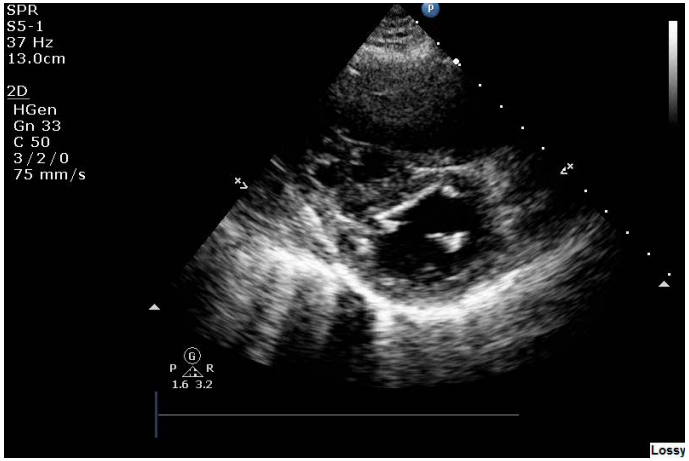


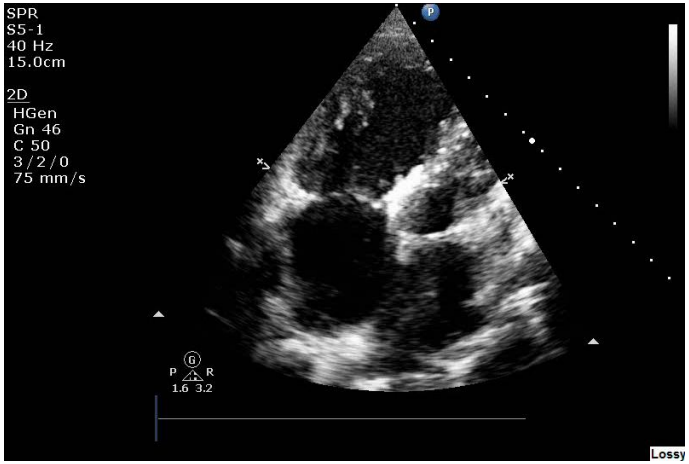
Chest X-Ray (20/11/2016)



TTE (21/11/2016)







During Admission

23+6 weeks

20.11.2016

November 2016

ECG

Chest X-Ray

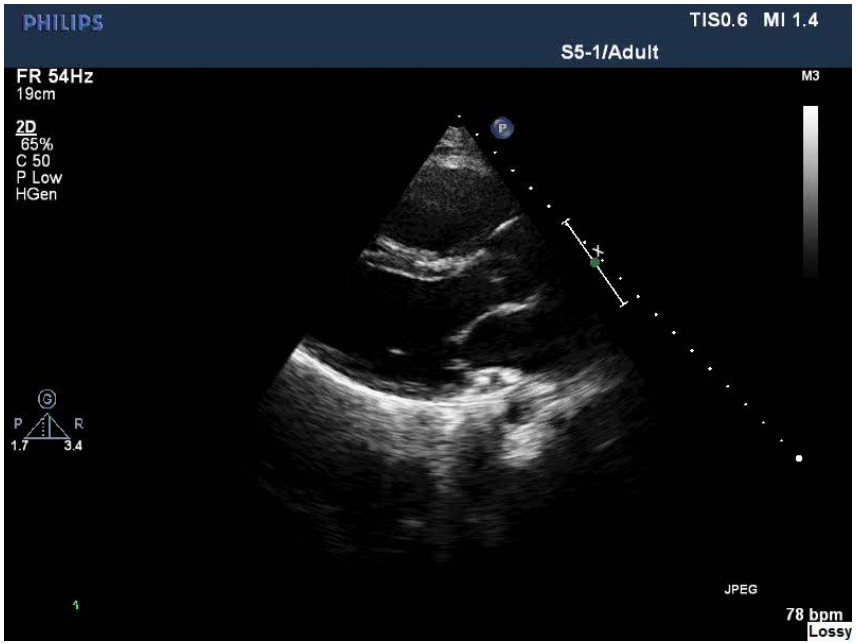
Echocardiogram

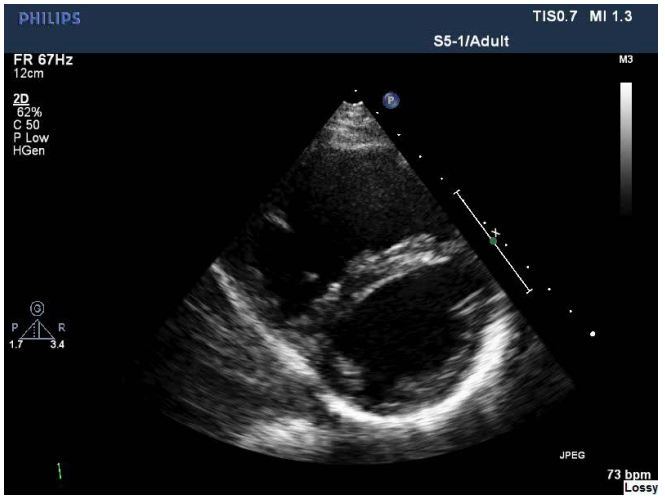
MDT (20/11/2016)

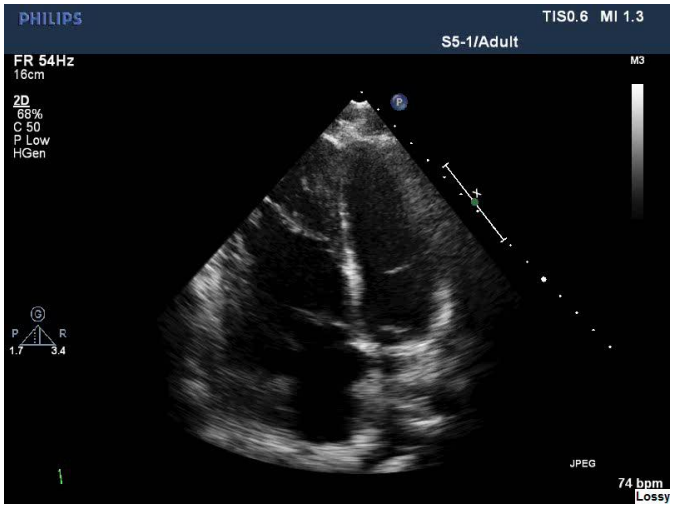
-Start inhaled iloprost therapy (5mcg 3 hourly)

Medication	19/11/2016	20/11/2016
Sildenafil 50mg td	√	√
Clexane 40mg od	√	√
Inhaled Iloprost 5mcg 3 hourly		√

TTE (25/11/2016)







Admission

24+4 weeks

November 2016

21.11.2016

ECG

Chest X-Ray

Echocardiogram

MDT (20/11/2016)

-Start inhaled iloprost therapy (5mcg 3 hourly)

25.11.2016

MDT (25/11/2016)

-Insertion of a PICC line

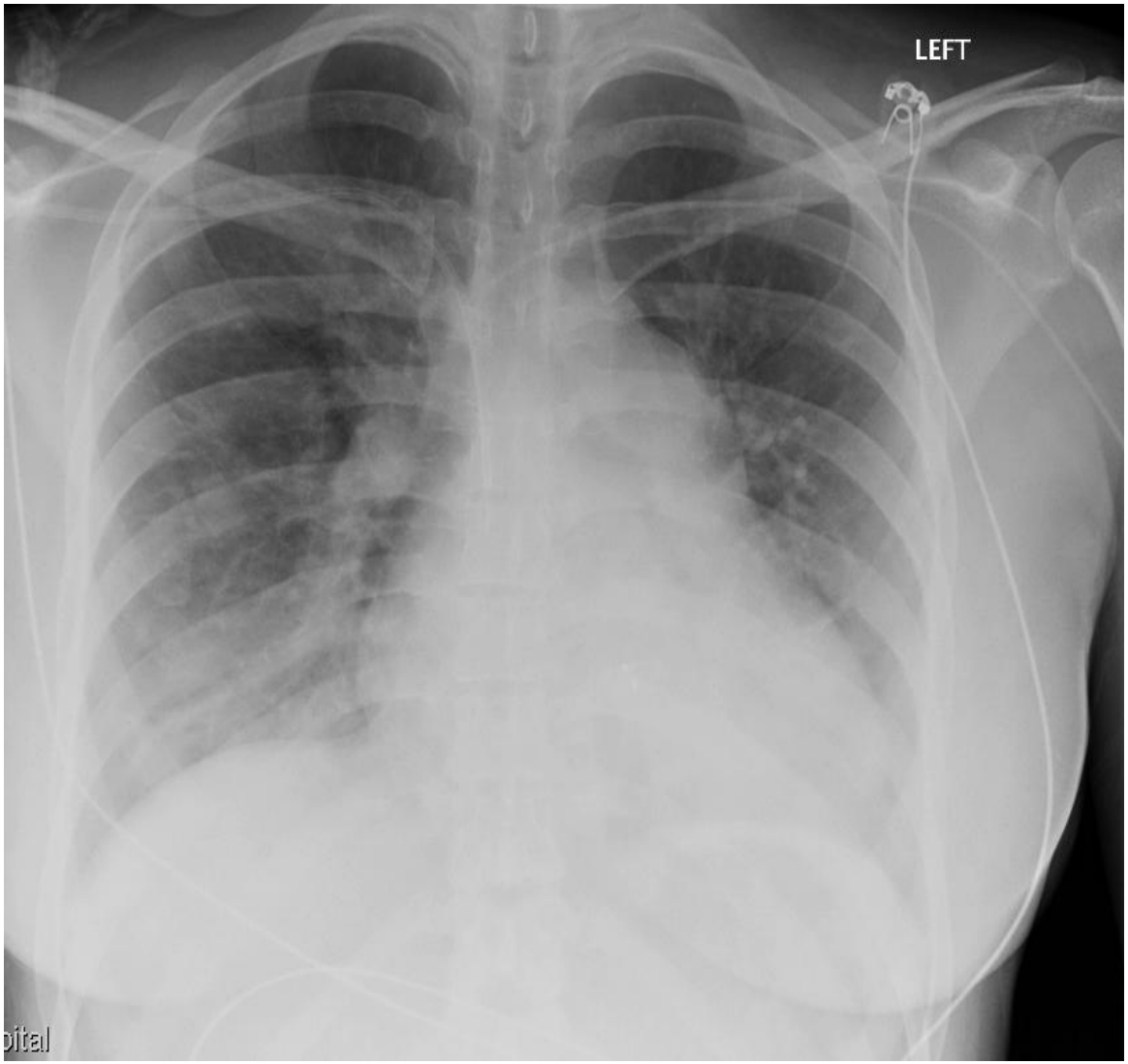
-IV Epoprostenol and escalation of therapy by 1-2ng/day according to tolerance

-Transfer to HDU

26 weeks

Medication	30/11/2016	2/12/2016	5/12/2016
Sildenafil	√ (50mg td)	√ (50mg td)	√ (25mg bd, 40od)
Clexane 40mg od	√	√	√
Inhaled Iloprost 5mcg 3 hourly	√		
IV Epoprostenol	√ (5ng.kg/min)	√ (6ng.kg/min)	√ (6.5ng.kg/min)
Dexamethasone			√ (12mg bd)

HDU Admission (06/12/2016)



Admission

26+1 weeks

November 2016

25th November 2016

MDT (25/11/2016)

-Insertion of a PICC line

-IV Epoprostenol and escalation of therapy by 1-2ng/day according to tolerance

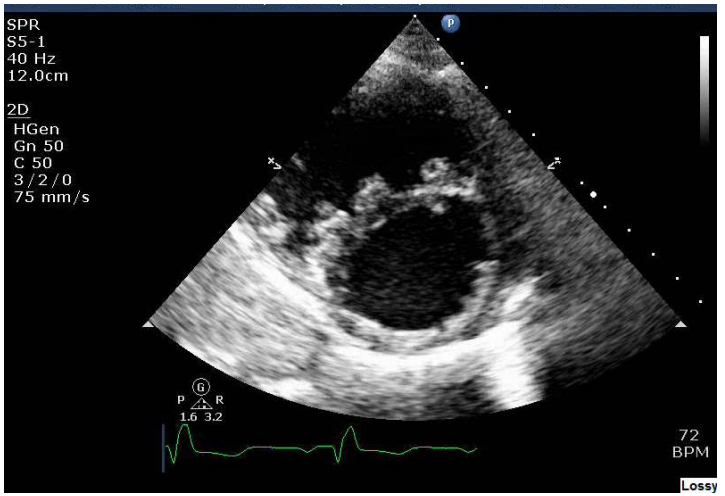
-Transfer to HDU

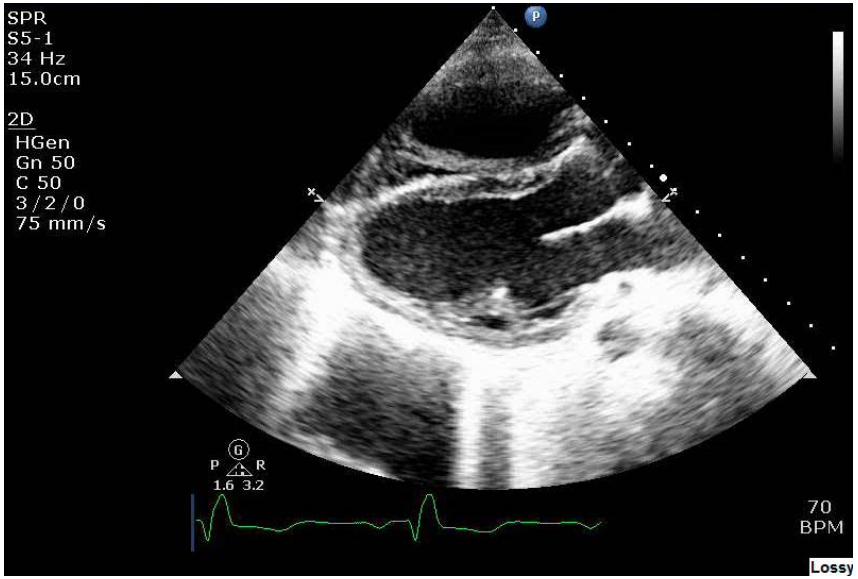
6th December 2016

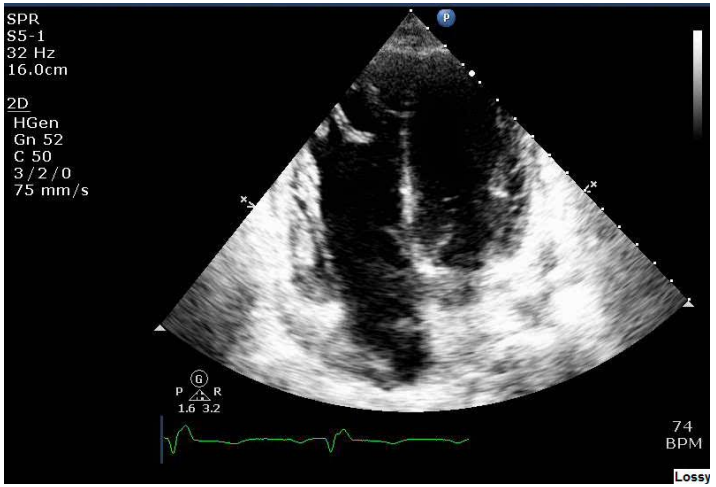
Chest X-Ray: Right lung lower lobe infection-
Respiratory support with CPAP and highflow oxygen
Reviewed on a regular basis by the maternal obstetrics team
Aim: Escalation of Epoprostenol by 0.5ng/kg/min 12 hourly up to 15ng/kg/min or maximum tolerated dose (not less than 10ng/kg/min)

Medication	6/12/2016	10/12/2016	12/12/2016
Sildenafil	√ (25mg td)	√ (25mg td)	√ (25mg td)
Clexane 40mg od	√	√	√
IV Epoprostenol	√ (7ng.kg/min)	√ (10ng.kg/min)	√ (12ng.kg/min)
Piptazobactam	√	√	
Salbutamol inhalers	√	√	
Intermittent CPAP	√	√	

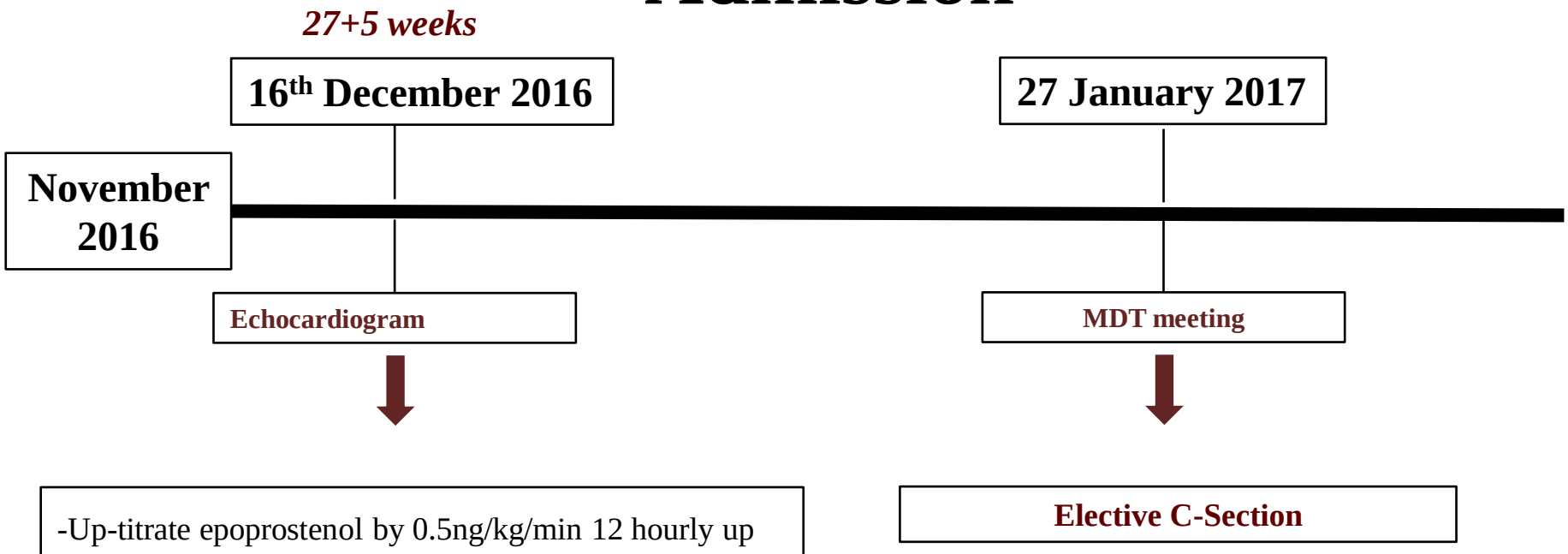
TTE (16.12.2016)





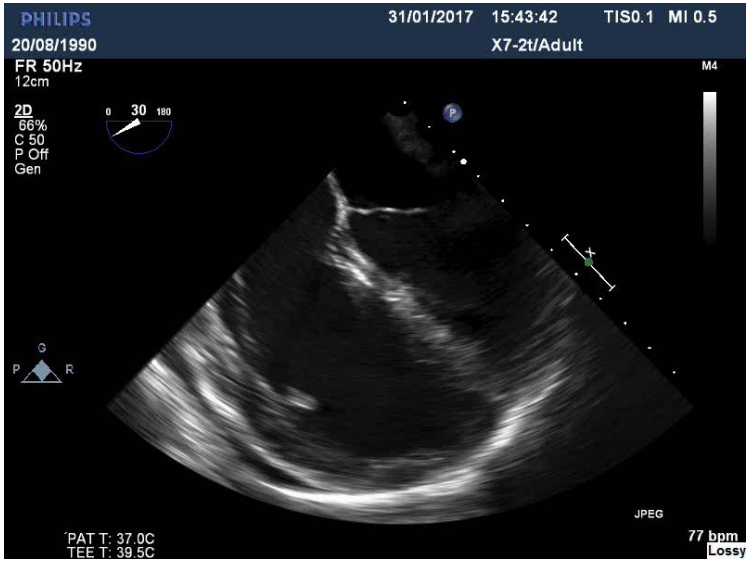


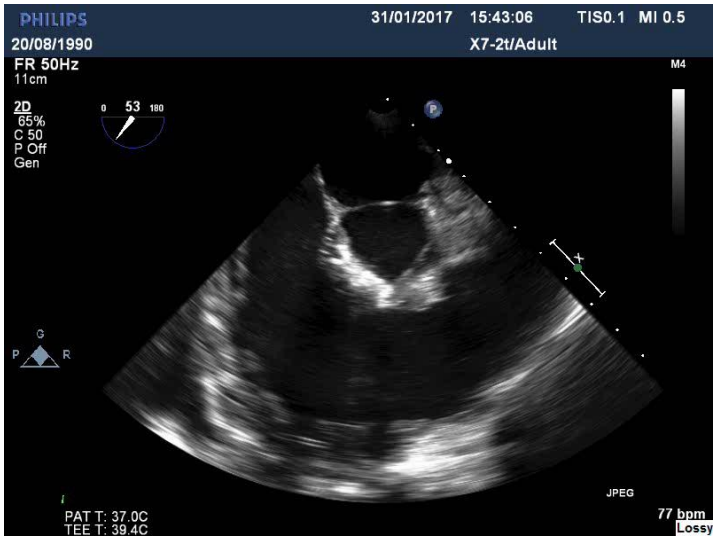
Admission


27+5 weeks
16th December 2016
27 January 2017
November 2016
Echocardiogram
MDT meeting
-Up-titrate epoprostenol by 0.5ng/kg/min 12 hourly up to 15ng/kg/min or maximum tolerated dose (not less than 10ng/kg/min)
-CPAP is required intermittently
Elective C-Section

Elective C-Section at 32+4 weeks (31/1/2016)

- **Under TOE investigation**





Admission

32+2 weeks

31 January 2017

Post-Op C-Section

May 2016

Elective C-Section

-Estimated 700mls blood loss intra-operatively

-Managed on continued epoprostenol infusion and continued sildenafil 25mg TDS

-Baby transferred to Chelsea and Westminster neonatal unit for monitoring

-Continued epoprostenol infusion and continued sildenafil 25mg TDS

-On **oxytocin infusion** due to PV bleeding

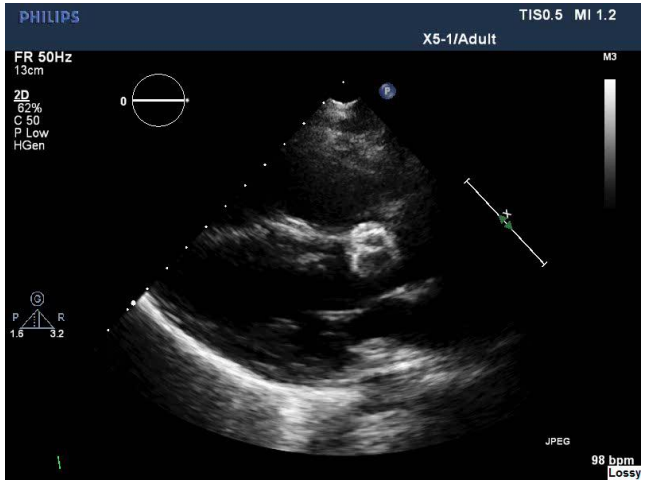
-On **epoprostenol infusion**

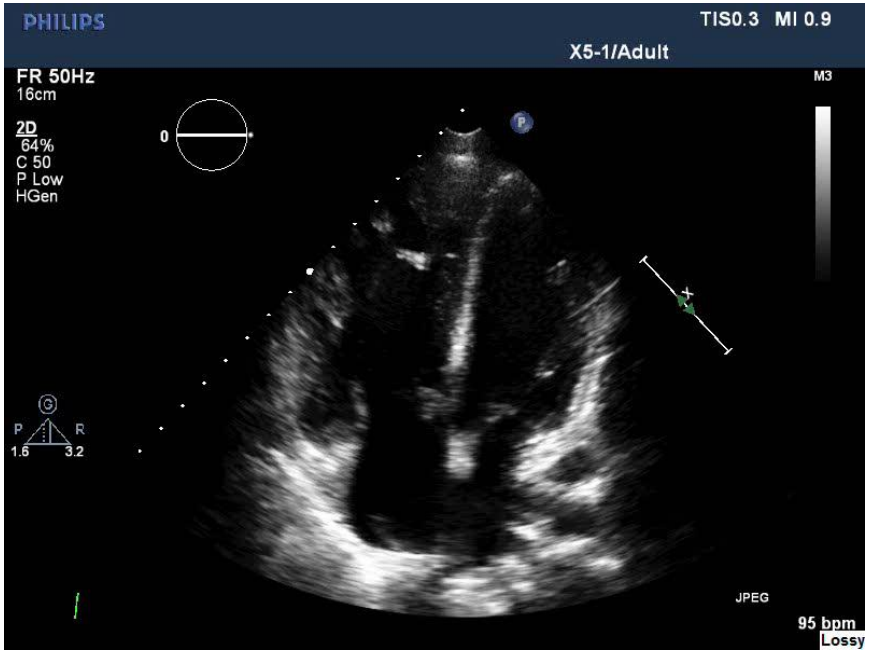
Plan:

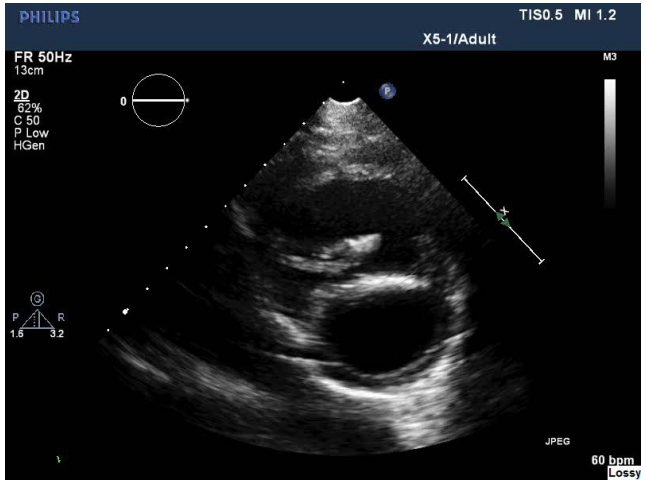
Physiotherapy

Negative fluid balance

TTE Pre-Discharge







Follow-up Plan

- Repeat echocardiogram in 4 weeks and revision of epoprostenol infusion
- Continue epoprostenol for 6 months and arrange a right heart catheterization
- Follow Hb level (given Ferinject 500mg prior to discharge)
- Long-term contraception
  - Started a progesterone only mini-pill, until she can be reviewed for Mirena Coil insertion in 3 months time.

Peripartum outcomes in PAH-CHD

Maternal mortality and morbidity remain high in PAH-CHD patients, who should be counseled on the risks of pregnancy and managed in a tertiary multidisciplinary environment to improve prognosis

	PAH-CHD ($n=94$)	Eisenmenger Syndrome ($n=30$)	PAH with systemic to pulmonary shunt ($n=51$)	PAH after defect correction ($n=13$)	p-Value of difference between all groups
Outcomes					
Maternal death	6 (6.4%)	5 (16.7%)	0 (0%)	1 (7.7%)	0.012
Heart failure	33 (35.1%)	20 (66.7%)	7 (13.7%)	6 (46.2%)	<0.001
PHC	10 (10.6%)	6 (20.0%)	1 (2.0%)	3 (23.1%)	0.012
Other complications					
SBE	2 (2.1%)	0 (0%)	2 (3.9%)	0 (0%)	0.423
Thromboembolic event	2 (2.1%)	1 (3.3%)	1 (2.0%)	0 (0%)	0.779
Arrhythmia	4 (4.3%)	1 (3.3%)	2 (3.9%)	1 (7.7%)	0.529

Multivariate predictor			
SaO_2	0.869	0.786–0.962	0.007
BNP	1.002	1.000–1.005	0.027
Pericardial effusion	11.985	1.650–87.053	0.014

Li Q, Dimopoulos K, et al, Eur J Prev Cardiol. 2019.11:2047487318821246

- PAH can develop at any stage of CHD patients' life, when it does impacts on quality of life exercise capacity, morbidity and mortality.
- Cautious approach for defects closure : 'I can close it' does not mean 'I should close it'.

- Pregnant women with PAH should be referred to PAH specialist centre with close MDT collaboration between pulmonary hypertension specialists, obstetricians, critical care specialists and neonatologists.
- Pregnancy is still associated with a significant risk of maternal mortality and rapid deterioration may occur requiring escalation of therapy and semi-urgent delivery.

Background

DoB:

- 9th March 1993 (M)

Cardiac Diagnoses:

- PA with intact ventricular septum and small ASD combined with MAPCAs

Previous Interventions:

- **19/3/1993:** Raskind septostomy (Aghia Sophia Children's Hospital)

Guy's Hospital (9/3/2003)

'This is a very complicated situation and the diagnosis **of pulmonary atresia with intact septum and MAPCA's is one we have never come cross before**. However, the problem is clearly that of insufficient pulmonary blood supply and I imagine that this child is severely cyanotic.

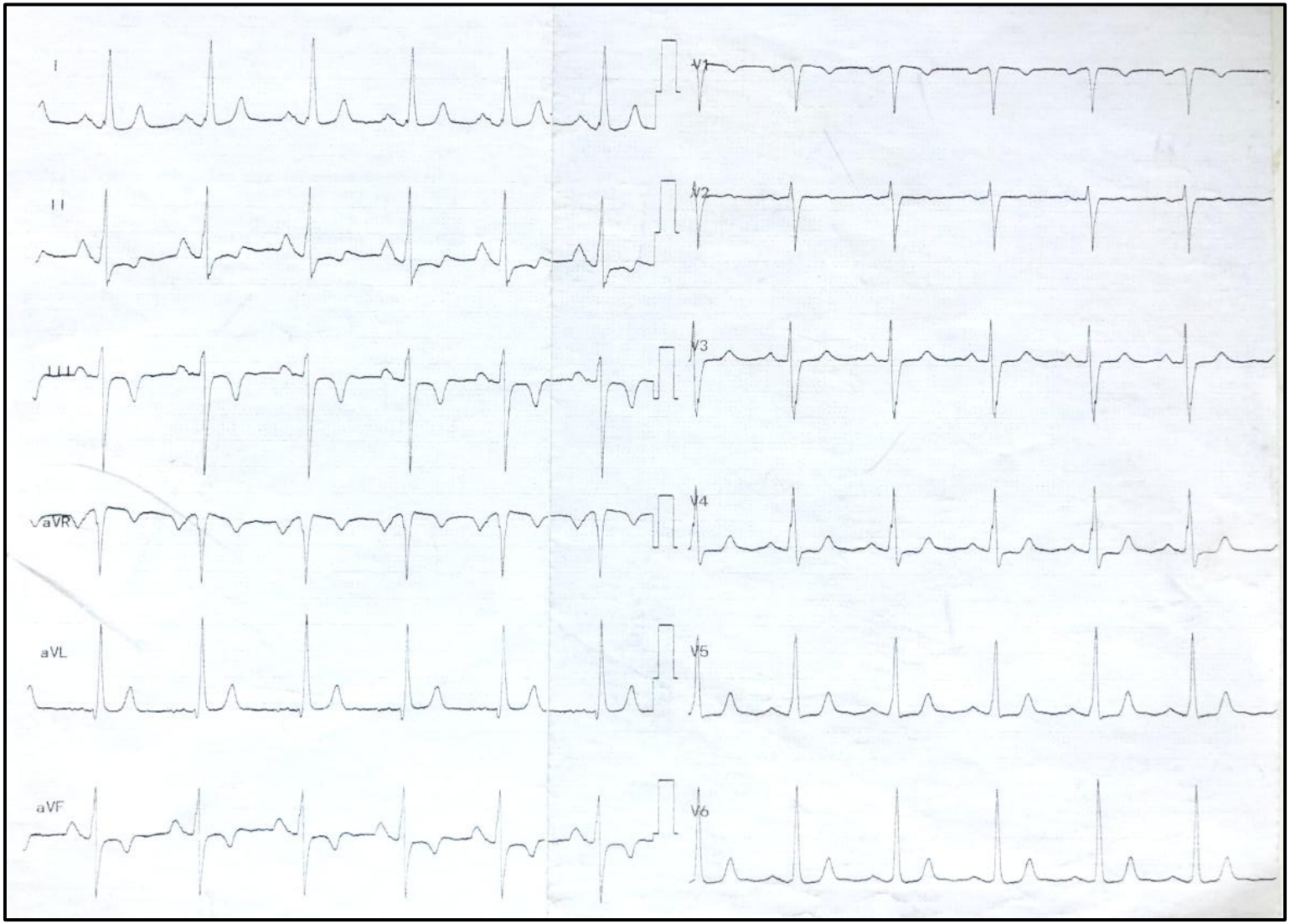
The **appearances of the collateral supply in the left lower zone** suggests that the region of the lung may well now have pulmonary vascular disease. Collateral also communicates well with the native central pulmonary arteries. The **collateral going to the right lung has very severe trifurcation stenosis** and that will have protected the distal pulmonary arteries.

It is very difficult to decide what may be best for the child. **Little useful surgery can be done** (shunt to central pulmonary artery to improve blood flow and encourage flow/ Fontan circulation)'

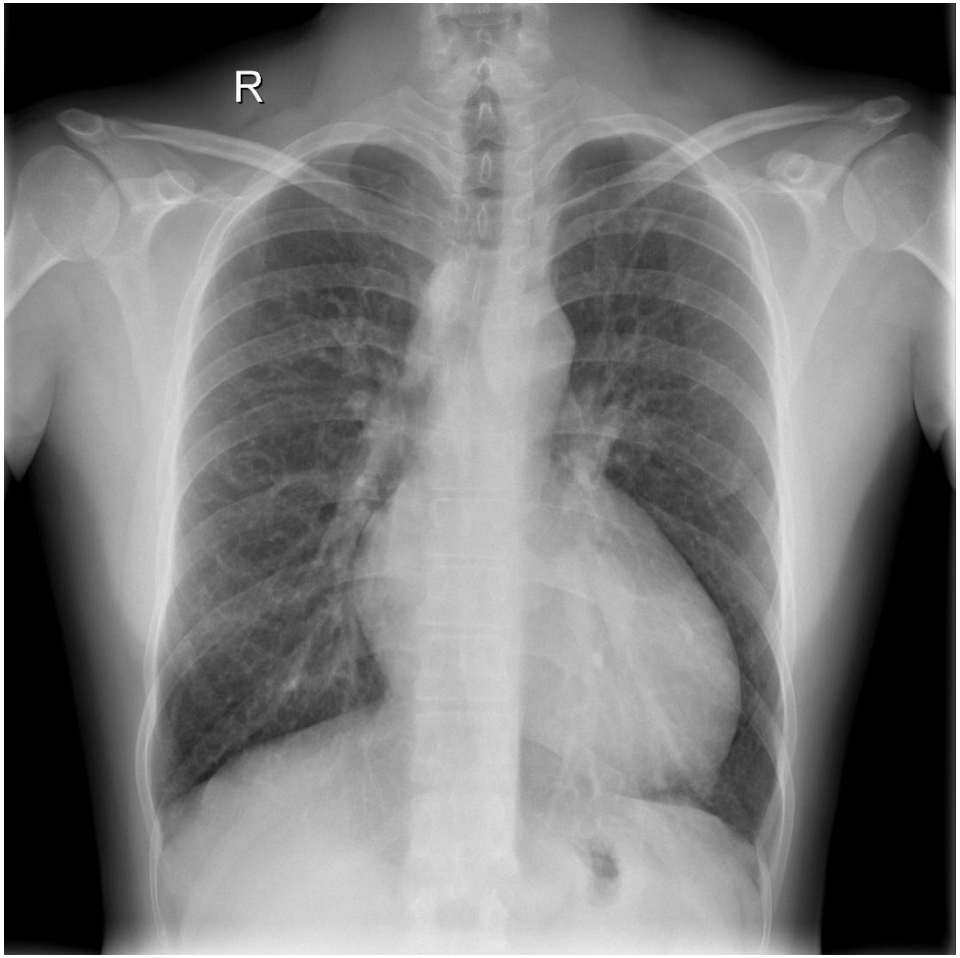
Mr D R Anderson

Consultant Cardiothoracic Surgeon

ECG (5/2019)



Chest X-Ray

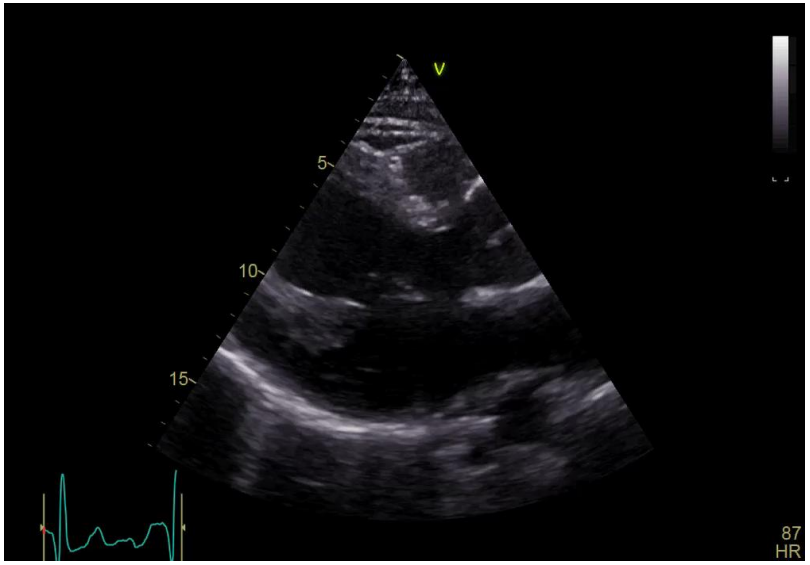


Labs

Περιγραφή εξέτασης	Ευρεθείσα Τιμή
WBC-Λευκά αιμοσφαίρια	8,08
RBC-Ερυθρά αιμοσφαίρια	**6,37**
HGb - Αιμοσφαιρίνη	**20,2**
HCT - Αιματοκρίτης	**59,5**
MCV - Μέσος όγκος ερυθρών	93,5
MCH - Μέση περιεκ.Hb/ερυθ	**31,7**
MCHC - Μέση πυκνότης Hb	34,0
RDW - Ευρος καταν.Ερυθρων	**15,5**
PLT - Αιμοπετάλια	211
HDW-Εύρος καταν. Hb	
MPV - Μέσος όγκος PLT	7,20
ΔΕΚ %	

ΤΥΠΟΣ ΛΕΥΚΩΝ	%	Τιμές Αναφοράς
Ουδετερόφιλα	56,5	40 - 74
Λεμφοκύτταρα	31,0	19 - 48
Μονοκύτταρα	6,0	3,4 - 9,0
Ηωσινόφιλα	1,8	0 - 7
Βασεόφιλα	**2,2**	0,0 - 1,5
Αλλα κύτταρα	2,5	0 - 4
Προμυελοκυτ.		
Μυελοκυτ		
Μεταμυελοκυτ		
Ραβδοπυρ.		
Βλάστες		
Άτυπα		

Περιγραφή εξέτασης	Ευρεθείσα Τιμή	Τιμές Αναφοράς
Σάκχαρο	89 mg/dL	70 - 105
Ουρία	40 mg/dL	18 - 55
Κρεατινίνη	1,0 mg/dL	0,72 - 1,25
Κάλιο	4,2 mmol/L	3,5 - 5,1
Νάτριο	**135 mmol/L**	136 - 145
Ασβέστιο	9,9 mg/dL	8,40 - 10,20
SGOT	34 U/L	5 - 34
SGPT	54 U/L	0 - 55
LDH	**350 U/L**	125 - 220
CPK TOTAL	84 U/L	30 - 200
CPK -MB	9,0 IU/L	12% της TOTAL CPK
Αλκαλική φωσφατάση	64 U/L	40 - 150
γ-GT	51 U/L	12 - 64
Αμυλάση	60 U/L	20 - 160
Ολική Χολερυθρίνη	**3,08 mg/dL**	0,20 - 1,20
Αμεση Χολερυθρίνη	**0,79 mg/dL**	0,00 - 0,50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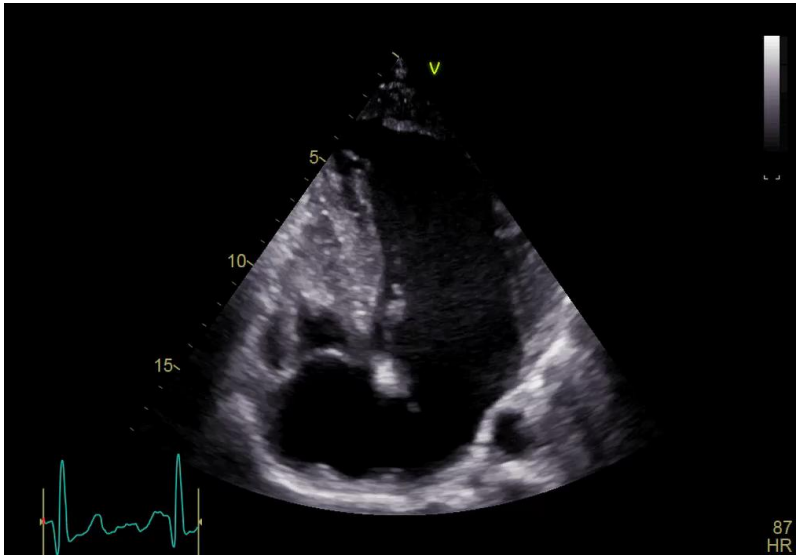
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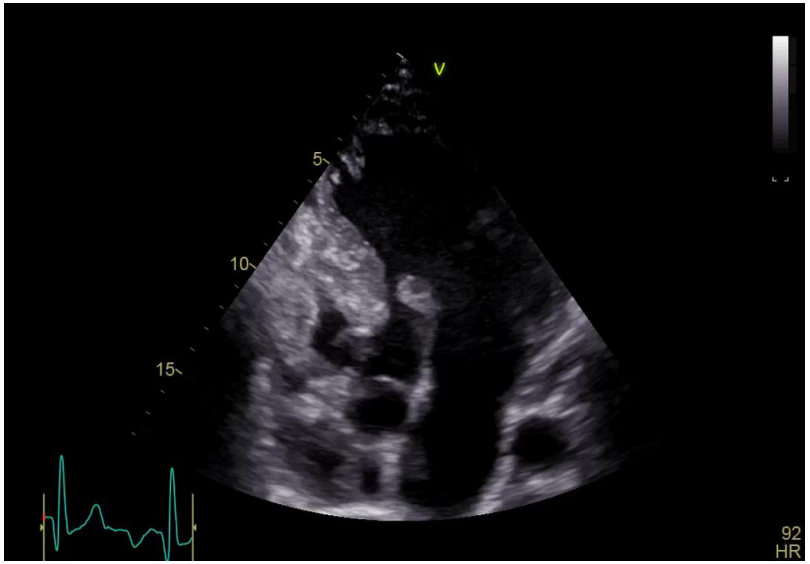
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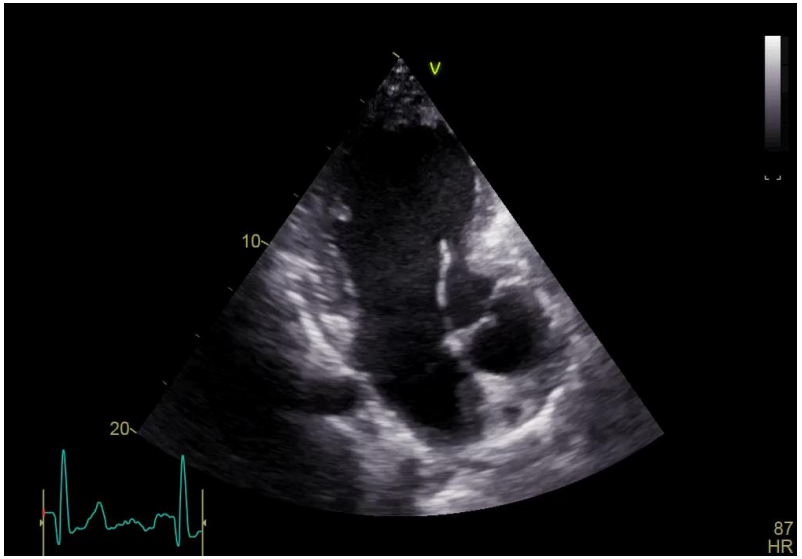
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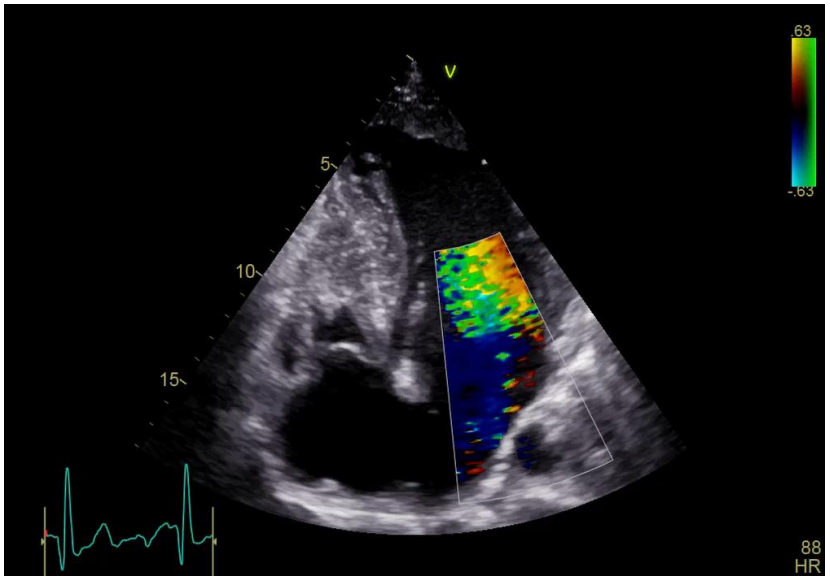
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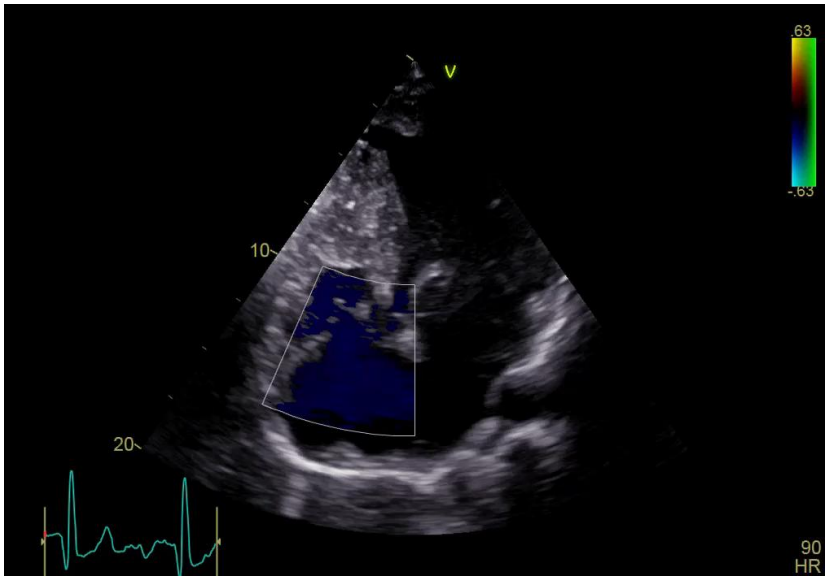
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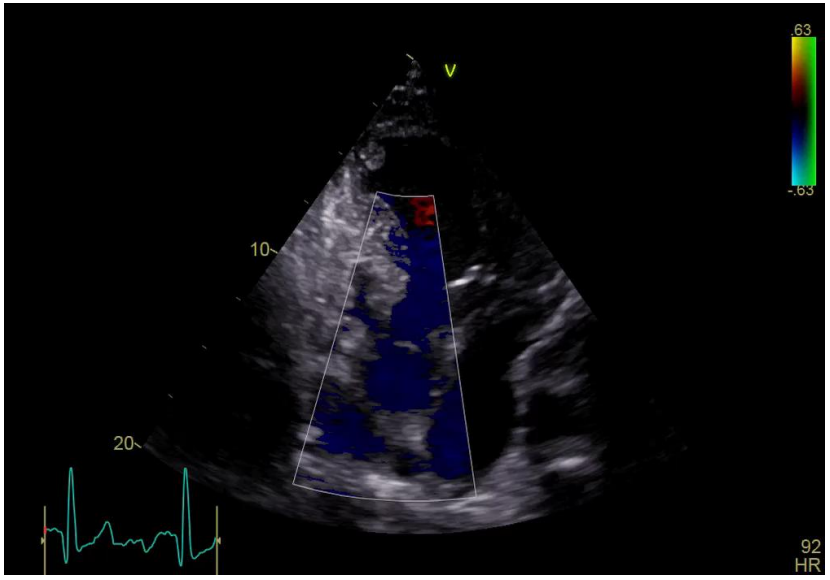
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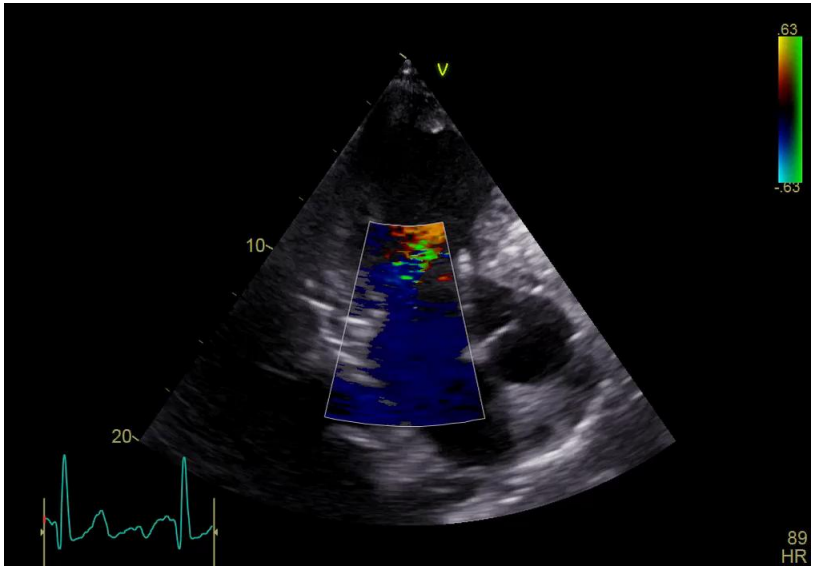
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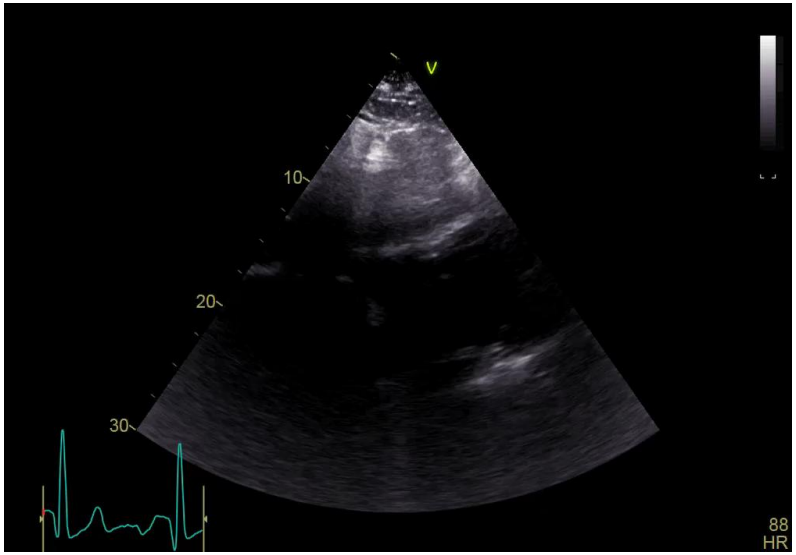
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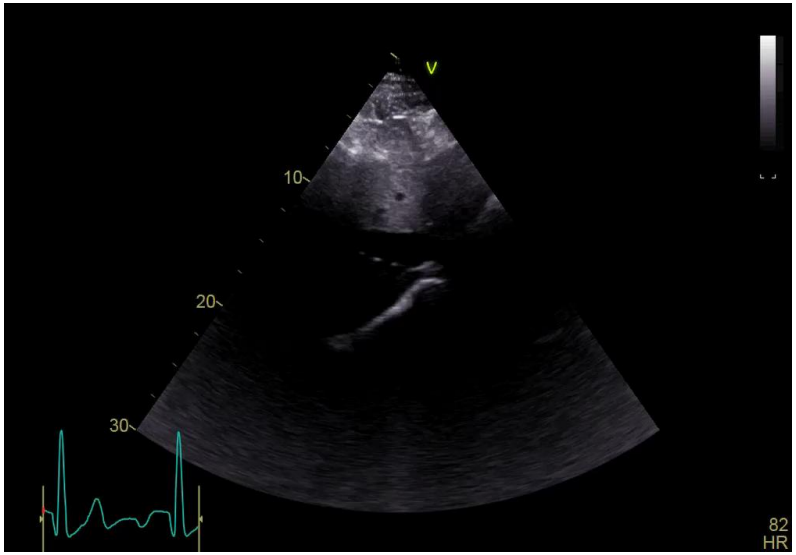
HR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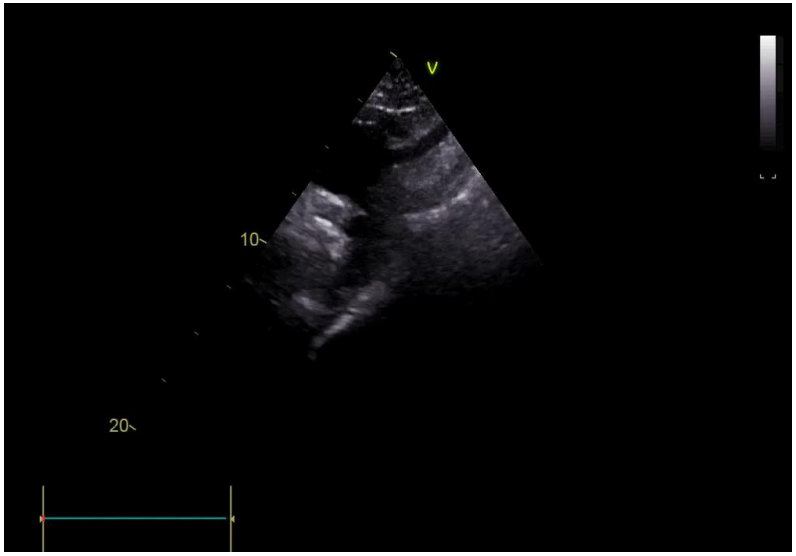
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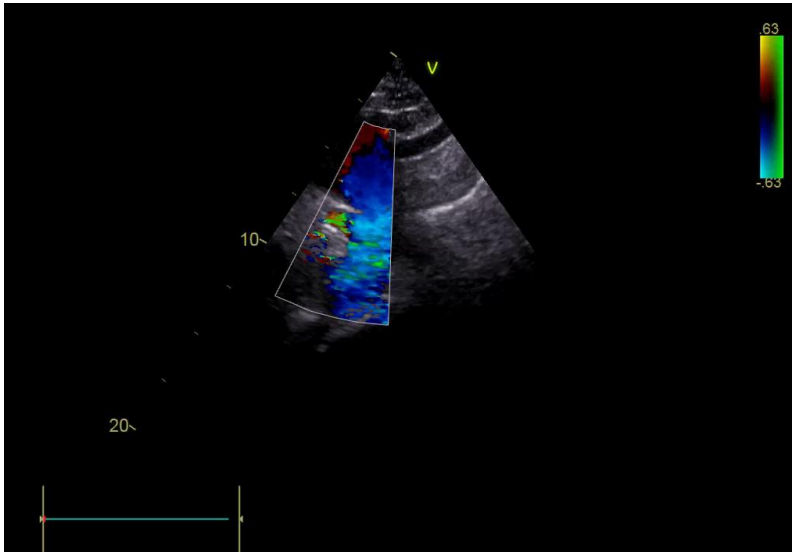
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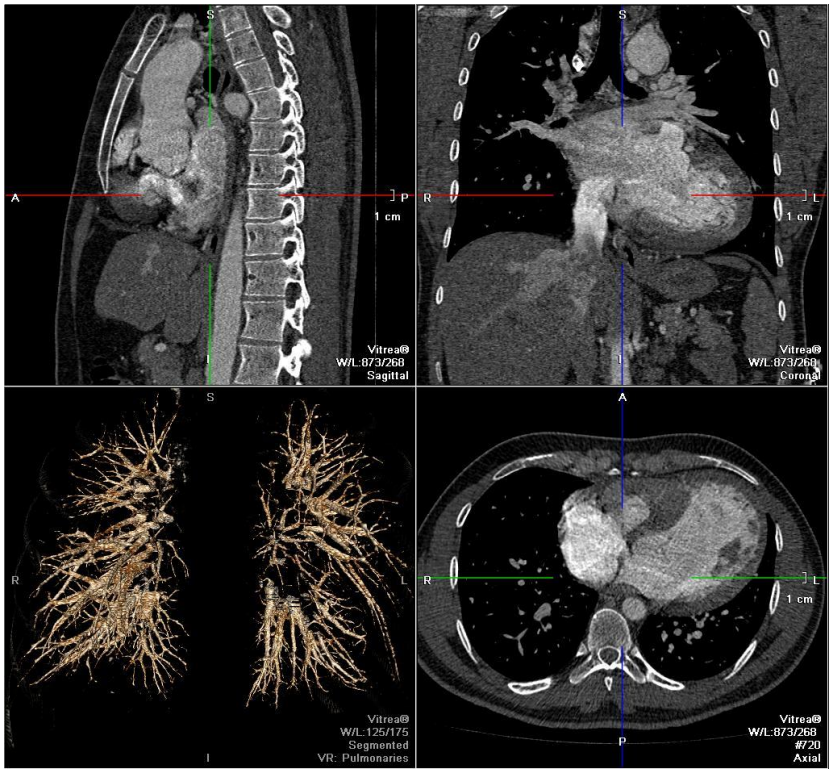
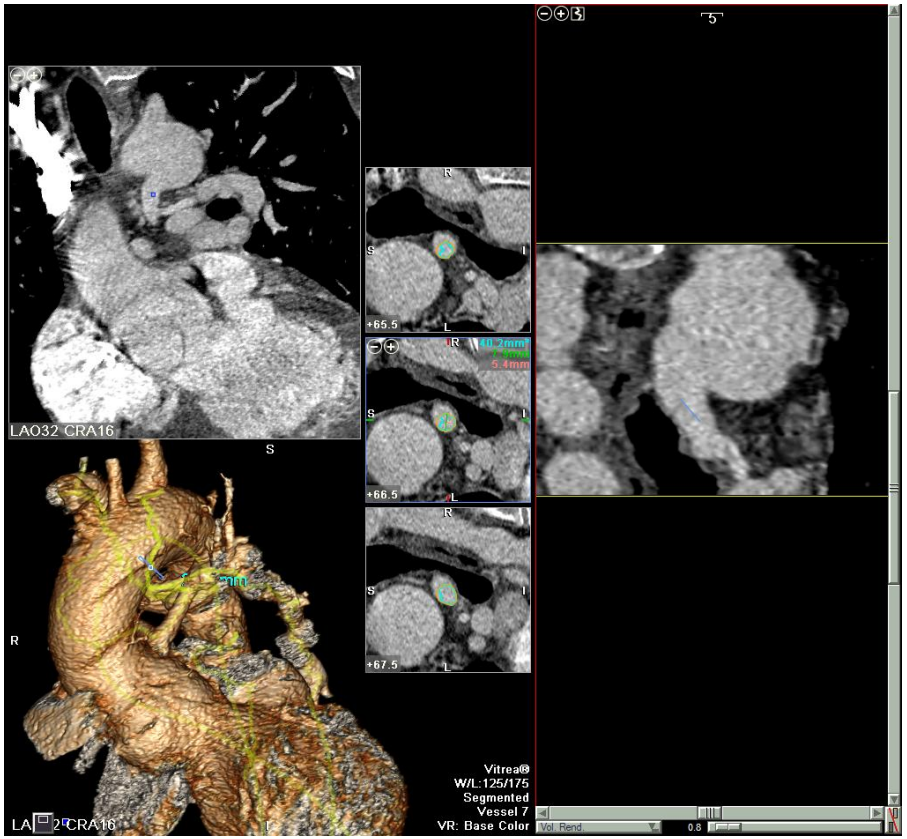
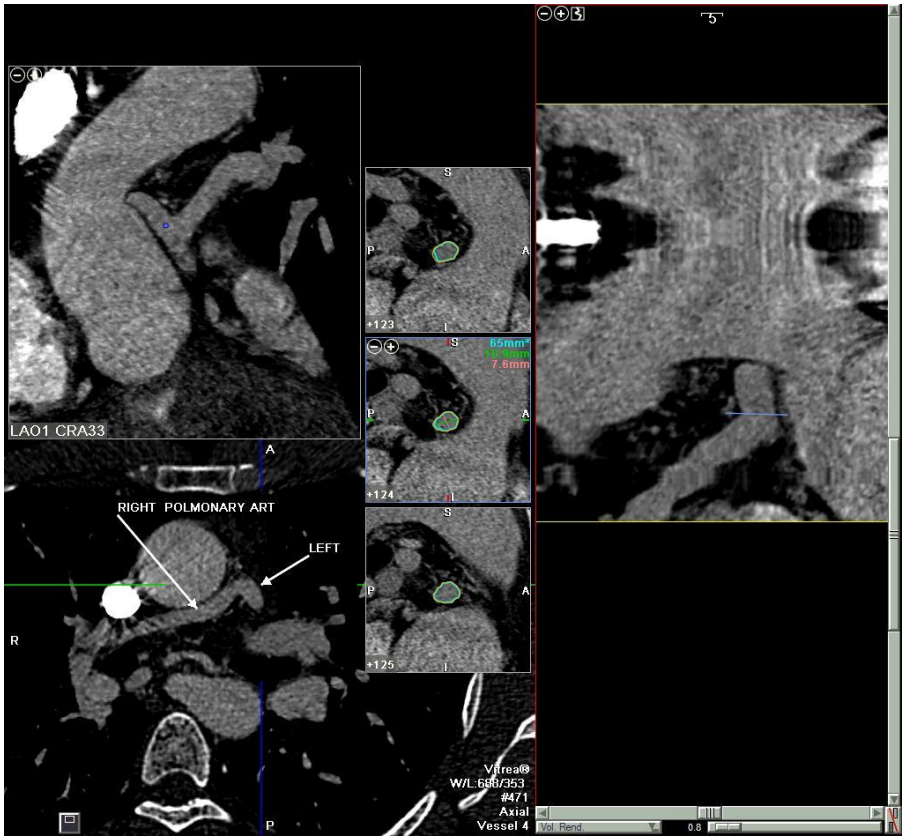
CT angiography



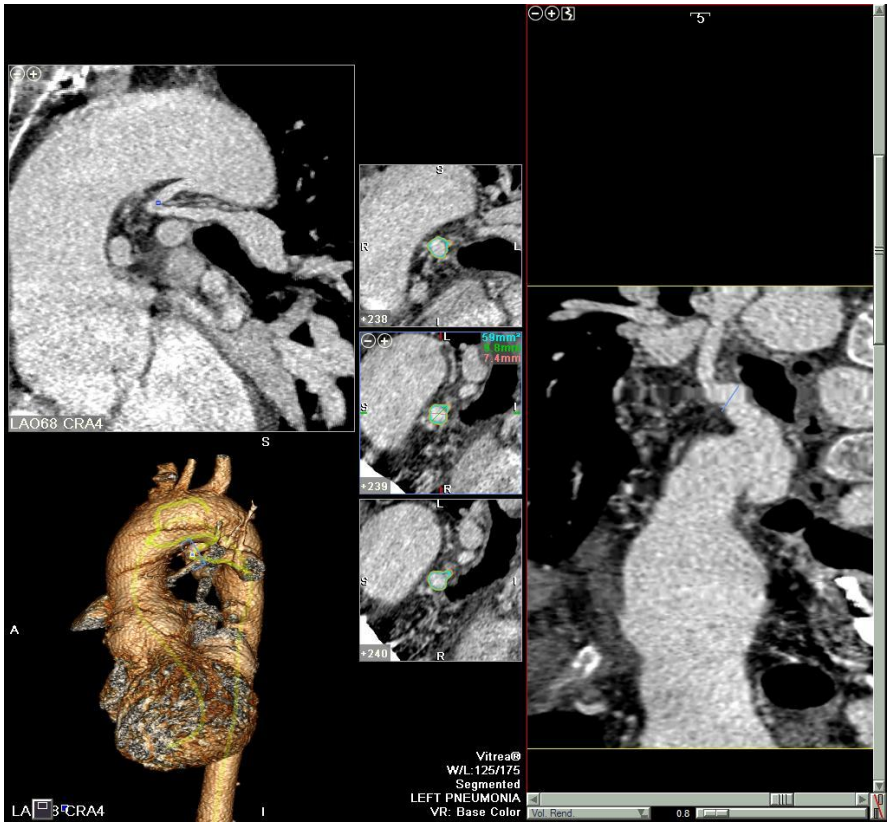
CT angiography

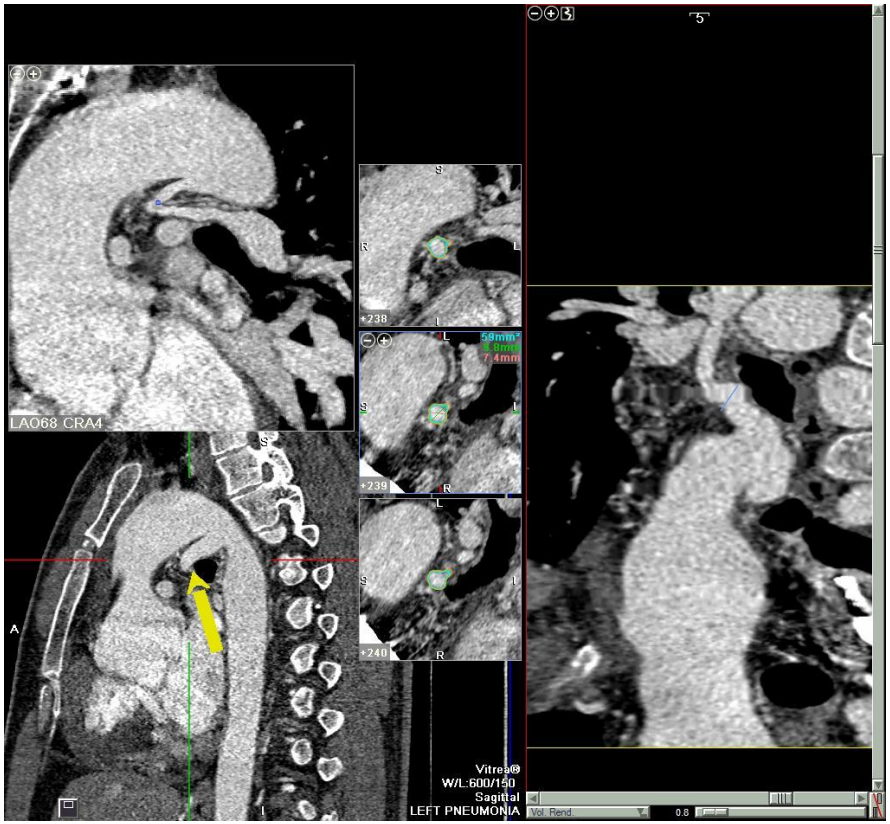




Non- confluent aortic root

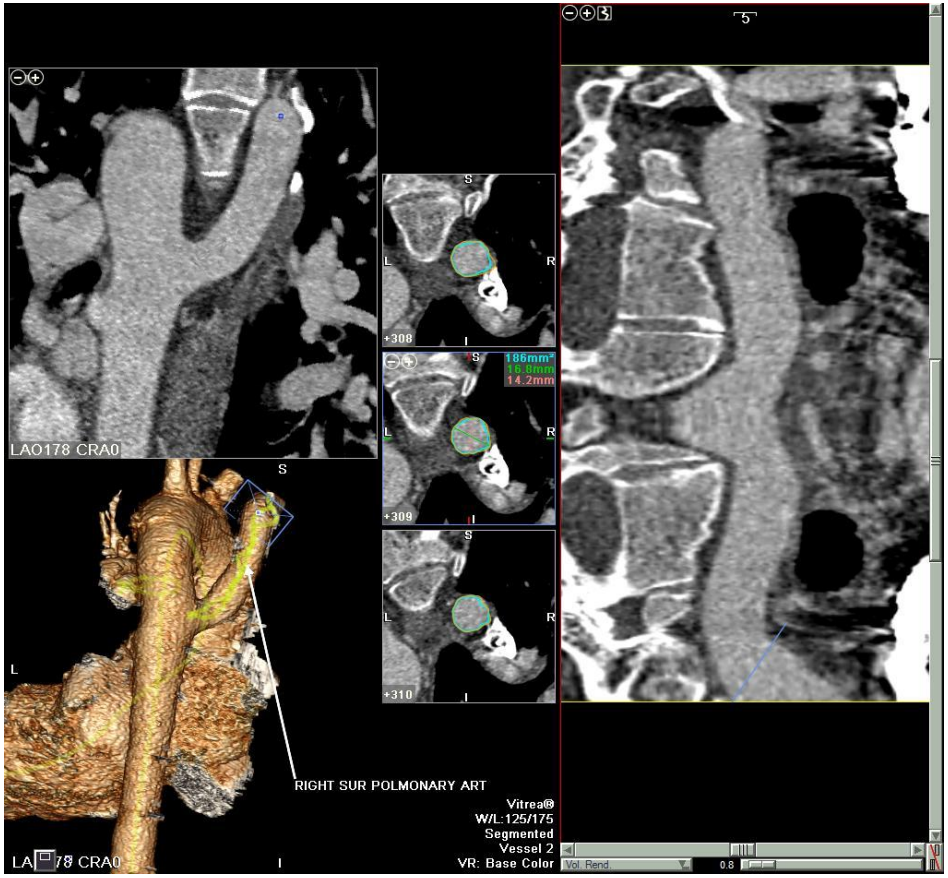
CT angiography



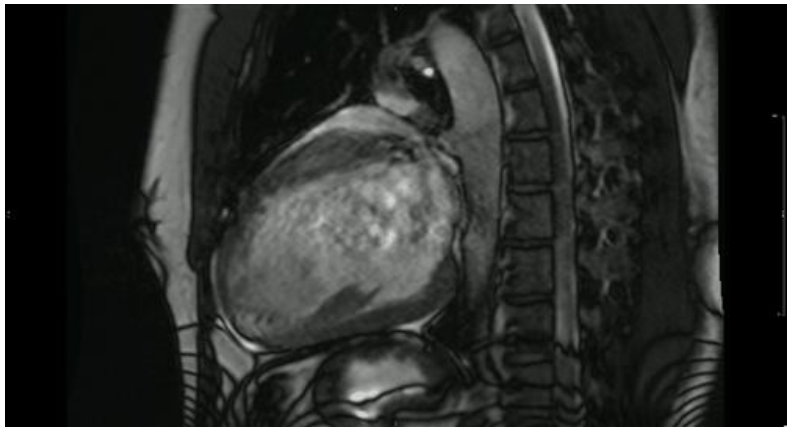


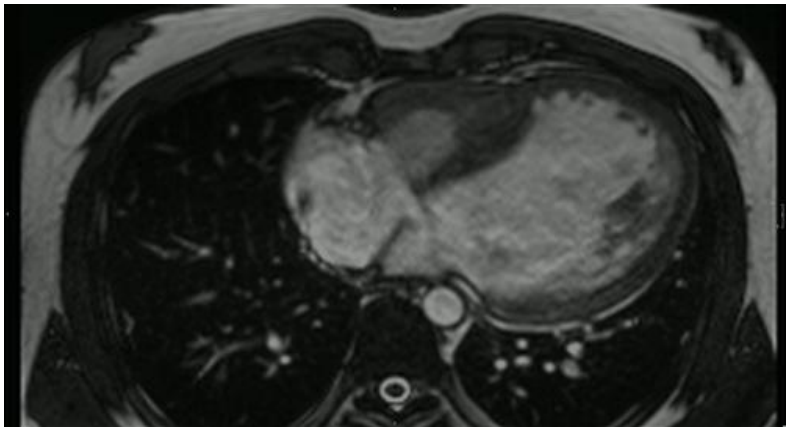
Aortic arch to left upper lobe with stenosis

CT angiography



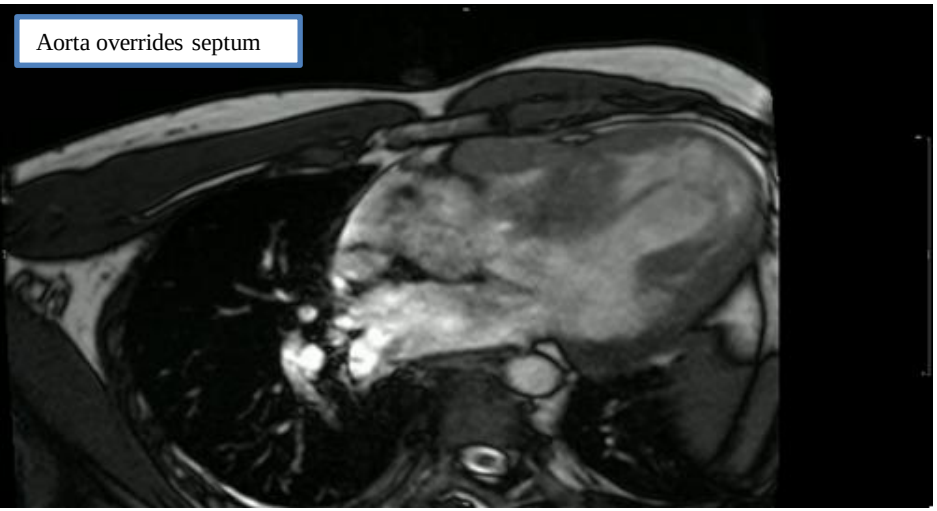
Right side descending aorta right upper lobe
Left side descending aorta left lower lobe with stenosis and right lower lobe

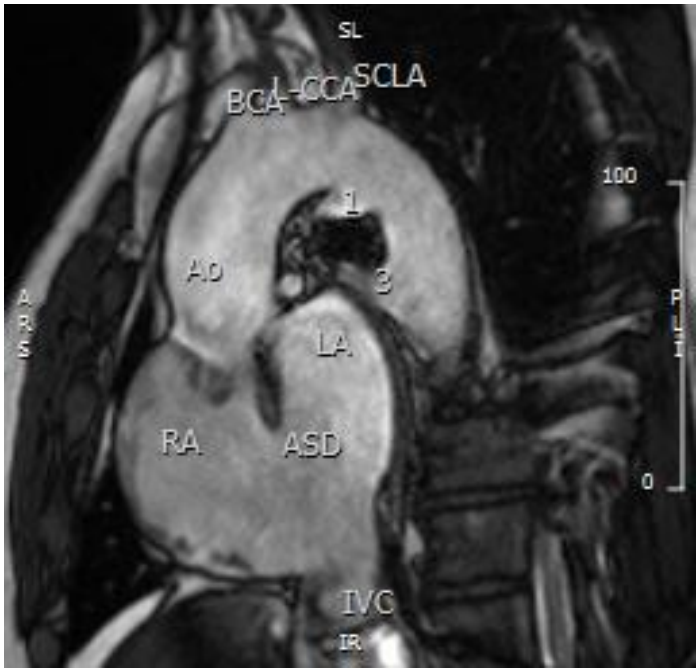




Dilated LV (EDVi 191ml/m2) with preserved EF 59%
Severely hypoplastic RV with severe hypertrophy-

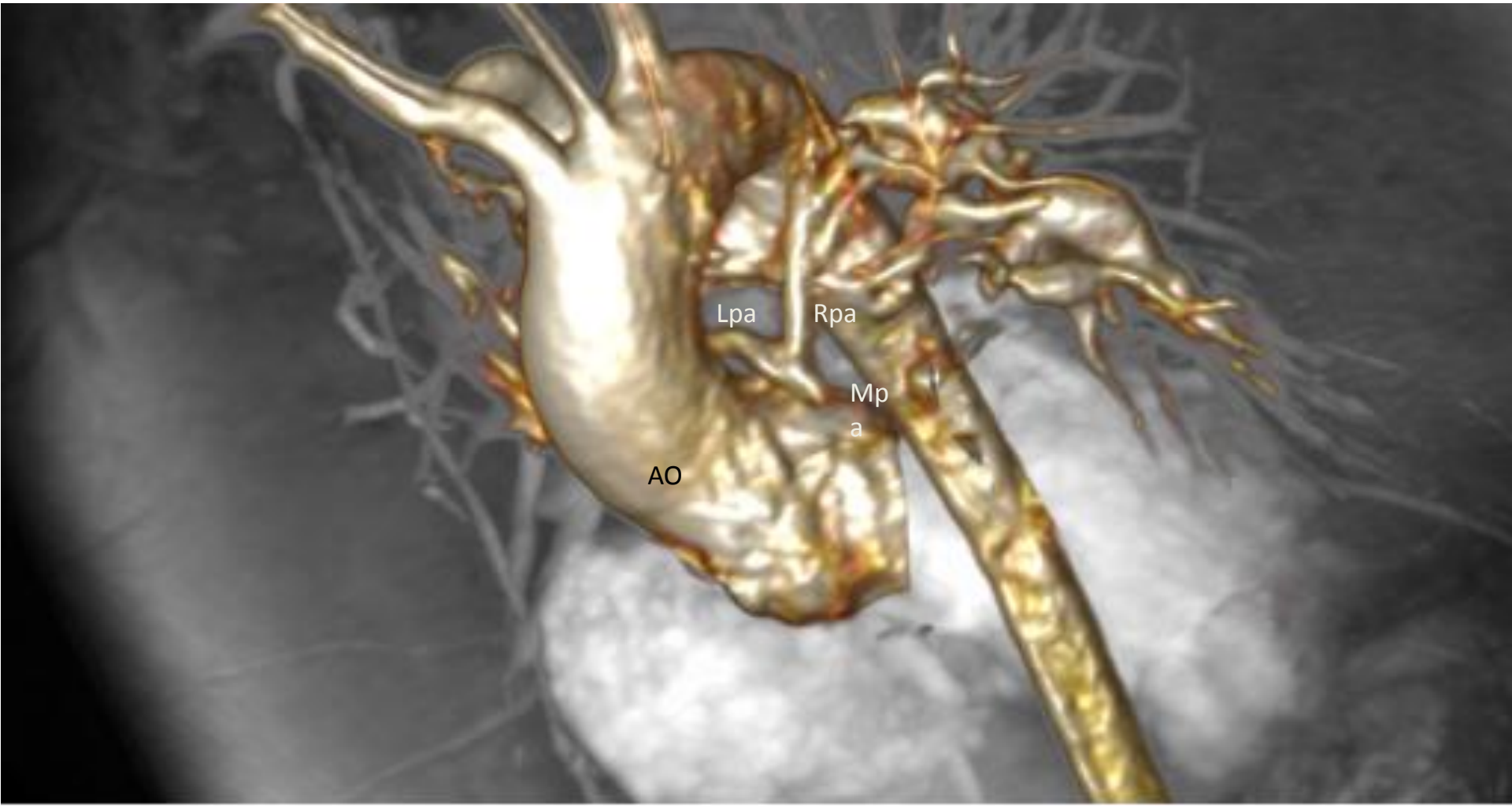
Aorta overrides septum

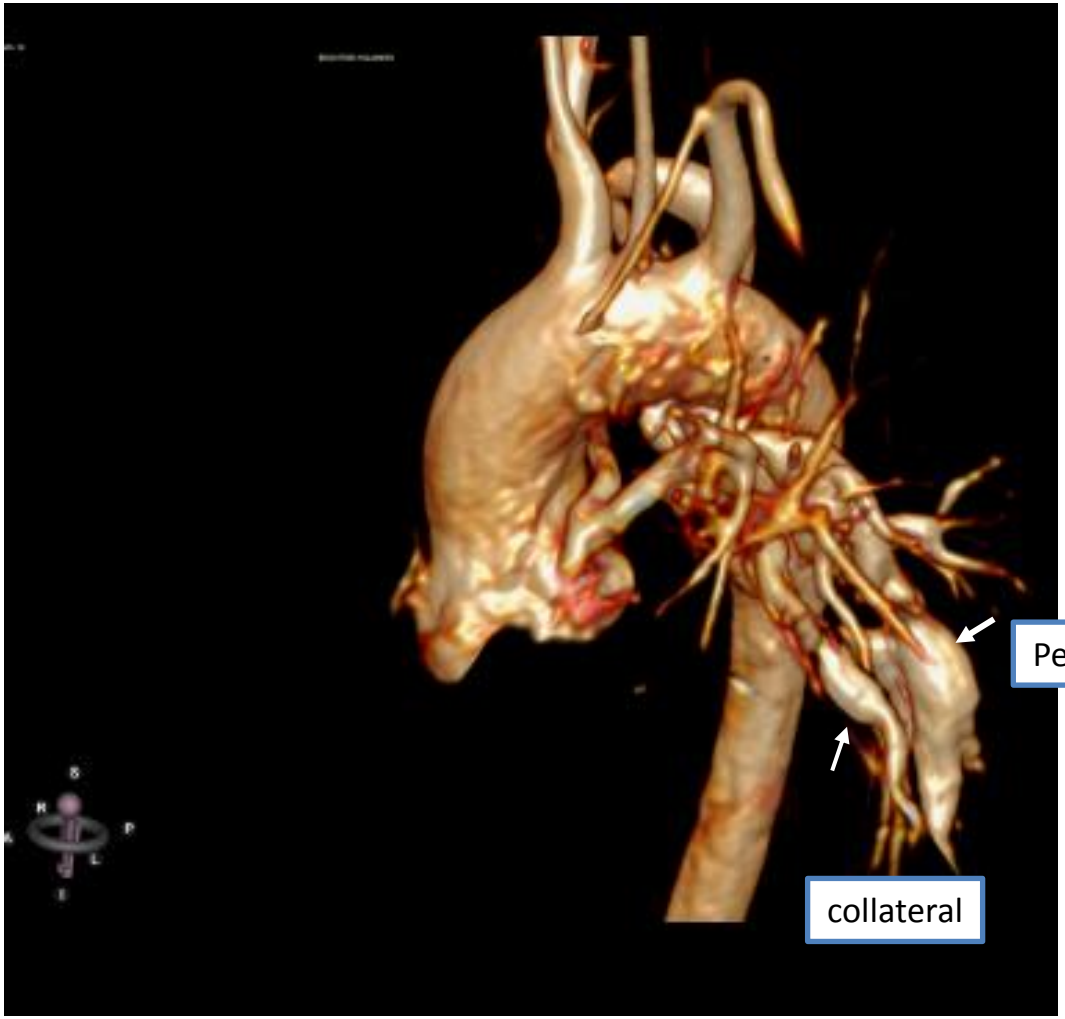


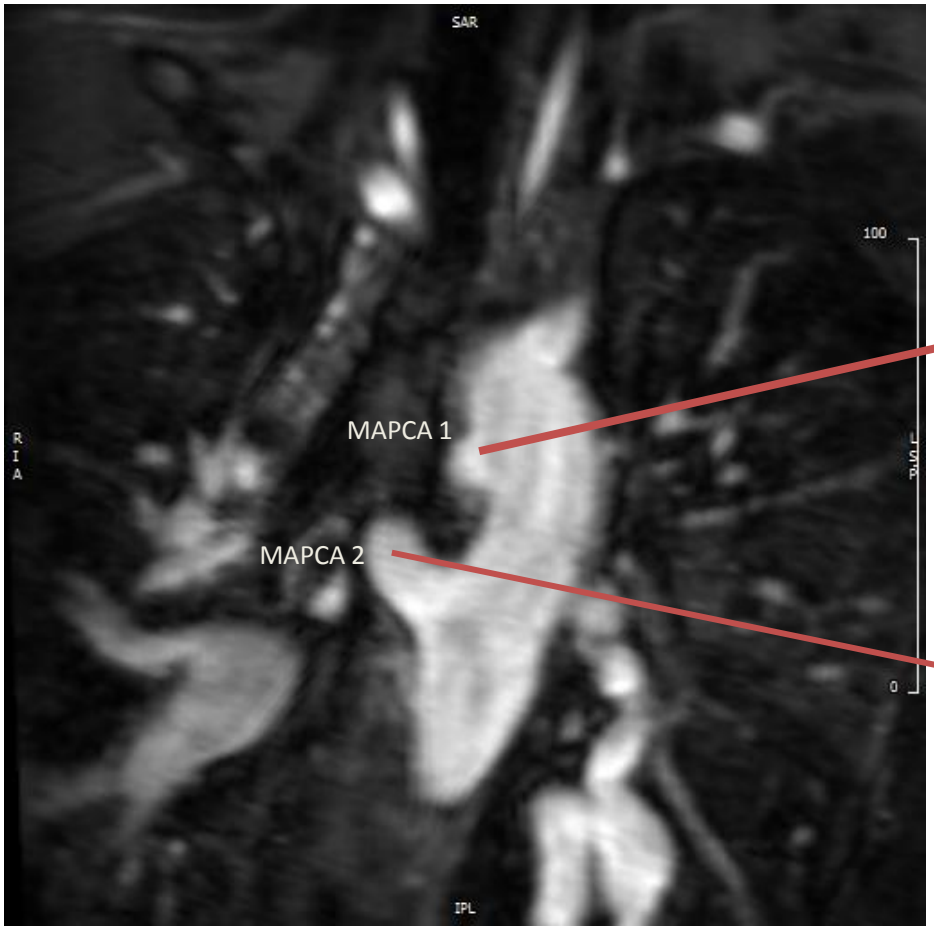


Septostomy- unrestricted ASD

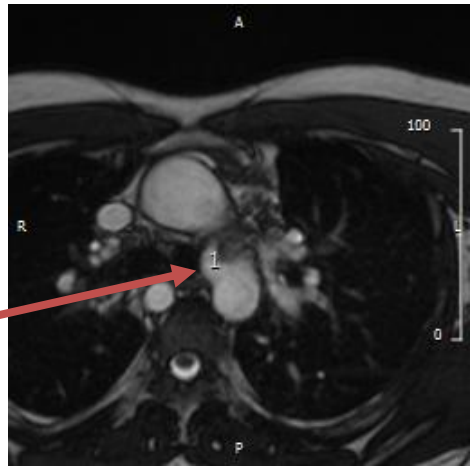
SA cine stack
Dilated LV, severely hypoplastic RV
Ao overriding septum

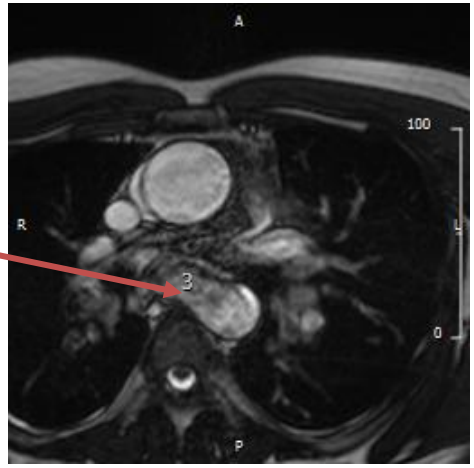

Lpa
Rpa
Mp
a
AO


Peripheral LPA
collateral



Right lung






S
Collateral 2
R
A
10
0
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S
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Large collateral MAPCA 2 from upper desc Ao
Feeding R-lung

Definition of Segmental PAH

‘Observed in discrete lung areas perfused by aortopulmonary collaterals in congenital heart diseases such as pulmonary or tricuspid atresia’.

World Pulmonary Hypertension Symposium in 2013, J Am Coll Cardiol. 2013;62:D34–D41

‘PH in one or more lobes of one or both lungs.’

ESC 2015 guidelines, Eur Heart J. 2016;37:67–119

‘PH that does not follow a homogeneous distribution, with some parts of the pulmonary vasculature being exposed to higher pressures than others’

Int J Cardiol.2013;164:106–110.

Segmental pulmonary hypertension encompasses any condition with abnormal underlying cardiac or vascular anatomy, usually including varied sources of pulmonary blood supply, which results in distal pulmonary vascular disease that affects various lung segments to differing degrees.

Dimopoulos et al, J Am Heart Assoc. 2018;7:e008587

Classification

1. **RV communicating directly with the entire pulmonary vascular bed** (eg, large VSD with peripheral PS and VA concordance)
2. **RV supplies part of the pulmonary vascular bed** (eg, congenital absence/interruption of a pulmonary artery supplied by large collaterals/isolated pulmonary artery of ductal origin or a PDA, hemitruncus arteriosus).
3. **RV with no direct communication** with the pulmonary vascular bed:

With well-formed (native) PAs (eg, truncus arteriosus with PA stenosis);

With ill-formed PAs and a pulmonary circulation supplied by collateral arteries, a PDA, or surgical shunts (eg, TOF] with pulmonary atresia, often referred to as complex pulmonary atresia throughout this article

Dimopoulos et al, J Am Heart Assoc. 2018;7:e008587

To Which WHO PH Group Does Segmental PH Best Belong?

- **Group 1:** Common histological features
- **Group 4:** pulmonary arterial obstructions (congenital branch PA stenosis, CTEPH)
- **Group 5**: fibrosing mediastinitis or pulmonary arterial compression by tumors

5. Pulmonary hypertension with unclear and/or multifactorial mechanisms

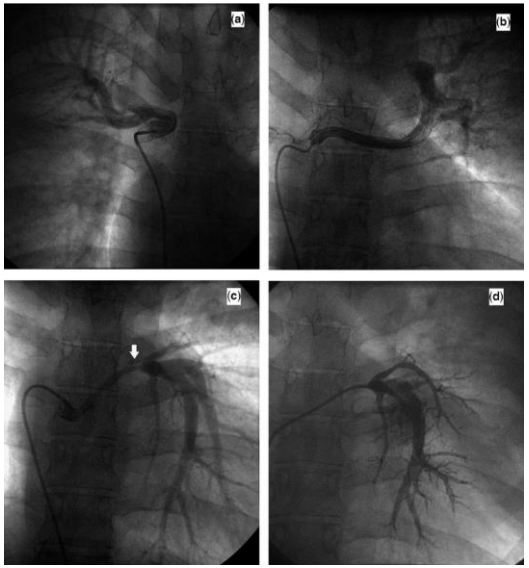
5.1 Haematological disorders: chronic haemolytic anaemia, myeloproliferative disorders, splenectomy
5.2 Systemic disorders: sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis, neurofibromatosis
5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
5.4 Others: pulmonary tumoral thrombothic microangiopathy, fibrosing mediastinitis, chronic renal failure (with/without dialysis), segmental pulmonary hypertension

- At present, it may seem appropriate to classify segmental PH within group 5, multifactorial PH, reinforcing the complexity and unique physiology of this condition.
- However, inclusion within group 1 (PAH related to CHD) may have the merit of reminding physicians this is a CHD-related condition, with significant similarities in pathophysiology to other CHD related PAH (eg, chronic cyanosis) and may help inclusion in future research.

Effect of PAH therapy in patients with segmental PH remains a debate

Patient	Parameter	Baseline	6 months	2 years	5 years	10 years	14 years
1	NYHA class	III	II	II	II	II	II
	O_2 sat (%)	85	81	85	82	82	83
	VO_2 max (ml/kg/minute)	10.5	11.7	13.6	12.7	16.7	12.4
	6MWD (m)	415	360	400	410	410	420
2	NYHA class	III	II	II	II	II	II
	O_2 sat (%)	80	84	81	82	82	–
	VO_2 max (ml/kg/minute)	13.2	13.2	13.0	12.7	12.5	–
	6MWD (m)	300	330	330	320	310	–
3	NYHA class	IV	II	II	II	II	II
	O_2 sat (%)	52	68	70	78 (2.5 years)	–	–
	VO_2 max (ml/kg/minute)	–	–	–	–	–	–
	6MWD (m)	–	–	–	–	–	–

O_2 sat = oxygen saturation; VO_2 max = maximal oxygen consumption; 6MWD = 6-minute walking distance

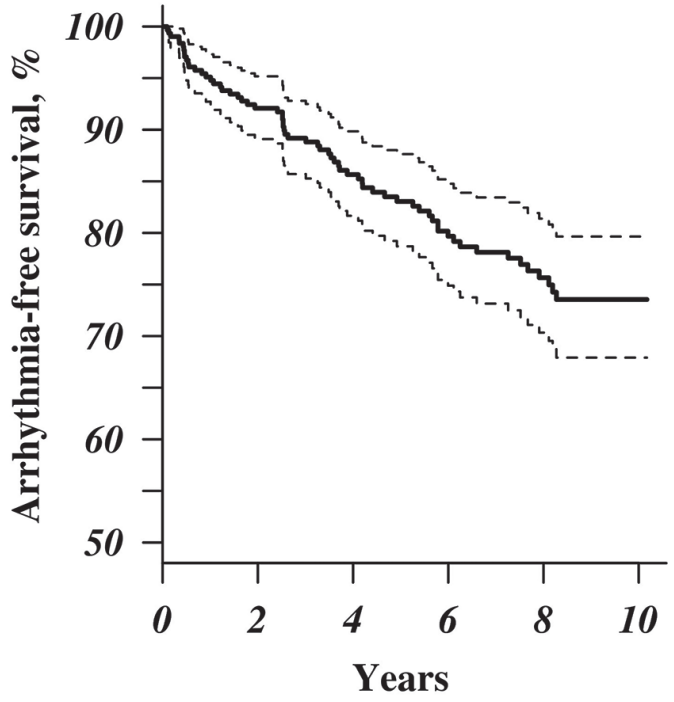


- 3 adult patients with unrepaired ToF, PA, MAPCAs and segmental PAH.
- Patients improved by 1–2 NYHA classes with modest exercise-tolerance increase, and remained stable without side effects during 2.5, 10, and 14 years.

Apostolopoulou et al, Cardiology in the Young (2017), 27, 1861–1864

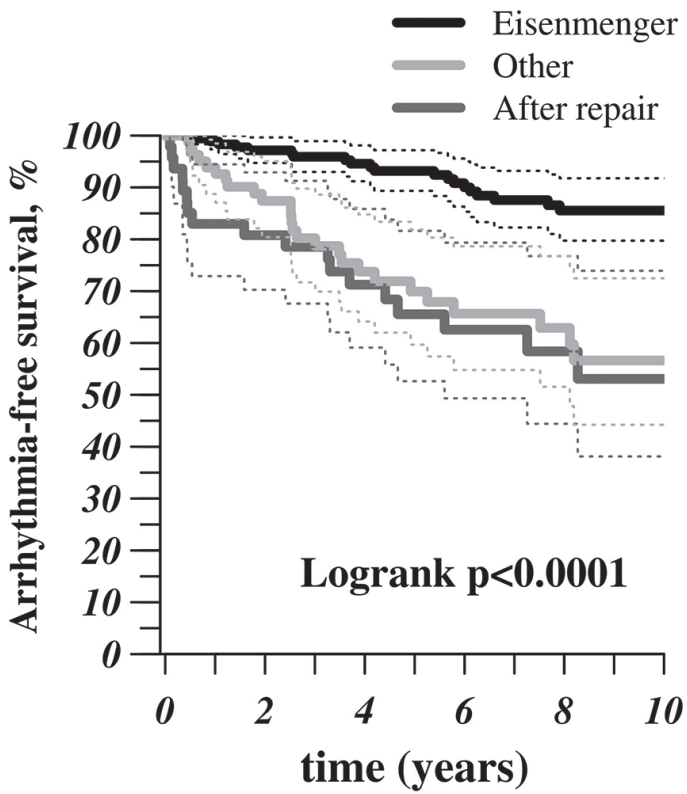
- A broad recommendation on the use of PAH therapies cannot be made at present given the lack of evidence, though anecdotal experience suggests these therapies may have a role and may be considered empirically on an individualized basis in patients with confirmed segmental PH.
- Because of significant heterogeneity, coupled with a small patient population, it is unlikely that adequately powered randomized controlled trials will ever be feasible.
- However, well-structured prospective registries with prespecified baseline and follow-up protocols may shed additional light on the natural and unnatural history, and optimal management of segmental PH

Arrhythmias in ACHD patients with PAH



n at risk	310	262	205	160	113	22

A



Eisenmenger	181	163	135	113	80	17
Other	82	62	43	29	20	4
After repair	47	37	27	18	13	1

B

M. Drakopoulou, H. Nashat, A. Kempny, R. Alonso-Gonzalez, L. Swan, S. J Wort, L C Price, C. McCabe, T. Wong, M. A Gatzoulis, S. Ernst, K. Dimopoulos Heart 2018;0:1–7