



Important points case consideration III: Interventions in GUCH

Worakan Promphan, MD.FSCAI.

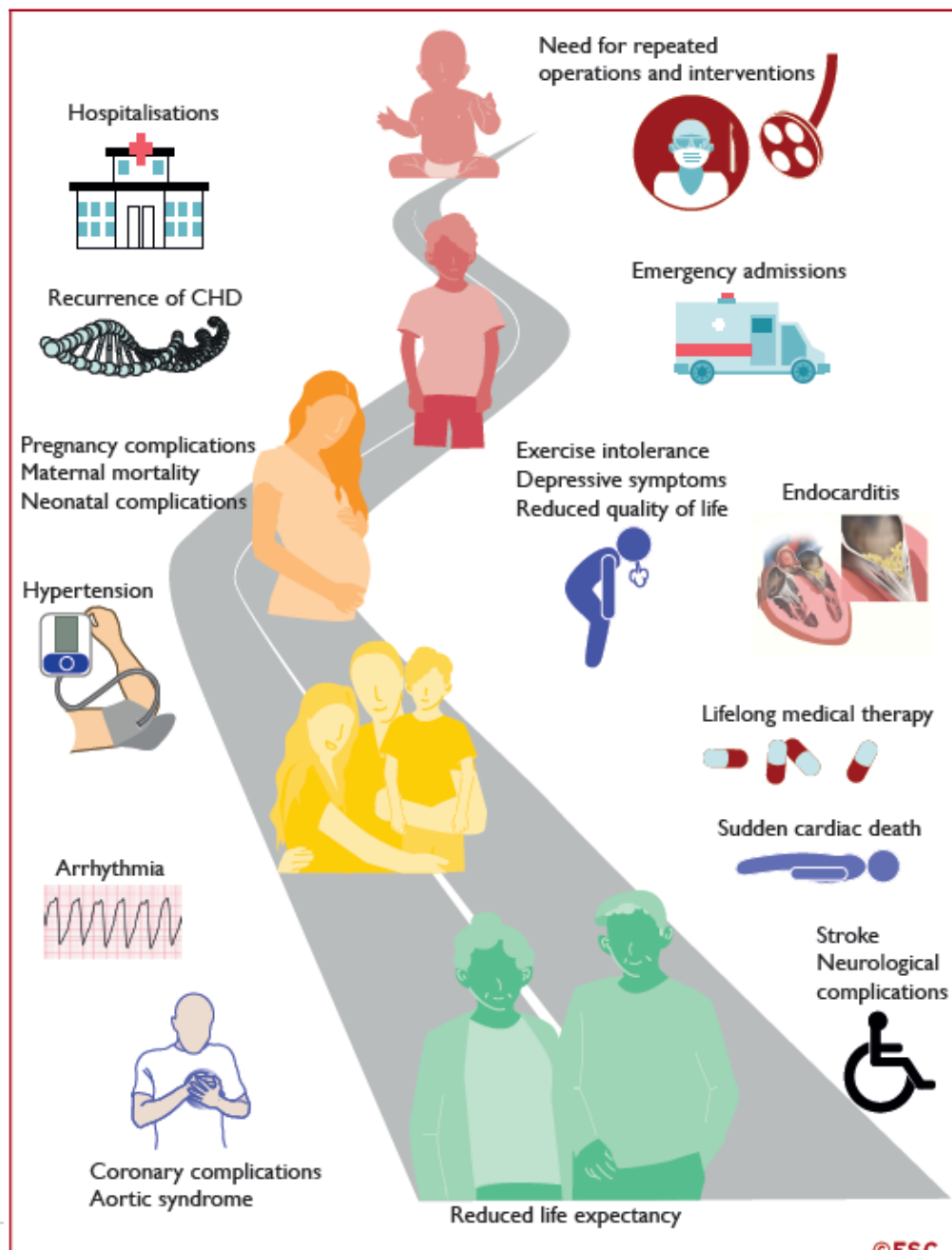
Kanyalak Vithessonthi, MD.

Worakan Promphan

Proctor: Abbott, Occlutech, Lifetech Scientific, Lepu Medical,
Venus Medtech

Kanyalak Vithessonthi

No disclosure

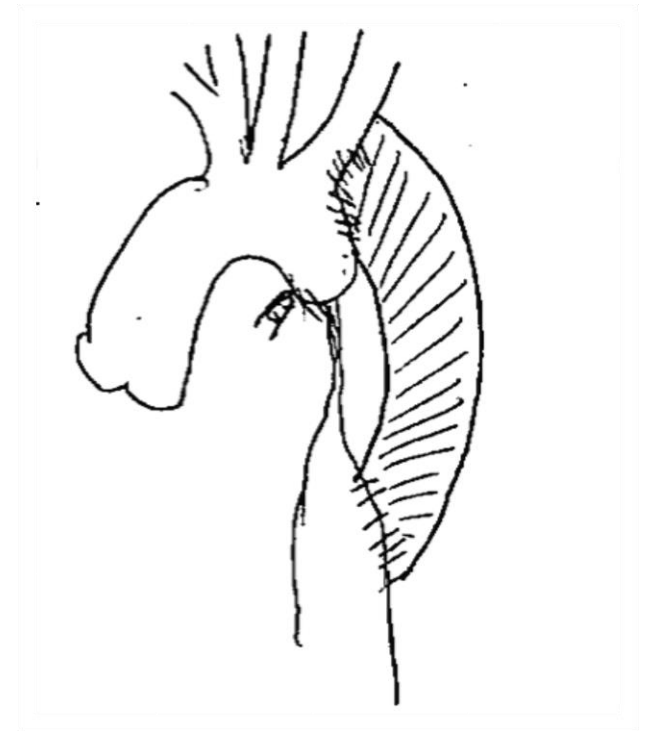


Congenital heart disease A lifelong chronic condition

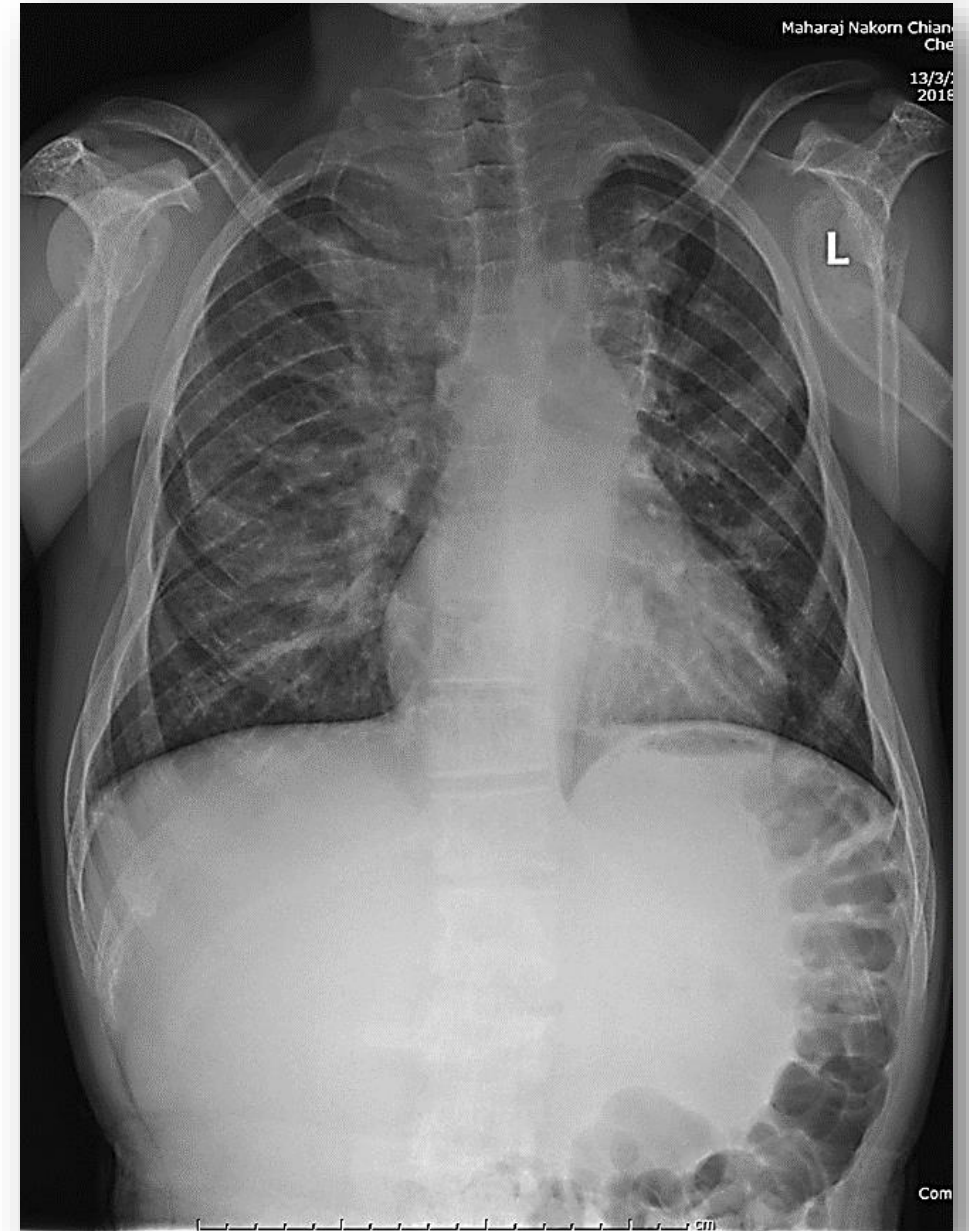
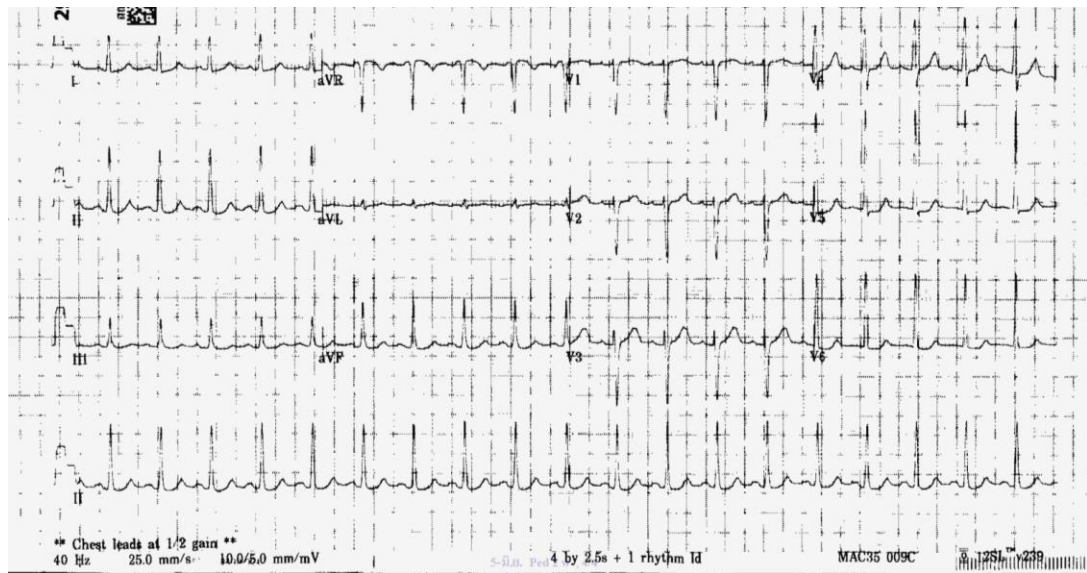


Case 1

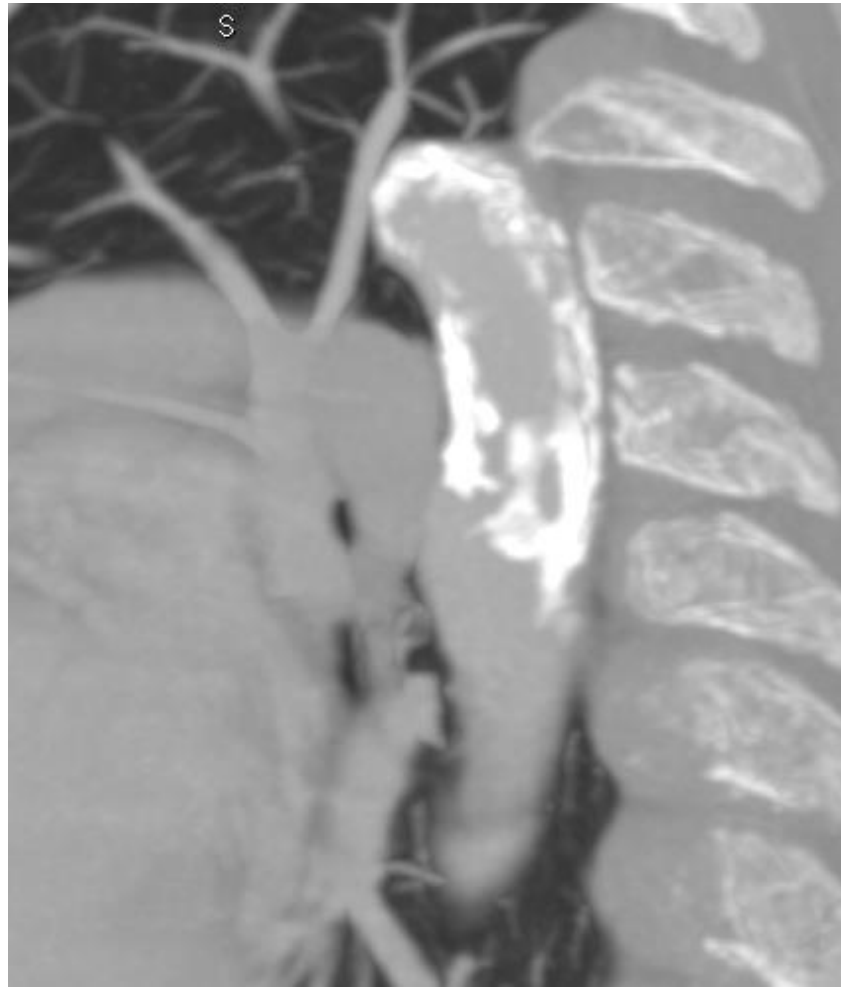
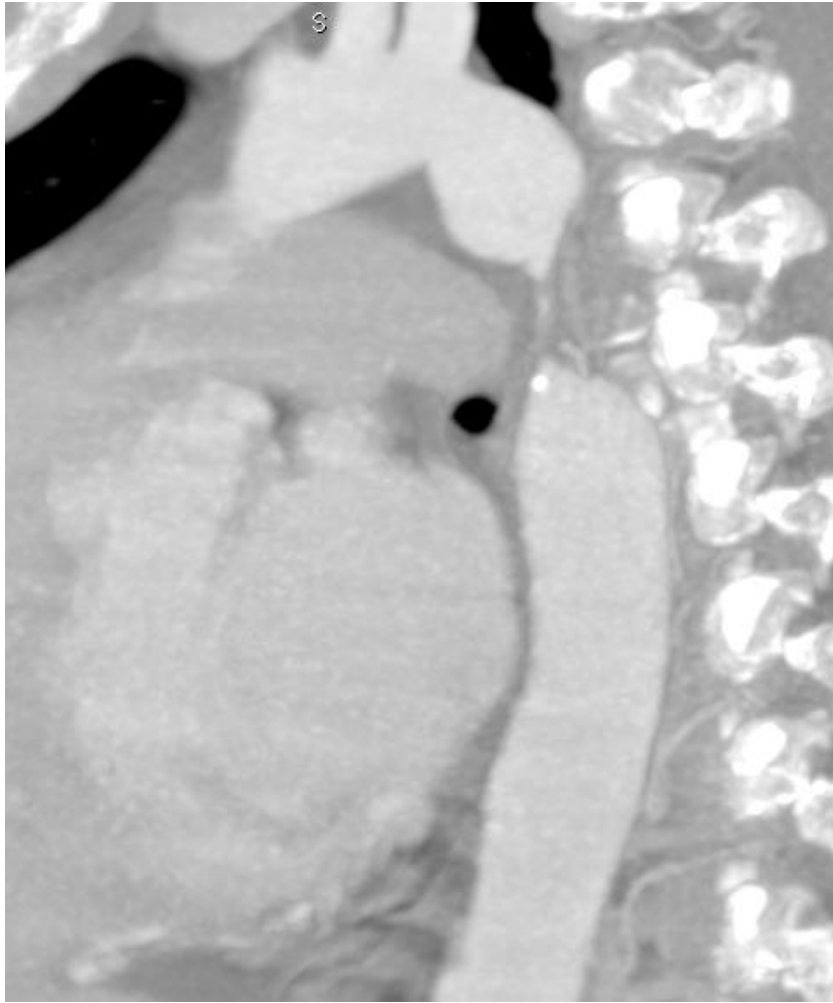
- 23-year-old male, thalassemia major, s/p splenectomy
- Regular blood transfusion monthly
- 2006 (11y): Hypertension
 - Long segment CoA (4-5 cm)
 - Aorto-aortic bypass, PTFE graft 22 mm
 - Uneventful post-op with normal BP and no medication



- Regular follow-up yearly: uneventful
- 2017 (23 y): New onset HT
 - Right arm 143/59 mmHg
 - Right leg 119/53 mmHg



23 years old, s/p aorto-aortic bypass for 11 years, recurrent HT, conduit stenosis with heavy calcification



CoA AHA Recommendations

Significant native or recurrent aortic CoA

1. UE/LE resting peak-to-peak gradient >20 mm Hg or mean Doppler systolic gradient >20 mm Hg.
2. UE/LE gradient >10 mm Hg or mean Doppler gradient >10 mm Hg
 - 2.1 decreased LV systolic function or AR
 - 2.2 with collateral flow

Therapeutic

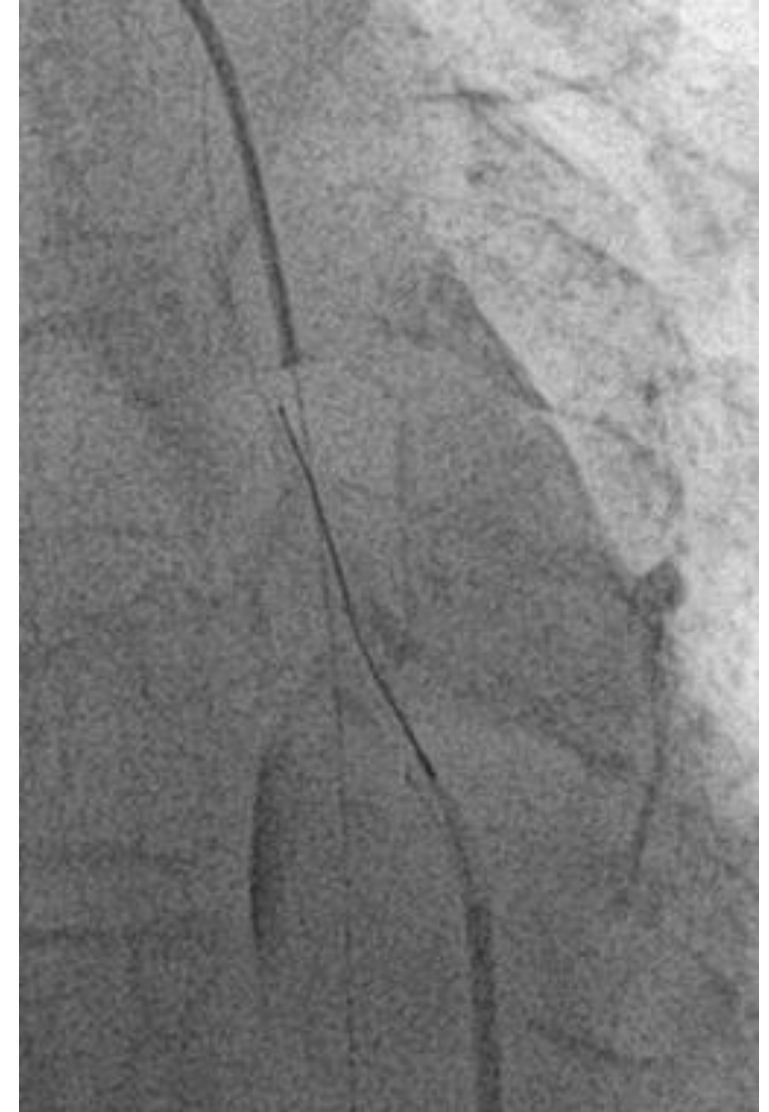
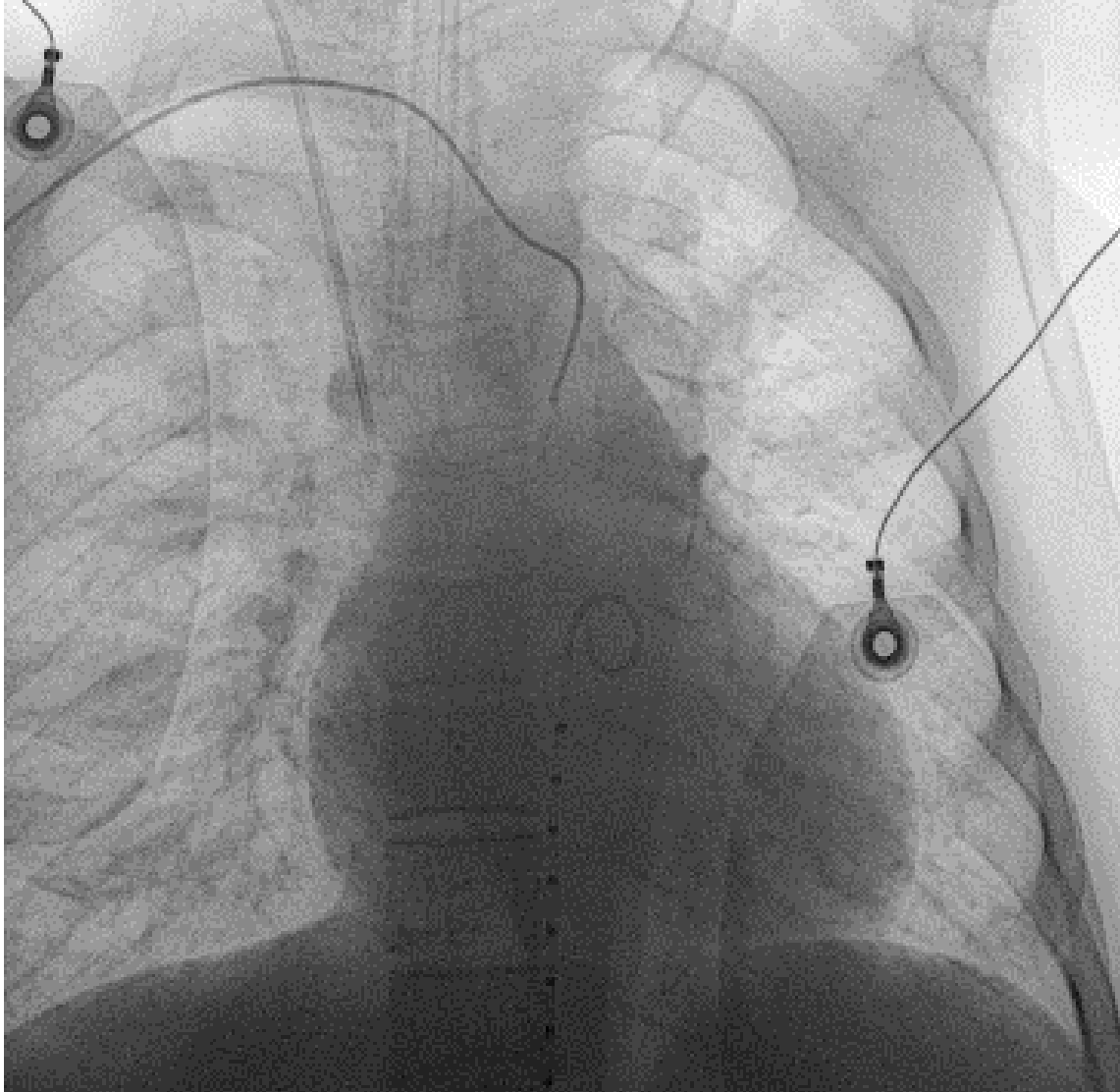
I	B-NR	6. Surgical repair or catheter-based stenting is recommended for adults with hypertension and significant native or recurrent coarctation of the aorta. ^{S4.2.6-1,S4.2.6-2,S4.2.6-8-S4.2.6-12}
I	C-EO	7. GDMT is recommended for treatment of hypertension in patients with coarctation of the aorta. ^{S4.2.6-13}
IIb	B-NR	8. Balloon angioplasty for adults with native and recurrent coarctation of the aorta may be considered if stent placement is not feasible and surgical intervention is not an option. ^{S4.2.6-14}

CoA ESC Recommendations

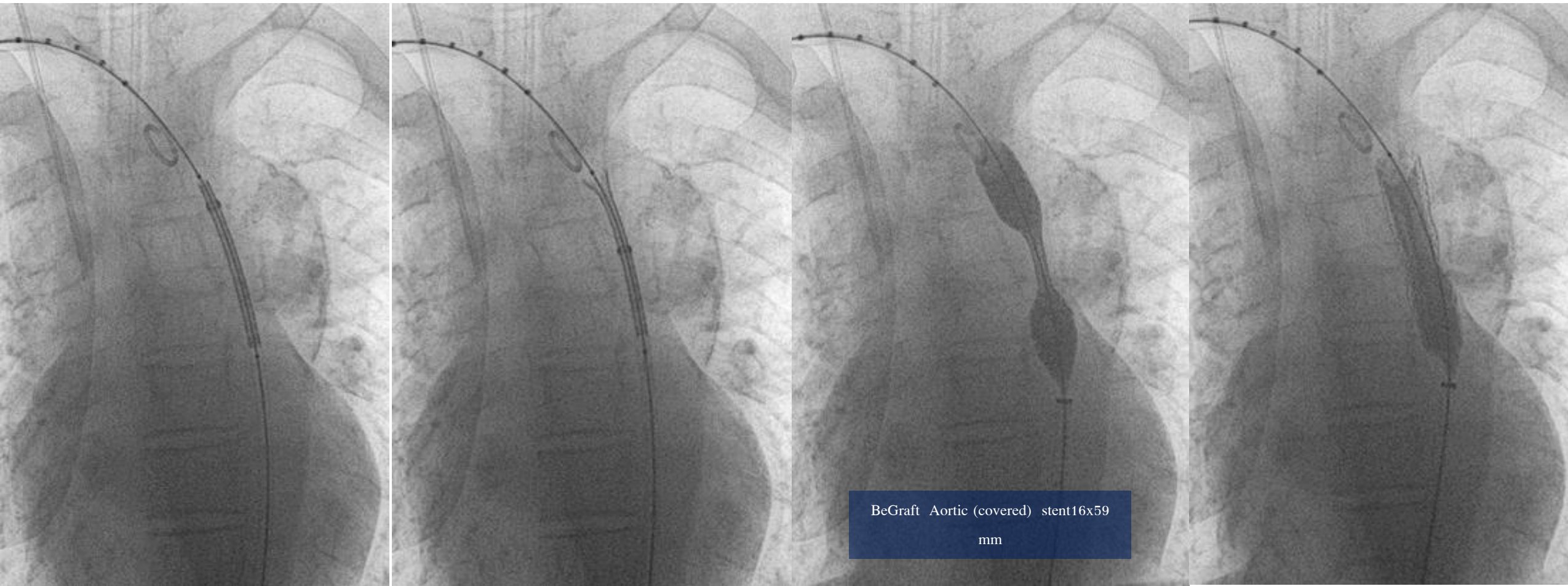
Recommendations	Class ^a	Level ^b
Repair of coarctation or re-coarctation (surgically or catheter based) is indicated in hypertensive patients ^c with an increased non-invasive gradient between upper and lower limbs confirmed with invasive measurement (peak-to-peak ≥ 20 mmHg) with preference for catheter treatment (stenting), when technically feasible.	I	C
Catheter treatment (stenting) should be considered in hypertensive patients ^c with $\geq 50\%$ narrowing relative to the aortic diameter at the diaphragm, even if the invasive peak-to-peak gradient is < 20 mmHg, when technically feasible.	IIa	C
Catheter treatment (stenting) should be considered in normotensive patients ^c with an increased non-invasive gradient confirmed with invasive measurement (peak-to-peak ≥ 20 mmHg), when technically feasible.	IIa	C
Catheter treatment (stenting) may be considered in normotensive patients ^c with $\geq 50\%$ narrowing relative to the aortic diameter at the diaphragm, even if the invasive peak-to-peak gradient is < 20 mmHg, when technically feasible.	IIb	C



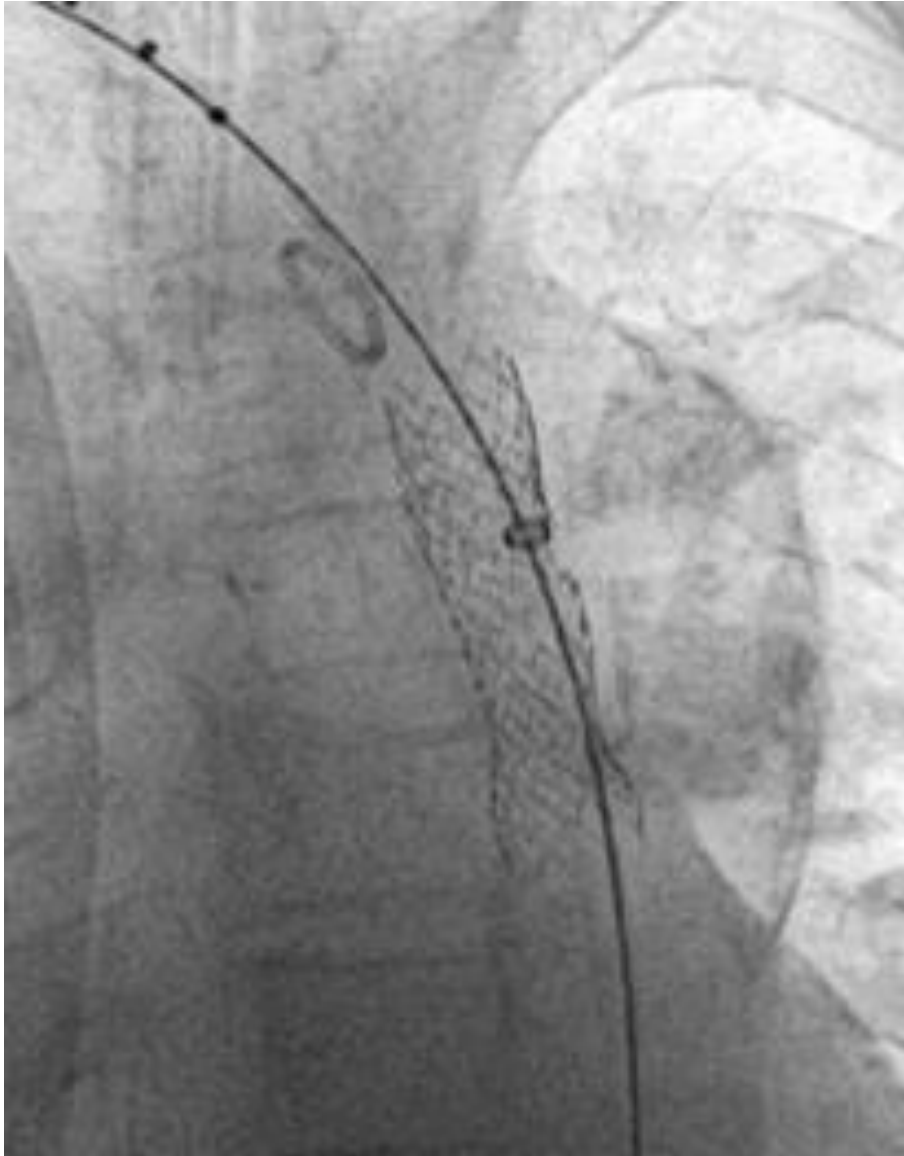
23 years old, s/p aorto-aortic bypass for 11 years, recurrent HT, conduit stenosis with heavy calcification



23 years old, s/p aorto-aortic bypass for 11 years, recurrent HT, conduit stenosis with heavy calcification

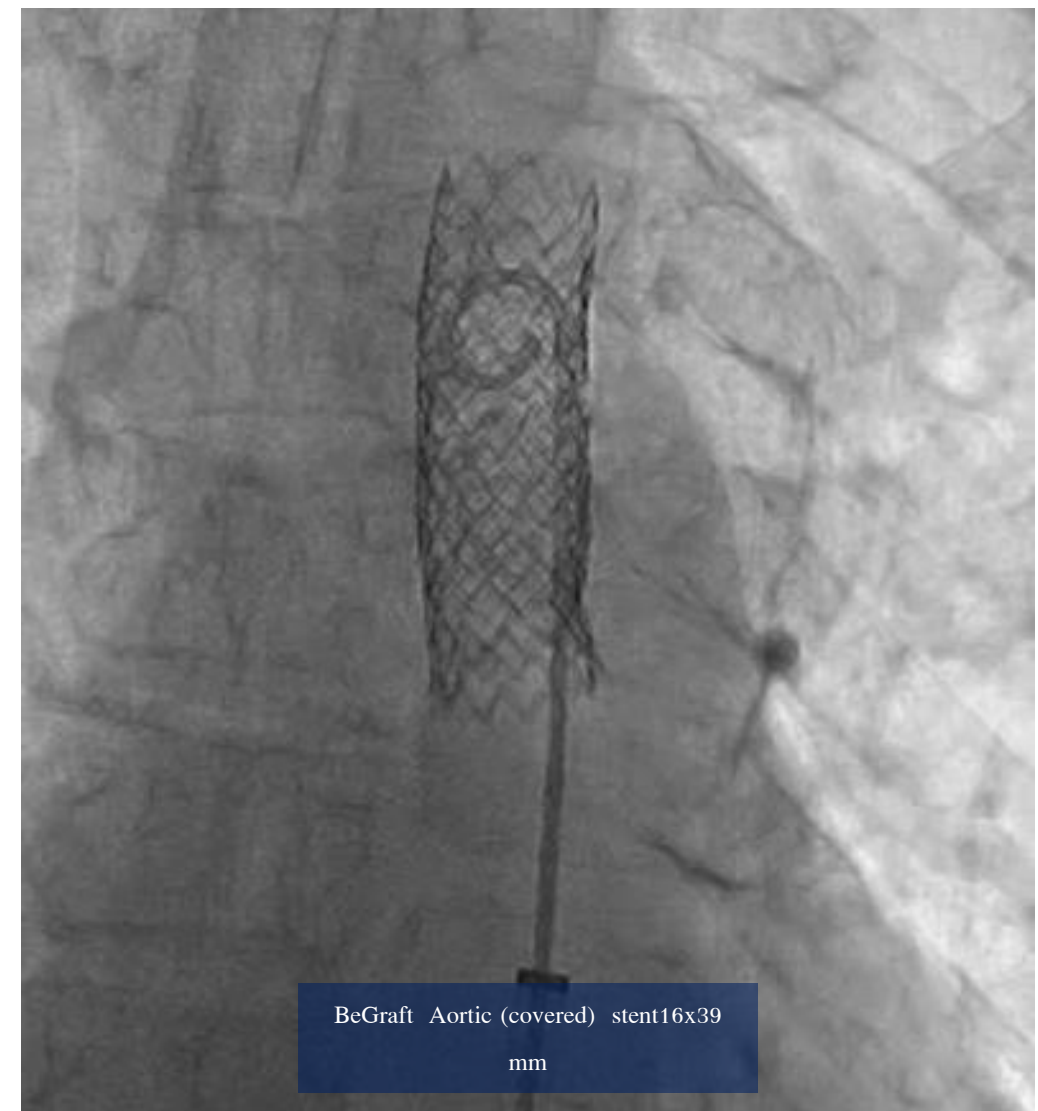
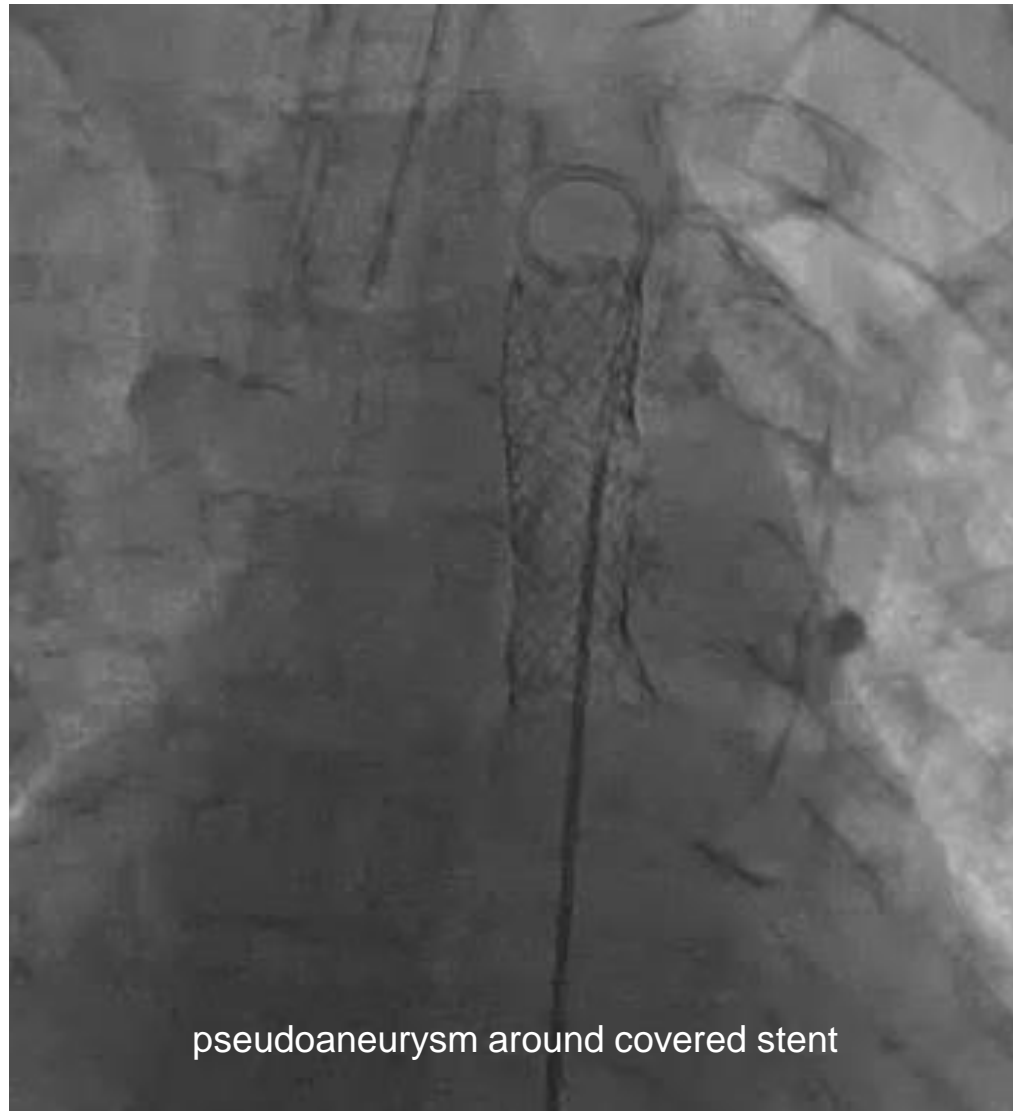


Krit Makonkaewkayoon, Noppon Taksaudom,
Tassaluck Tonghong, and CMU team



23 years old, s/p aorto-aortic bypass for 11 years, recurrent HT, conduit stenosis with heavy calcification





Case 2

- BP 110/70 mmHg, HR 72/min, SpO₂ 98%
- PMI 6ICS, MCL, no heave, no thrill
- Single S₂, gr. 2/6 to and fro murmur at LUSB
- Normal breath sound

- 17-year-old female
- ToF
- 2003: RMBT shunt
- 2007: Closure VSD, TAP, clipping RMBT shunt
- 2017: Hemoptysis

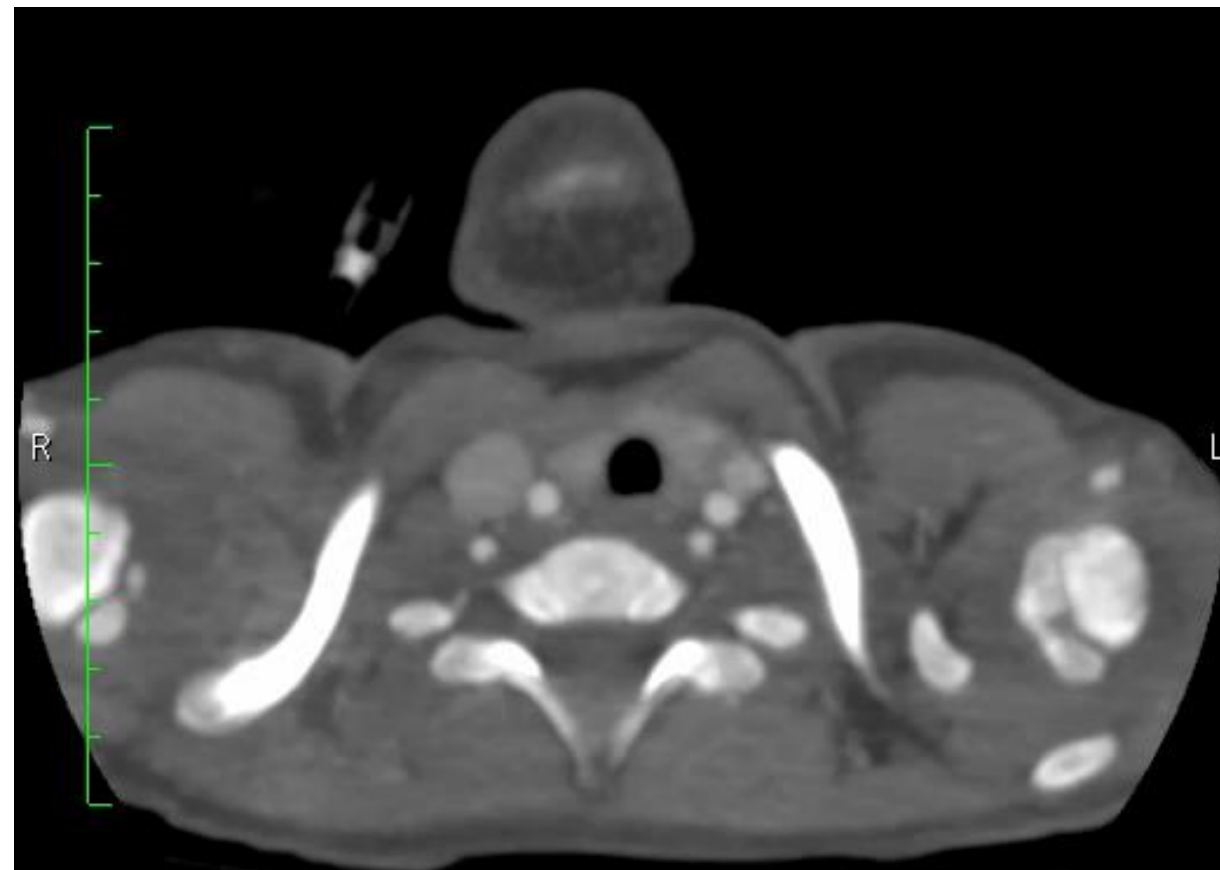


TTE

- No residual VSD
- RVOT aneurysm 40 mm
- PA trunk short and dilated, 30 mm in diameter
- Huge LPA 25 mm, RPA was not well seen
- Free PR , TAPSE 18 mm
- mild to moderate MR



CCT+CMR



Disconnected the distal RPA and lobar branches

Distal RPA 2.9 mm, proximal RPA 13 mm, distance 20 mm

Normal LPA

Forward flow Right : Left lung = 1 : 32

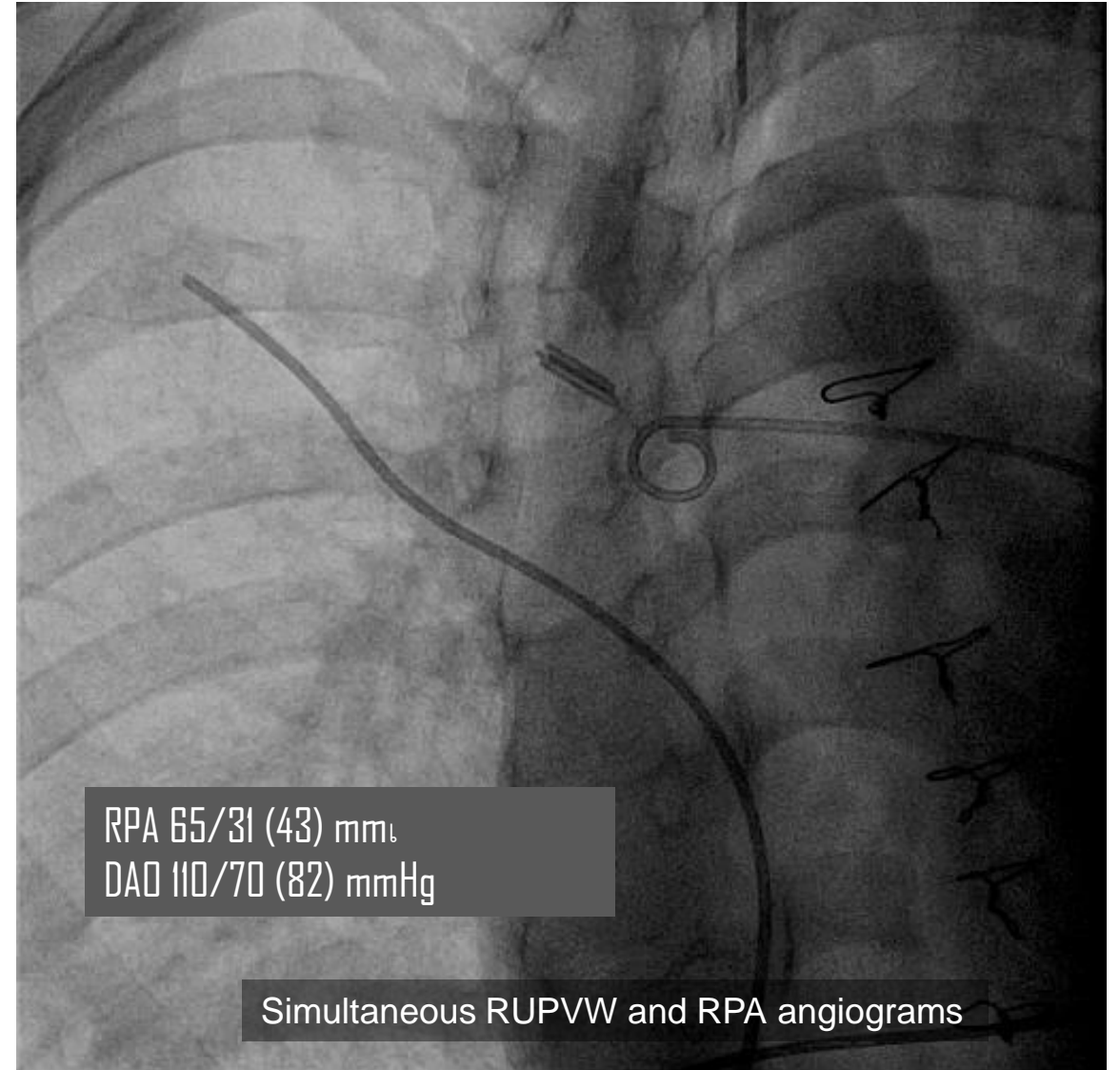
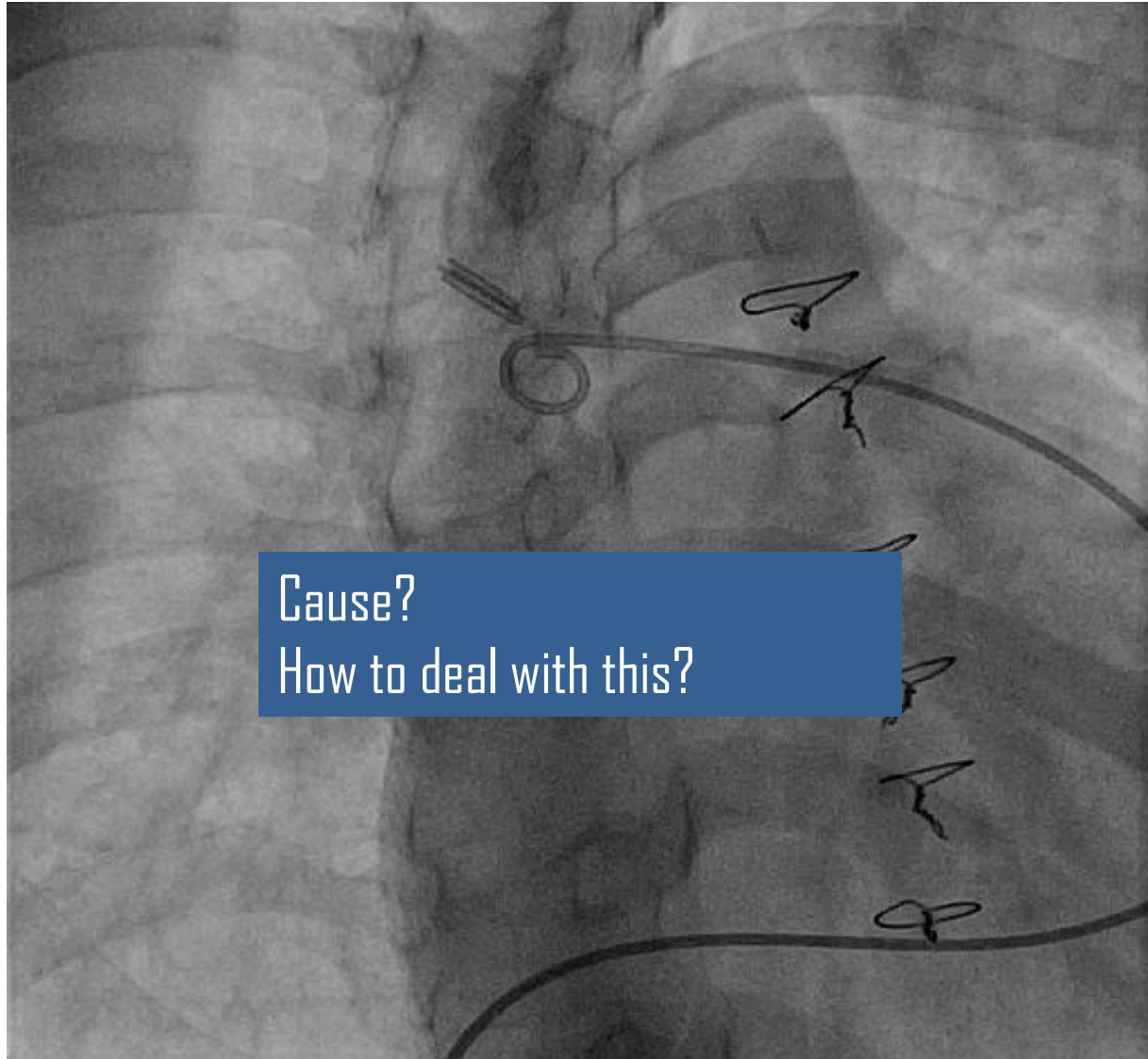
RVOT dilated 34 x 37 mm

RVEDVi 188.4 mL/m²

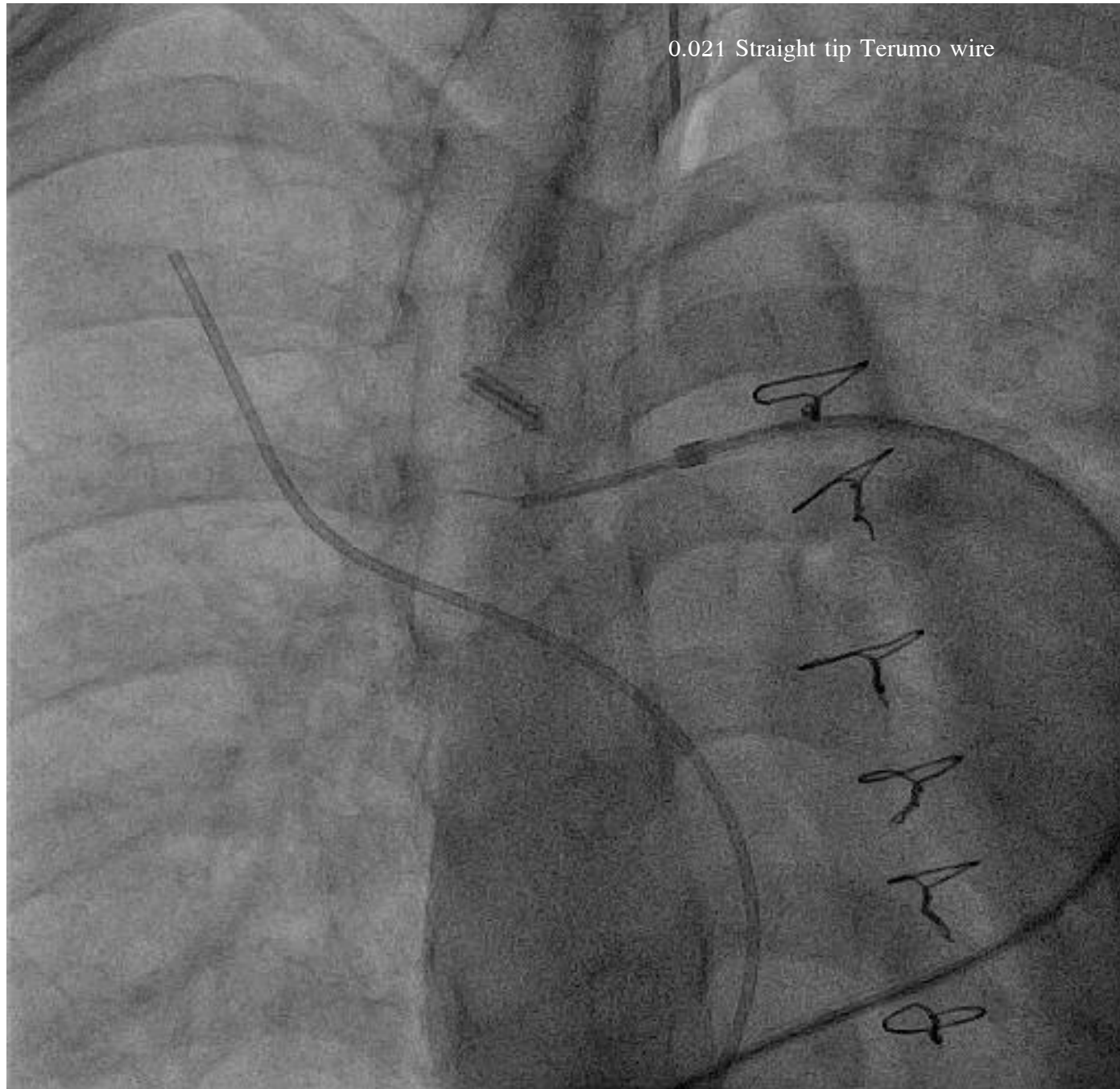
RVEF 62.8 %

Moderate PR, RF 40.1%, mild TR

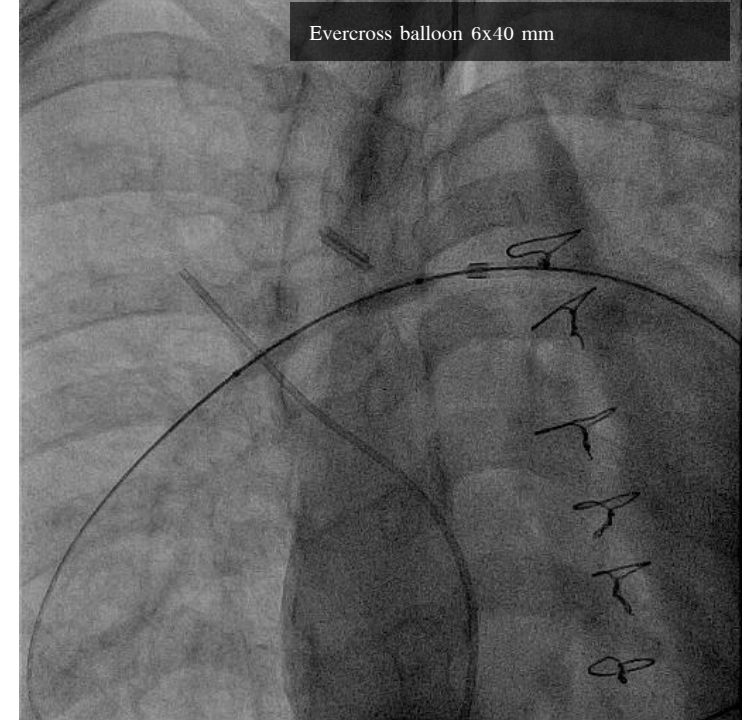
RPA angiogram confirmed interrupted RPA



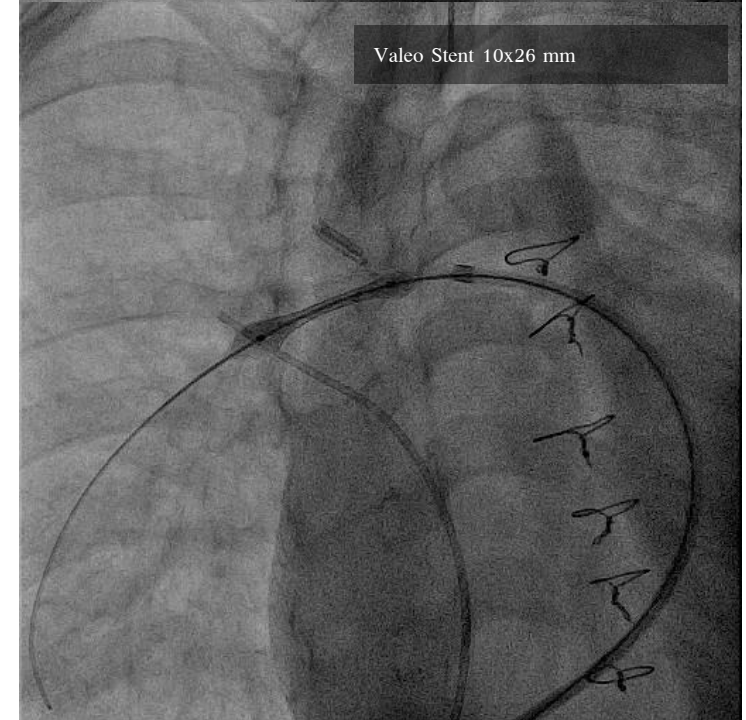
0.021 Straight tip Terumo wire



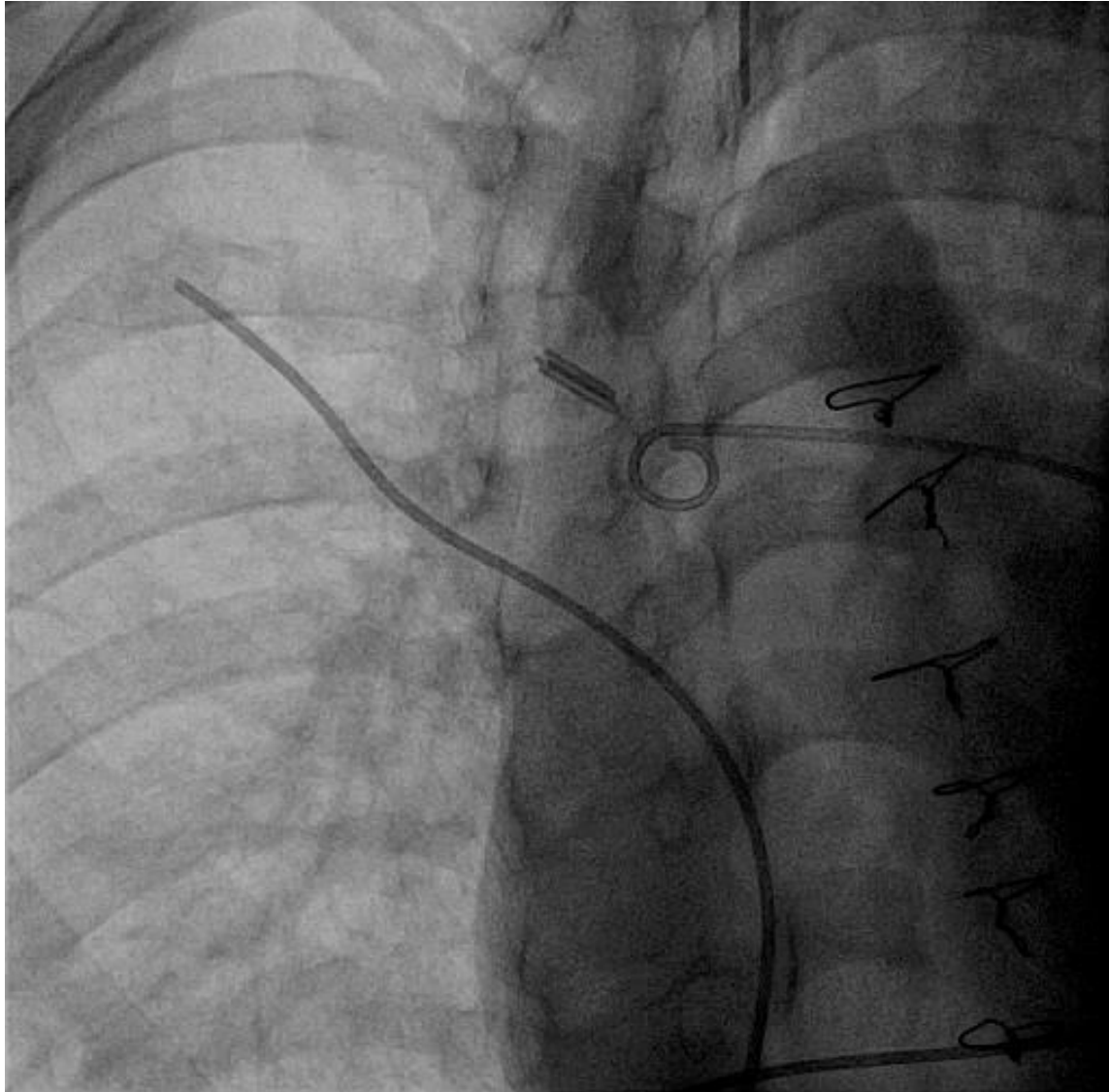
Evercross balloon 6x40 mm



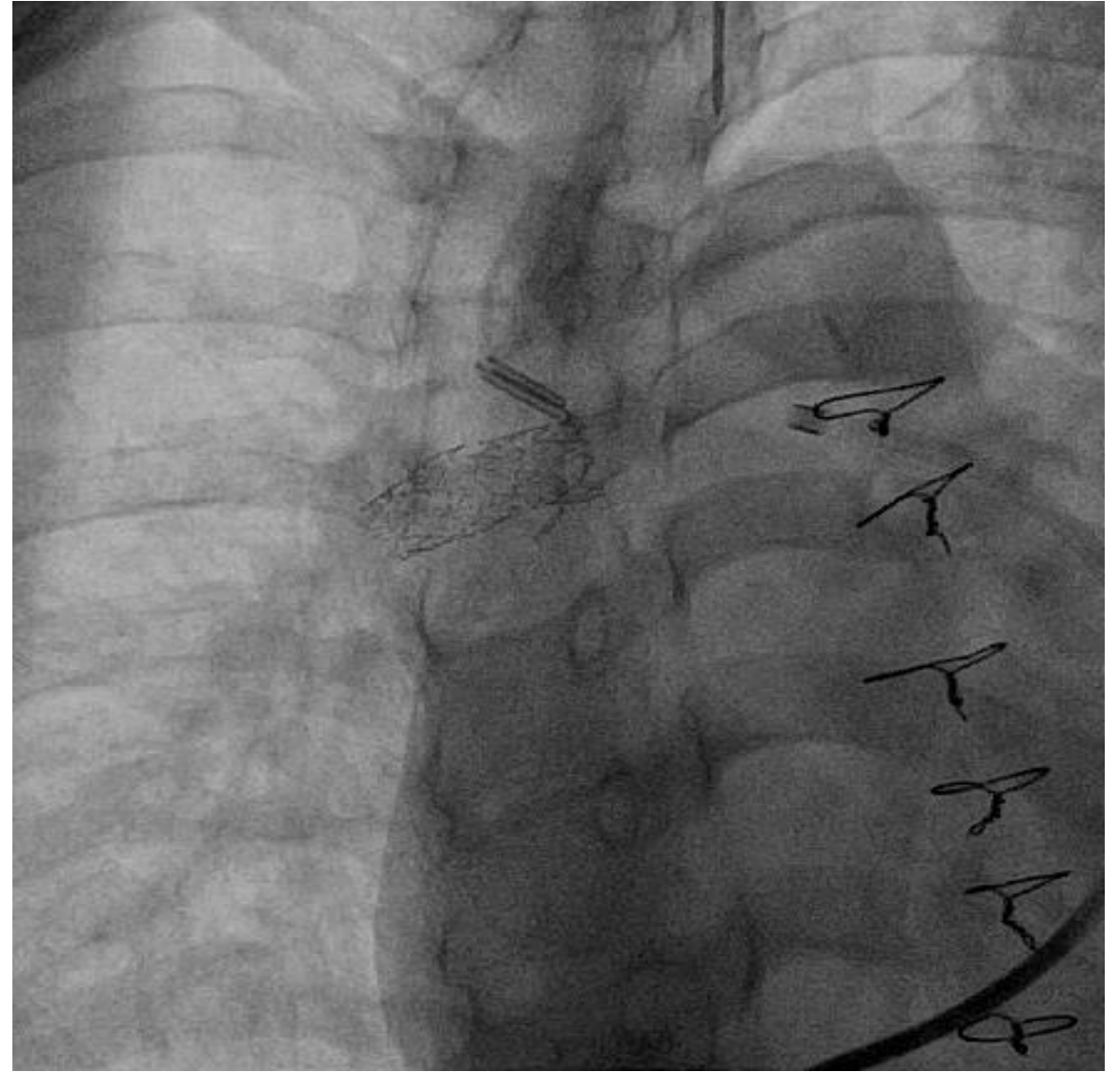
Valeo Stent 10x26 mm



Pre-procedure



Immediate post



CCT follow-up (6 months post)

- Patent newly implanted stent, no intimal proliferation
- Distal RPA 8.8 mm, proximal RPA 17.6 mm.
- LPA 23.1 mm
- Forward flow Right : Left lung = 1 : 3.5
- RVEDVi 163.8 mL/m²
- Normal RV systolic function, RVEF 56 %
- Severe PR, regurgitation fraction 40.3 %, mild TR



Case 3

23-year-old male: progressive dyspnea ~ 6 months

Single ventricle physiology

- Unbalanced ventricle, large inlet-outlet VSD, straddling TV, windsock MV
- DORV, d-TGA
- Severe infundibular-valvular PS

2003 (7 yrs old): **Bidirectional Glenn operation (BDG)**, leaving antegrade PS

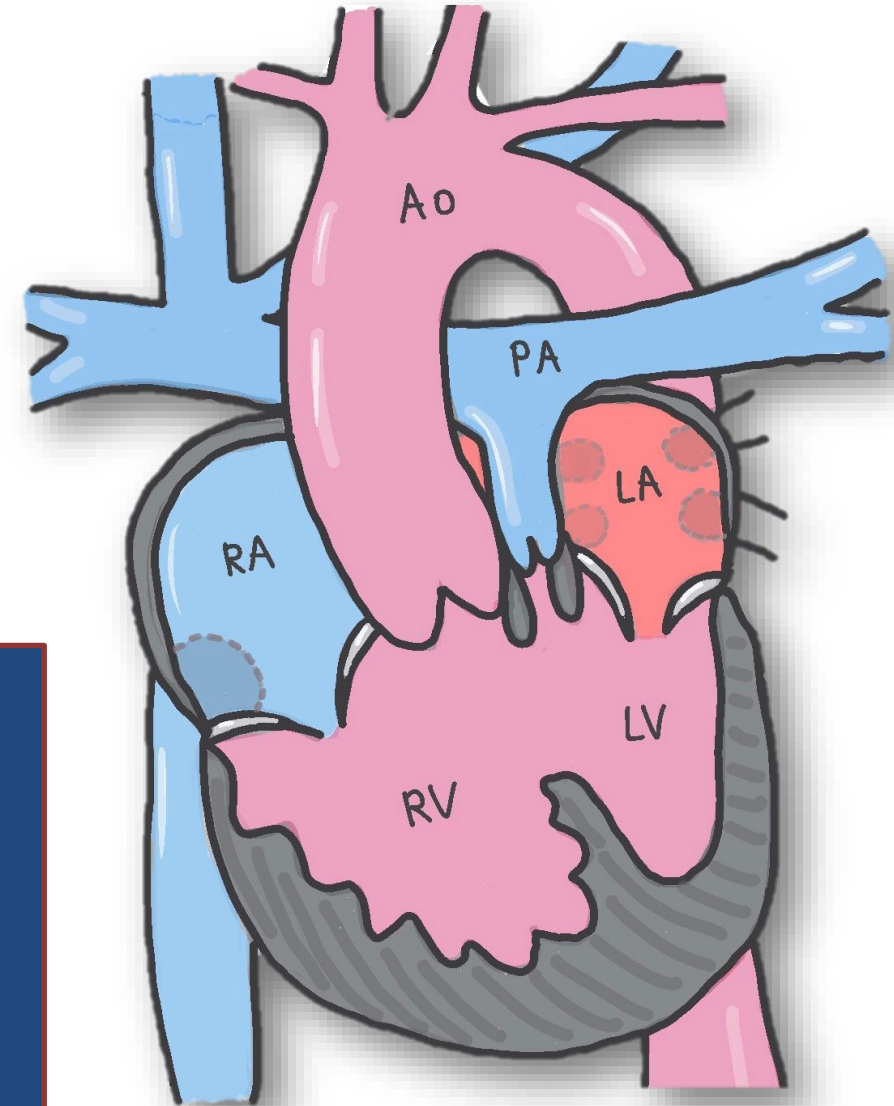
- CVP 20-22 mmHg
- LAP 10-12 mmHg
- O₂sat 85% at FiO₂ 0.6
- Fontan index 3.9

(Fontan index = $\text{PVR} + \text{LVEDP} / \text{Qp} + \text{Qs}$)

2007 (11 yrs old): **cardiac cath**

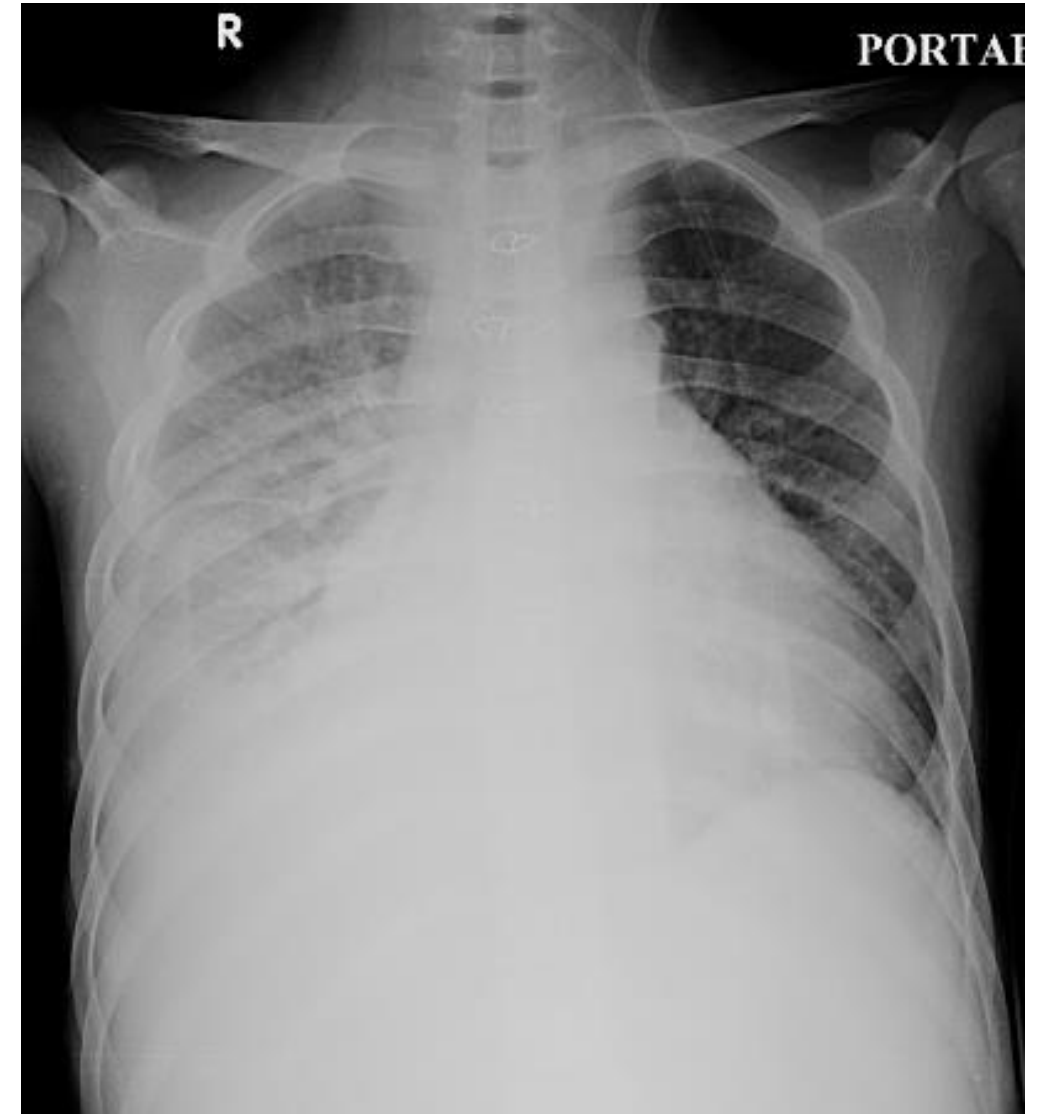
- RPA 25/20 (23) mmHg
- RA sat 57.3%
- Ao sat 70.6%
- Qp:Qs 0.32, Rp:Rs 0.27

Plan: Conservative treatment



2014 (17 yrs old): Clinical right sided HF

- Mild puffy eyelids
- SpO₂ 70%
- PSM gr 3/6 at LMPSB
- SEM gr 3/6 at LUPSB
- Hepatomegaly
- Pitting edema 2+





PR 55Hz
Image 1788
20cm

2D
76%
C 50
P Low
HRes

G
P 1.9 (R) 3.8

120 BPM

M3 PR 14Hz
Image 1715
18cm

2D
78%
C 50
P Low
HRes

CF
61%
2.5MHz
WF High
Med

G
P 1.9 (R) 3.8

JPEG

120 bpm

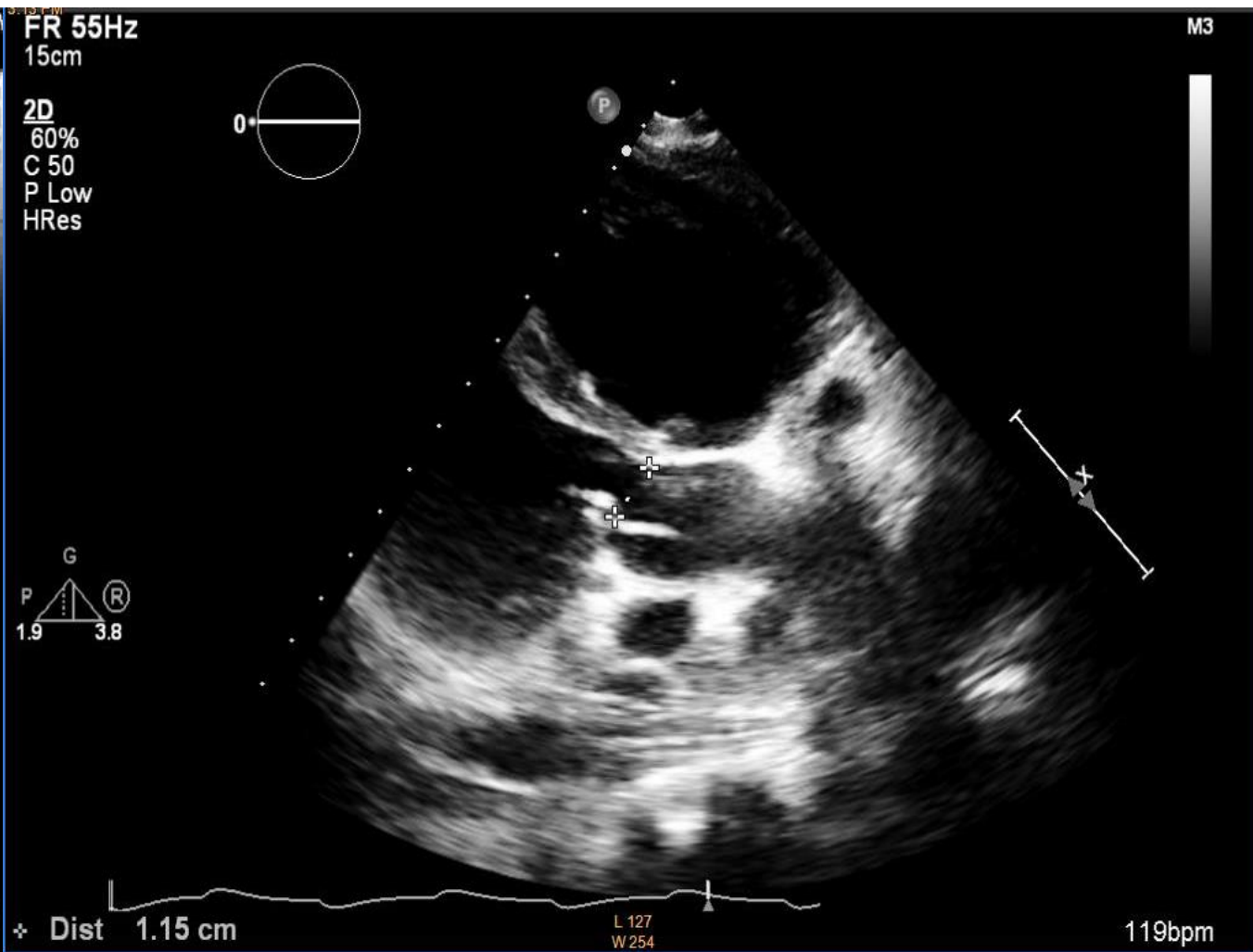
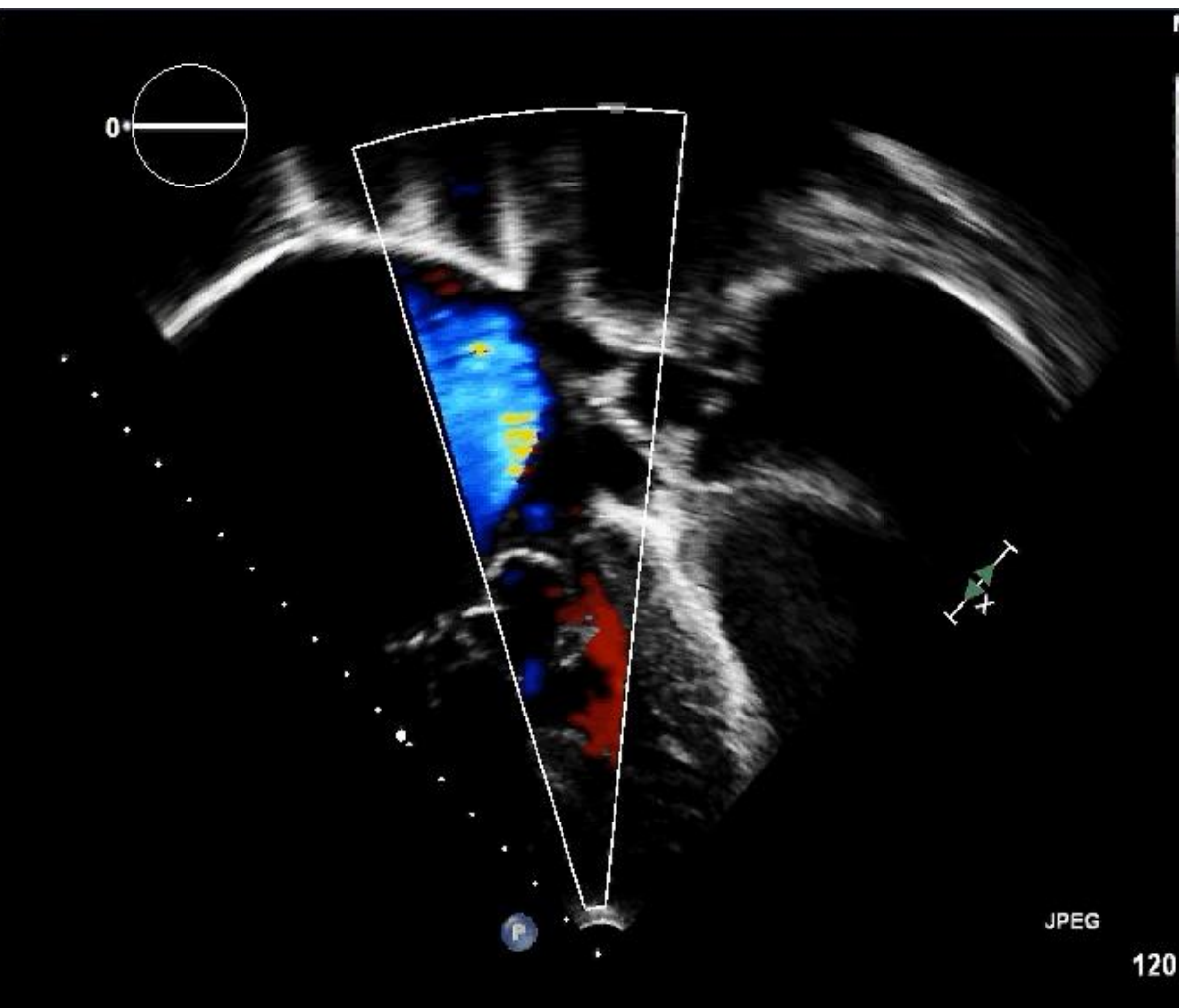
120 BPM

M3 M4
+58.8

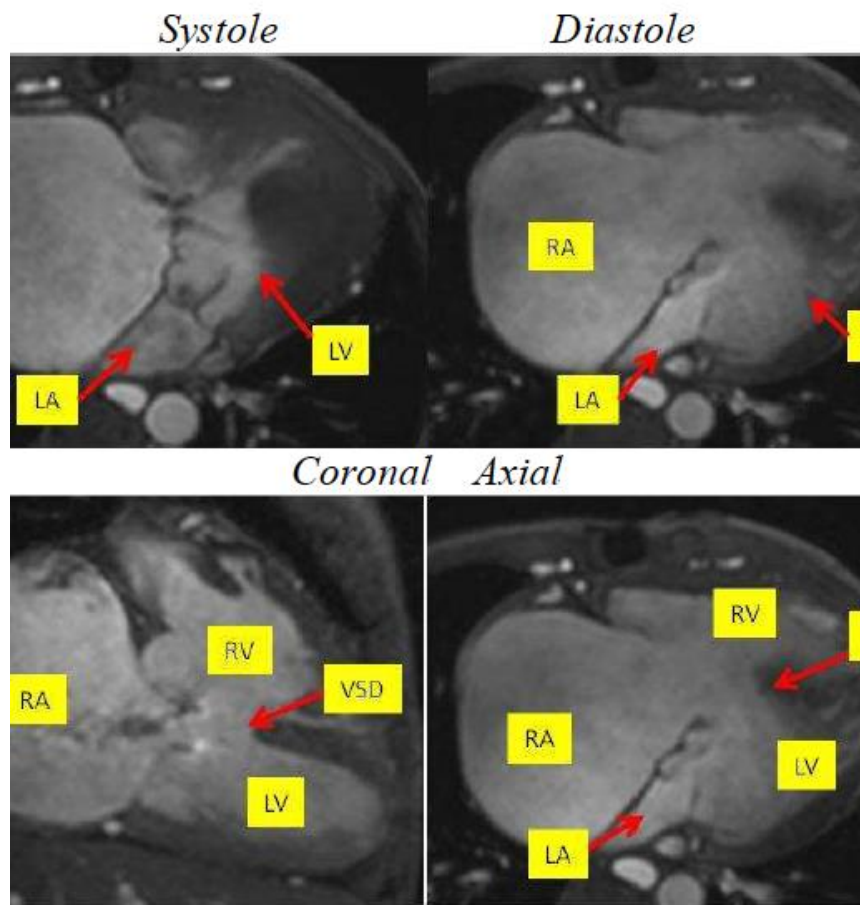
-58.8
cm/s

JPEG

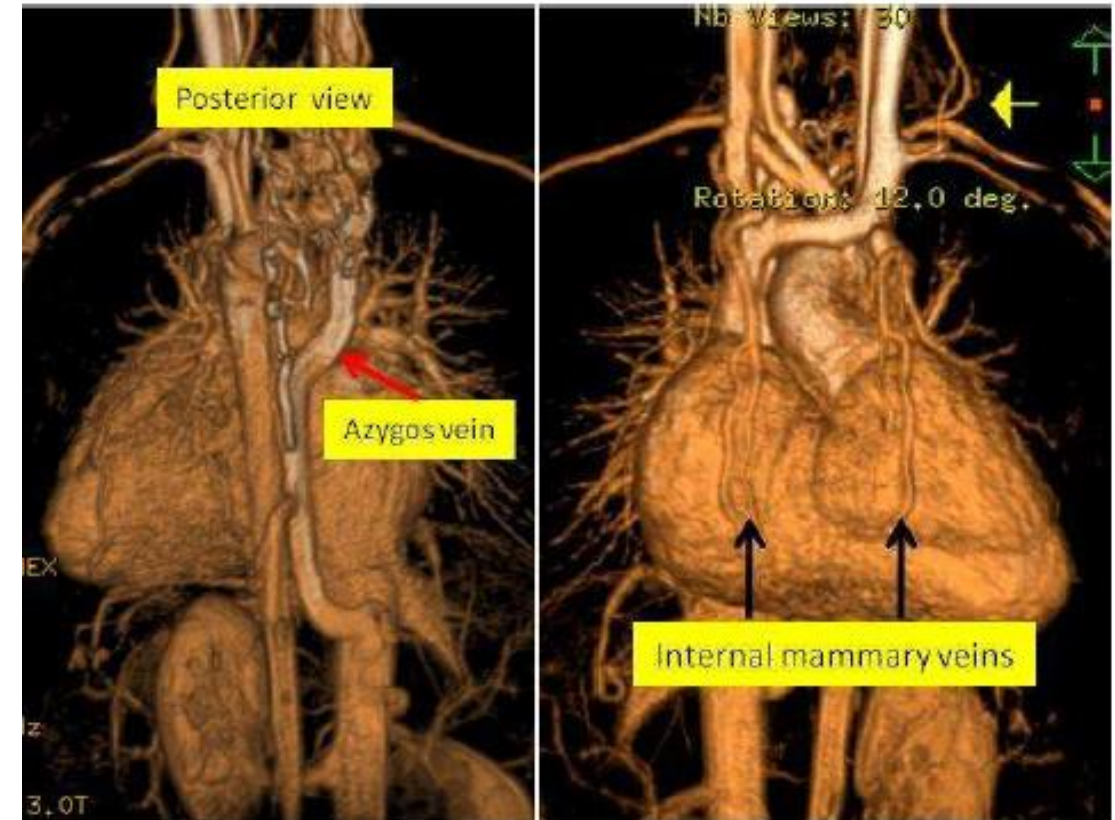
120 bpm



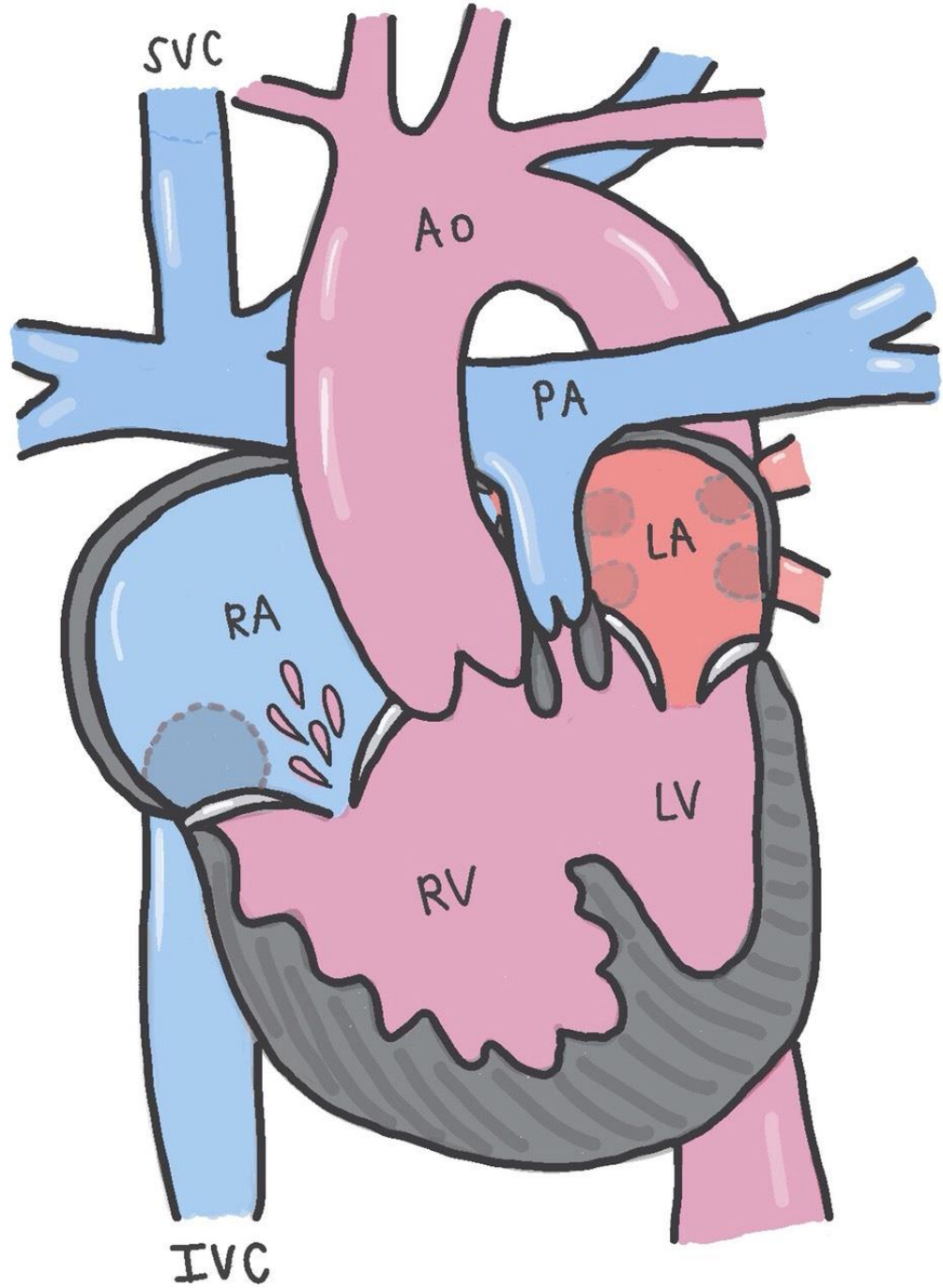
CMR 2014



- Supero-inferior ventricle
- Large inlet VSD 4 cm
- Severe RA dilatation
- Biventricular dilatation
- LVEF 54.6%
- Mild impaired RV systolic function
- Severe TR, severe PS



- Patent Glenn shunt
- Flow outward from RPA
- McGoon ratio 1.6, Nakata 294 mm²/m²
- Multiple venous collateral dilatation



- Single ventricle, severe PS, s/p bidirectional Glenn shunt at age 7 years
- Severe TR
- Inadequate atrial communication

At age 17 year (2014)

- Single ventricle, severe PS s/p bidirectional Glenn shunt at severe TR,
- inadequate atrial communication

Operation:

- TV Replacement: Perimount Magna Mitral 29 mm
- Atrial septectomy
- VVIR pacemaker implantation



PERIMOUNT MAGNA MITRAL

s/p TVR: Magna (Bovine pericardial valve) 29 mm, atrial septectomy,
VVIR at age 18 years (2014)

immediate post-op



1 year post-op



Medications: lasix, aldactone, enalapril, lanoxin, warfarin x 6 months, continue ASA

Unbalanced ventricle, large VSD, d-TGA, severe PS

- S/P BDG, leaving antegrade flow at age 7 years (2003)
- S/P TVR: Magna Bovine pericardial valve 29 mm, atrial septectomy, VVIR at age 18 years (2014)

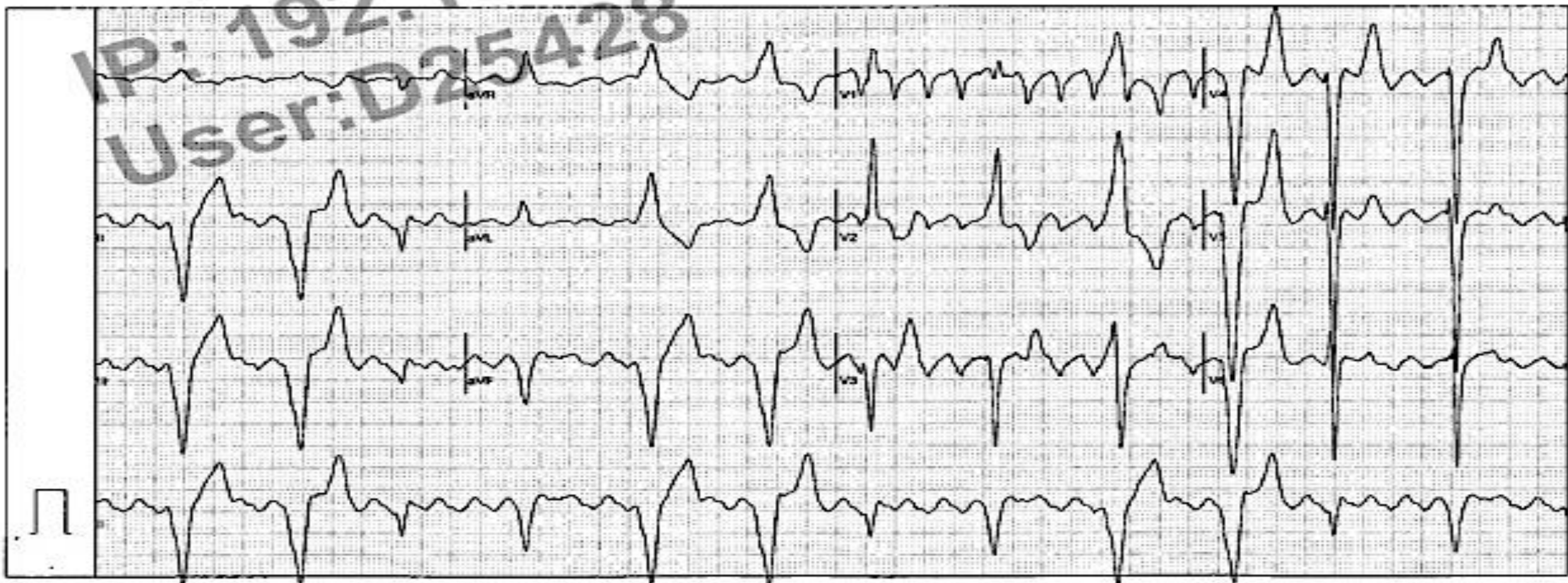
Baseline O_2 sat 75-85%

2018 (age 22 years):

- Atrial flutter → RFA, start warfarin
- Echo: bulging atrial septum, moderate TS, mean PG 8 mmHg, mild MR, good ventricular function
- cardiac catheterization for hemodynamic evaluation
 - Mean PA pressure 26 mmHg, Ao sat 76.1%

QT/QTc: 166/114 ms
 PR: 106 ms
 P: 174 ms
 RR/PP: 374/375 ms
 P/QRS/T: 93/72/0 degrees

nonspecific intraventricular block
 Lateral infarct
 , age undetermined
 Abnormal ECG



ECG Report

4/29/1996 M
10:45:30.8
9:46 AM
Image 1/18
16cm

2D
65%
C 50
P Low
HGen
CF
50%
4522Hz
WF 452Hz
2.5MHz

0 BPM

TIS 0.8 MI 0.8

M3 M4
+69.6

-69.6
cm/s

*** bpm



Adult Echo
4/29/1996 M
10/16/2018
9:44 AM
13Hz
16cm

2D
65%
C 50
P Low
HGen

CF
50%
4522Hz
WF 452Hz
2.5MHz



TISO.8 MI 0.8

M3 M4
+69.6
-69.6
cm/s

0 BPM

*** bpm



Cardiac Catheterization

	Pressure (mmHg)	O ₂ sat (%)		Pressure (mmHg)	O ₂ sat (%)
LSVC	34/18 (25)		LPA	32/20 (24)	
RA	20/12 (15)	69.9	LUPV	19/12 (16)	94
IVC	19/11 (15)		LLPV	21/13 (16)	93.3
RV	82/4 (15)		RUPV	17/12 (14)	95.7
MPA	34/19 (26)	78.8	RLPV	19/11 (15)	95.1
RPA	32/19 (25)	78.9	AO	86/48 (65)	76.1

Hb 17.4%, LVEDP 14 mmHg

Qp:Qs 0.383, Rp/Rs 0.512,

PVR 3.9 WU.m²

Fontan index 3.82

McGoon 1.62, Nakata index 215

Estimated Fontan pressure 40 mmHg

Unbalanced ventricle, large VSD, d. TCA, severe PS

Estimated Fontan Pressure 40 mmHg

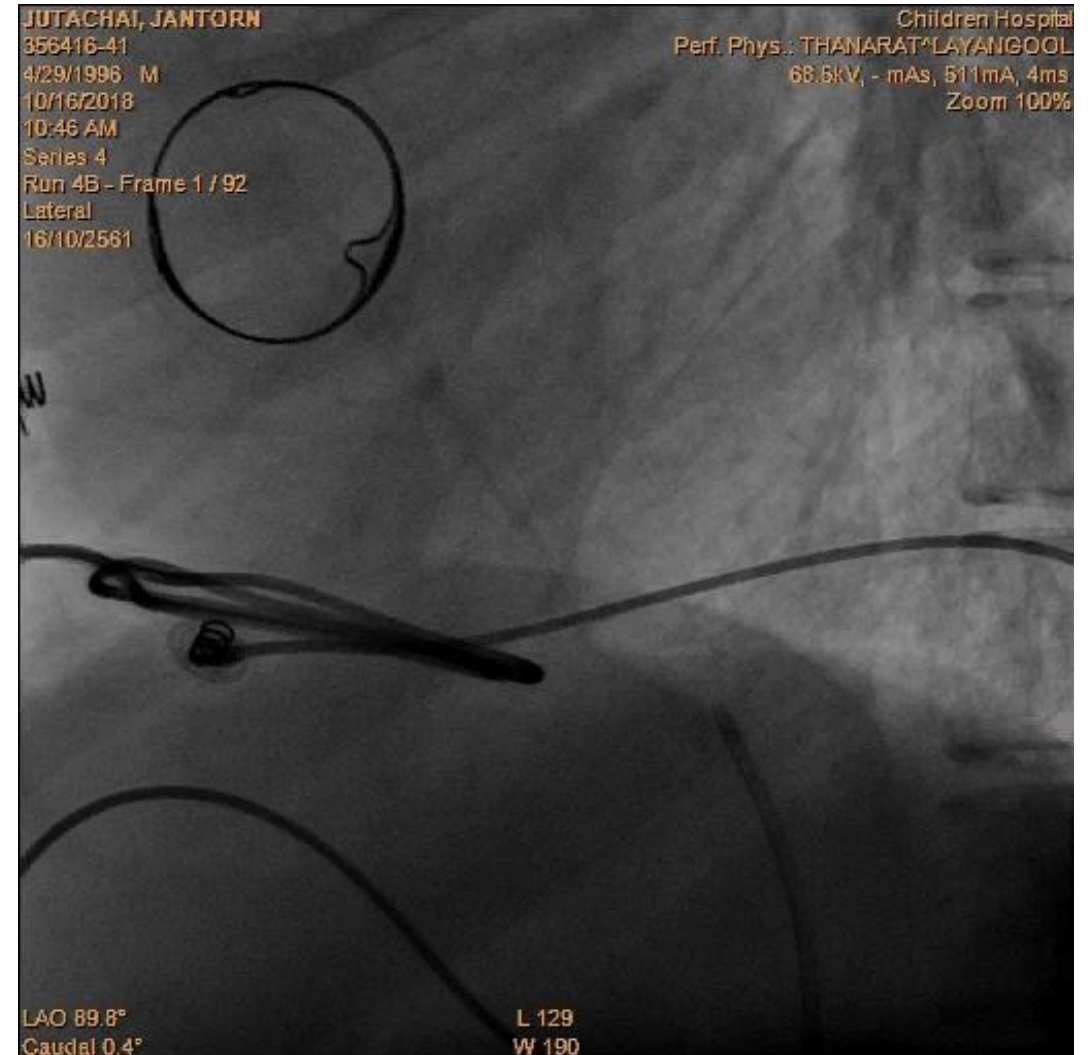
TPG = PAP-LAP = 25-16 = 9 mmHg

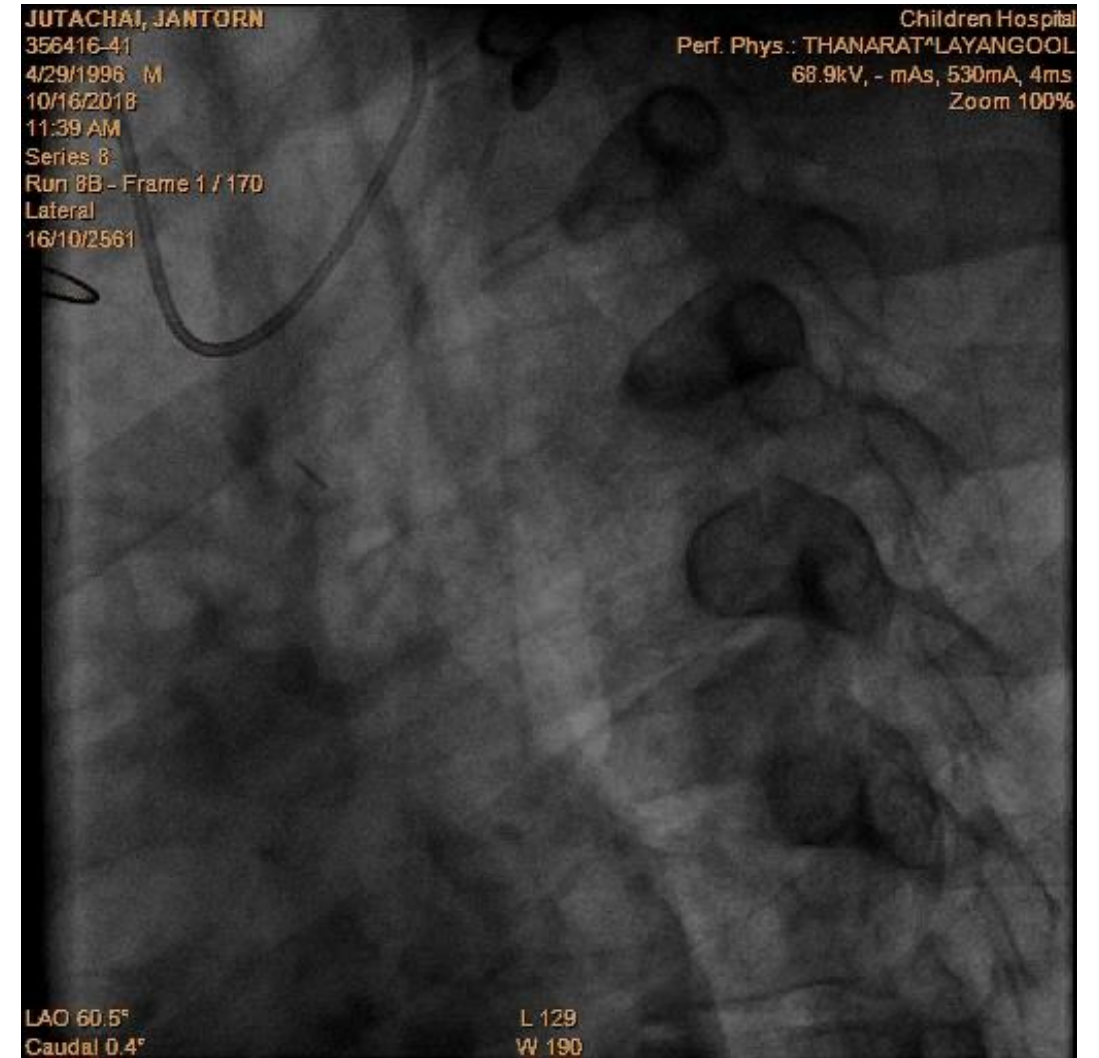
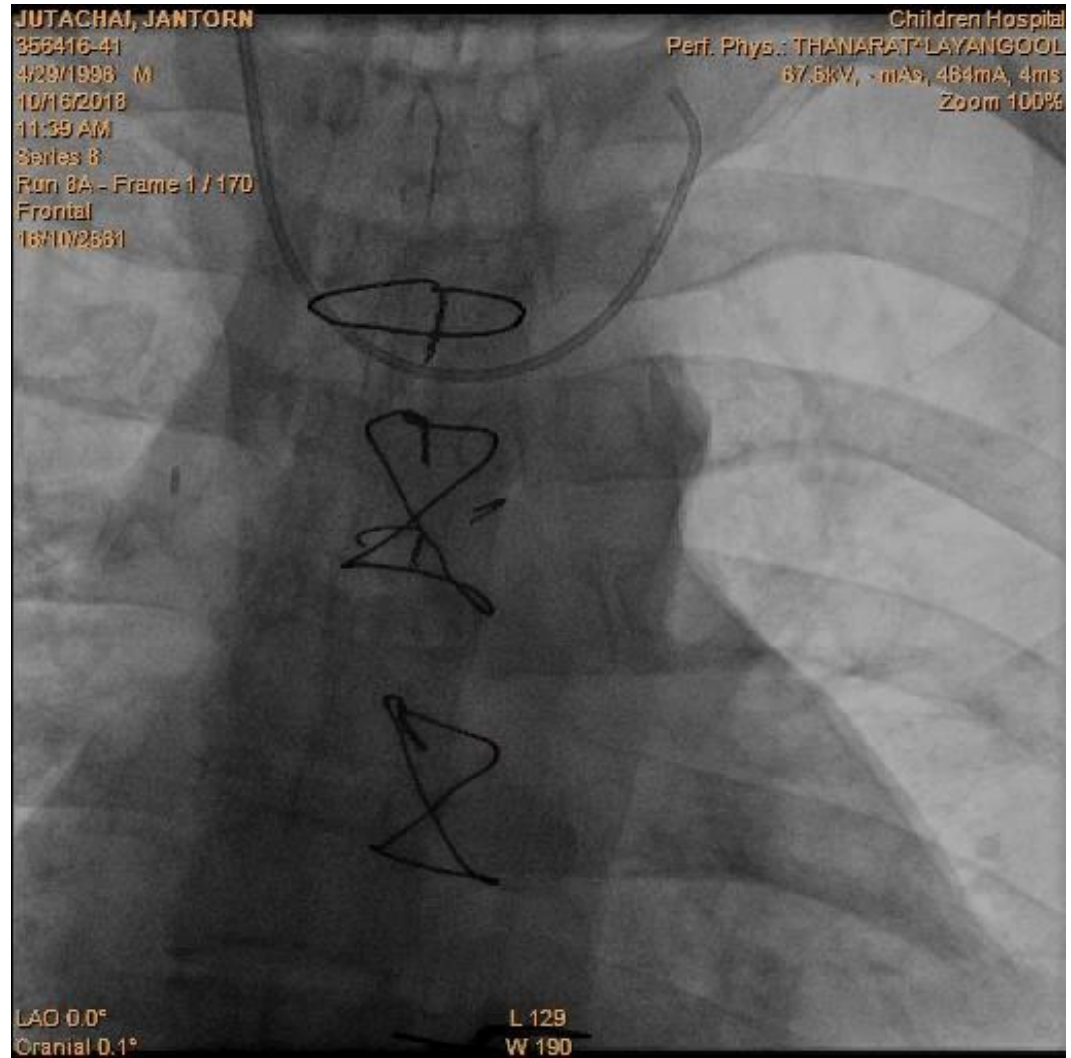
Qp:Qs 0.38, TPG 9 mmHg

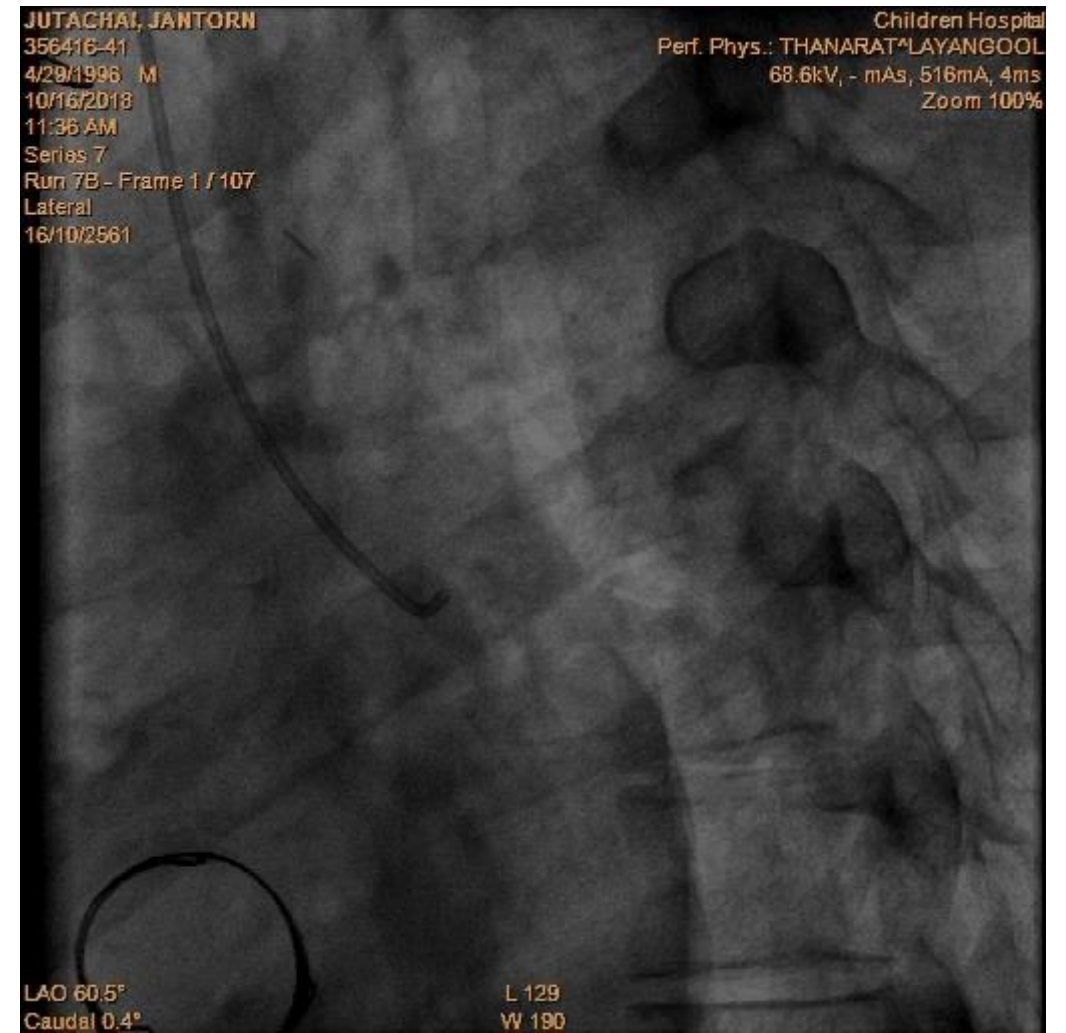
If Qp:Qs = 1 → est.TPG = 24 mmHg

Estimated PAP at Qp:Qs 1 = LAP + est.TPG
= 16 + 24 = 40 mmHg

Angiograms







2020 (age 24 years):

- **Progressive dyspnea**
- **FC III-IV, swelling,**
- **O₂sat 63-70%** (room air)



24-year-old, male

Single ventricle, severe PS

- S/P BDG, leaving antegrade flow at age 7 years (2003)

- S/P TV Replacement: Magna Bovine pericardial valve @29mm, atrial septectomy, VVIR at age 18 years (2014)

- S/P RFA, S/P Pacemaker reimplantation at age 23 years (2019)

Baseline O₂sat 75%

Adult Echo

TIS1.0 MI 0.9

X5-1

17Hz

22cm

2D

85%

C 50

P Low

HGen

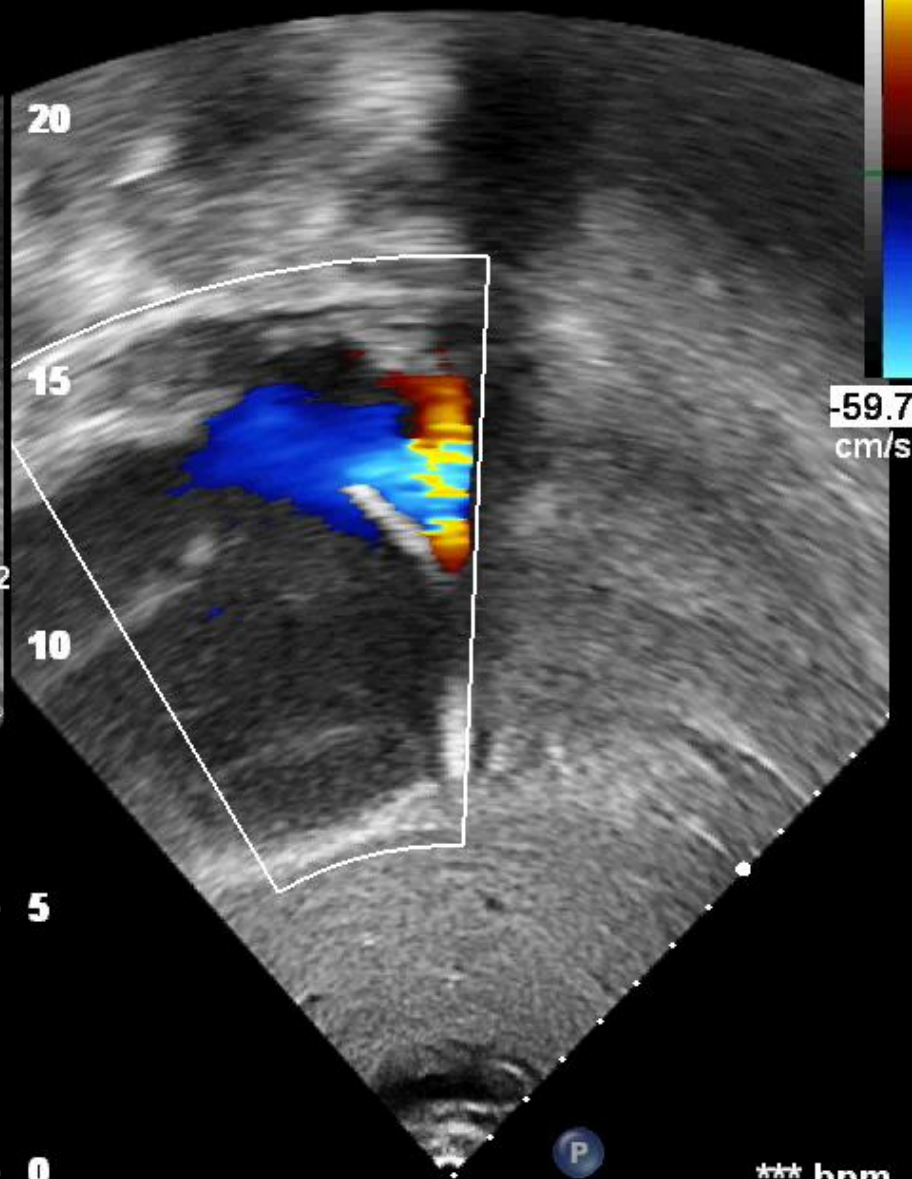
CF

50%

3875Hz

WF 387Hz

2.5MHz



M4
+59.7

-59.7
cm/s

P

*** bpm

8/1/2020 10:28 AM

JPEG

Adult Echo

X5-1

18Hz

19cm

2D

79%

C 50

P Low

HGen

CF

50%

4000Hz

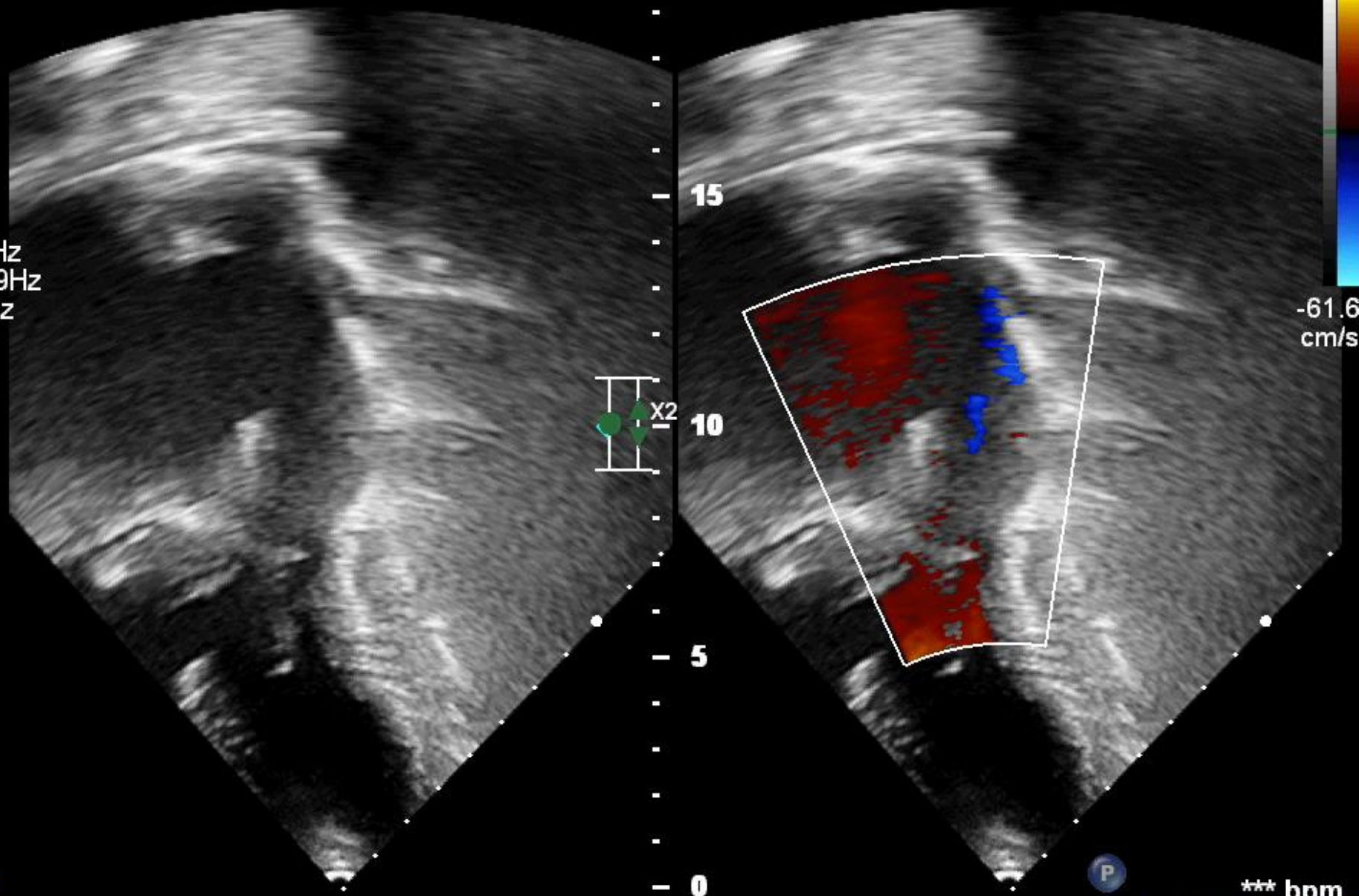
WF 399Hz

2.5MHz

TIS1.0 MI 0.9

M4
+61.6

-61.6
cm/s



Adult Echo

X5-1

22Hz

19cm

2D

79%

C 50

P Low

HGen

CF

50%

4000Hz

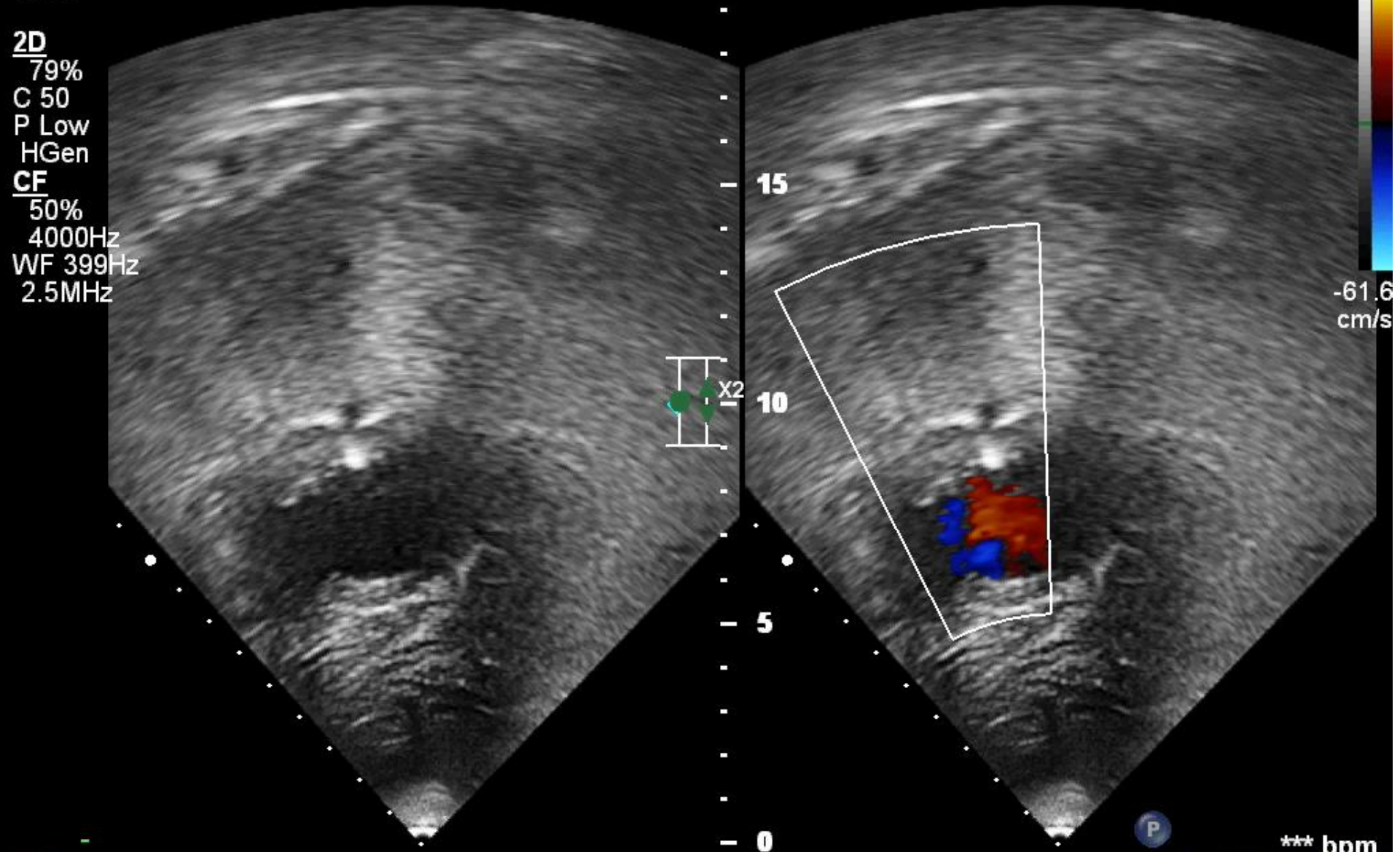
WF 399Hz

2.5MHz

TISO.9 MI 0.9

M4
+61.6

-61.6
cm/s



JPEG

*** bpm

8/1/2020 10:32 AM

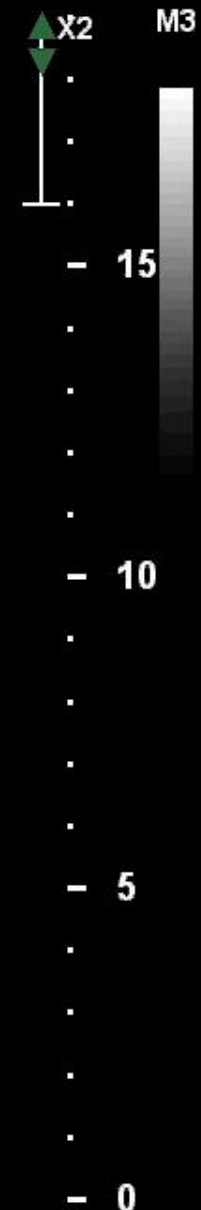
Adult Echo

X5-1
50Hz
19cm



TIS0.4 MI 1.2

2D
68%
C 50
P Low
HGen



G
P R
1.6 3.2

P

JPEG

*** bpm
8/1/2020 10:41 AM

ECHO at age 24 years

- s/p Rt. Bidirectional Glenn shunt, leaving antegrade flow
- Dilated IVC and hepatic vein
- Marked dilated RA, predominant right to left shunt
- Severe tissue valve stenosis at TV position, mean PG 10 mmHg
- Windsock MV, with moderate MR, mean MV inflow PG 5 mmHg
- Severe subvalvular and valvular PS, PG 120 mmHg
- No AR
- Patent Glenn, Lt arch

Age 7 yr: BDG, leaving PS

Age 18 yr: TV replacement, atrial septectomy, VVIR

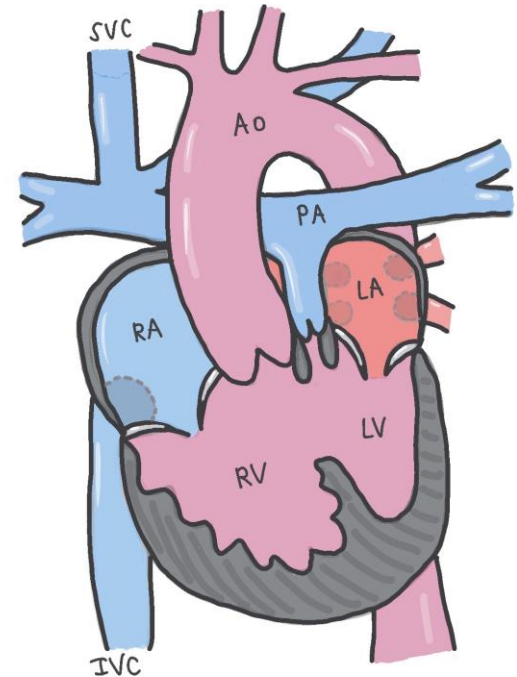
Age 22 yr Atrial flutter S/PRFA

Age 24 yr: Severe TS, moderate MR, moderate MS



WHAT'S NEXT???

How to manage this patient?



Special considerations and recommendations for intervention in univentricular heart (1)

Recommendations	Class	Level
It is recommended that adults with unoperated or palliated UVHs undergo careful evaluation in specialized centres, including multimodality imaging as well as invasive work-up to decide whether they may benefit from surgical or interventional procedures.	I	C
Only well-selected symptomatic cyanotic patients, after careful evaluation [low pulmonary vascular resistances, adequate function of the AV valve(s), preserved ventricular function], should be considered candidates for a Fontan circulation.	Ila	C
Patients with increased pulmonary blood flow – unlikely at adult age – should be considered for PA banding or tightening of a previously placed band.	Ila	C

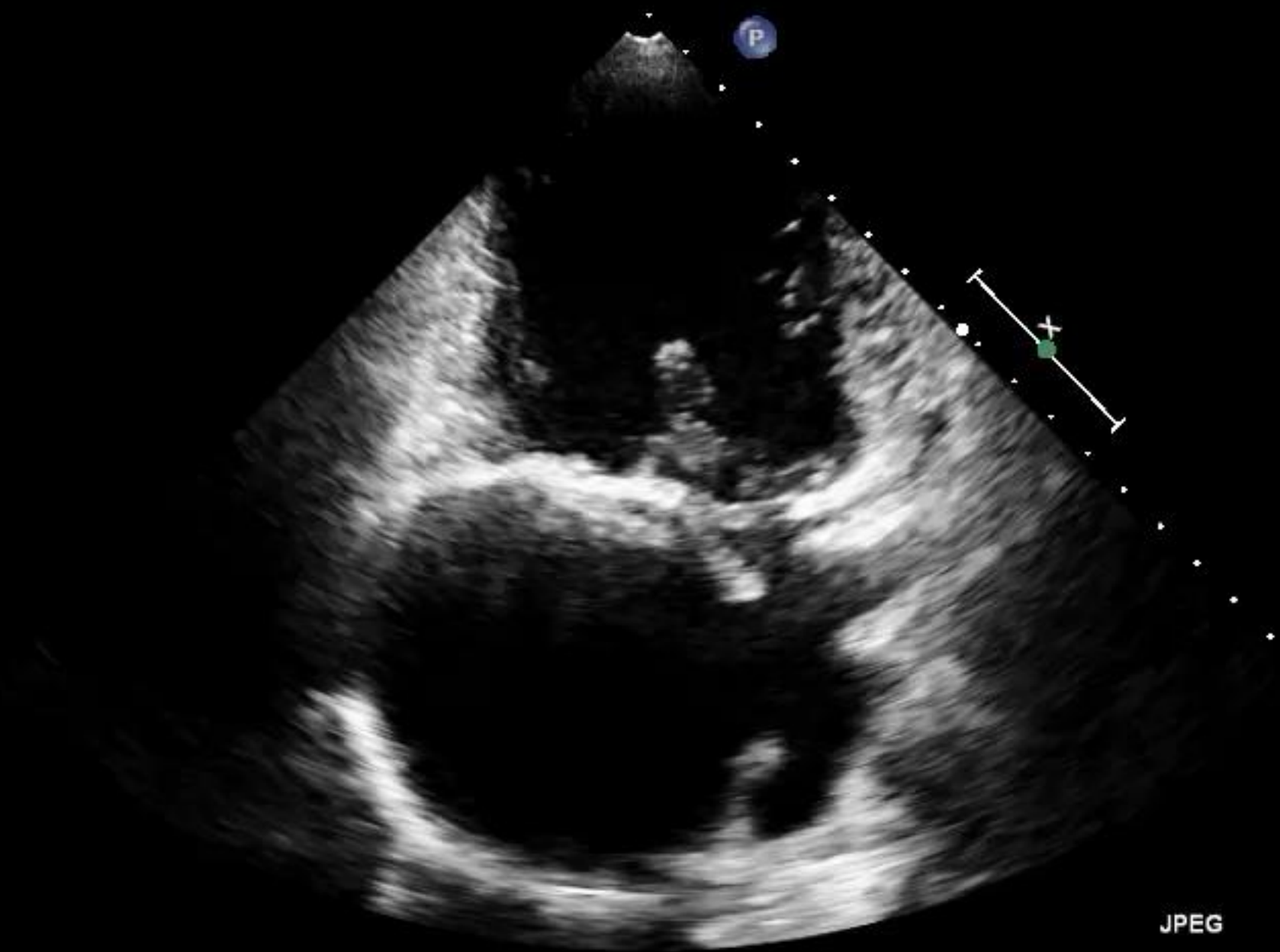
©ESC

- Transcatheter tricuspid valve-in-valve
- +/- balloon pulmonic valve

FR 45Hz
18cm

2D
65%
C 50
P Low
HGen

M3



JPEG

64 bpm

FR 16Hz
17cm

2D
58%
C 50
P Low
HGen

CF
66%
2.5MHz
WF High
Med



JPEG

61 bpm

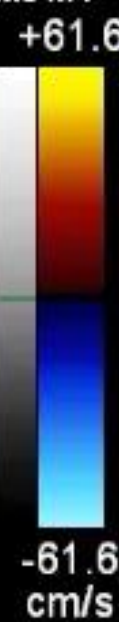
PR 3.12
17cm

2D

59%
C 50
P Low
HGen

CF

66%
2.5MHz
WF High
Med



JPEG

59 bpm

FR 47Hz
17cm

2D
62%
C 50
P Low
HGen

90 DEGREE CCW



JPEG

67 bpm



FR 8Hz
16cm

2D

62%

C 50

P Low

HGen

CF

66%

2.5MHz

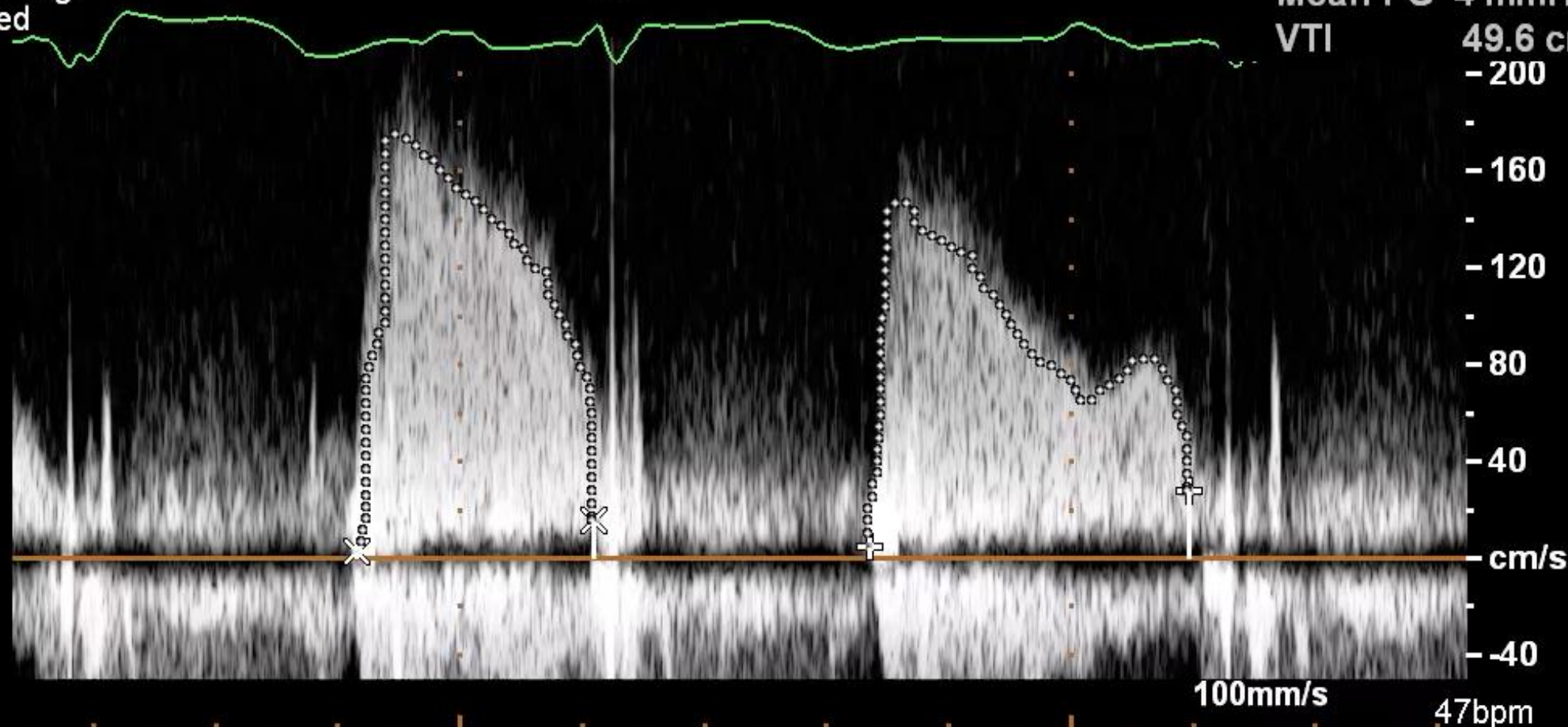
WF High

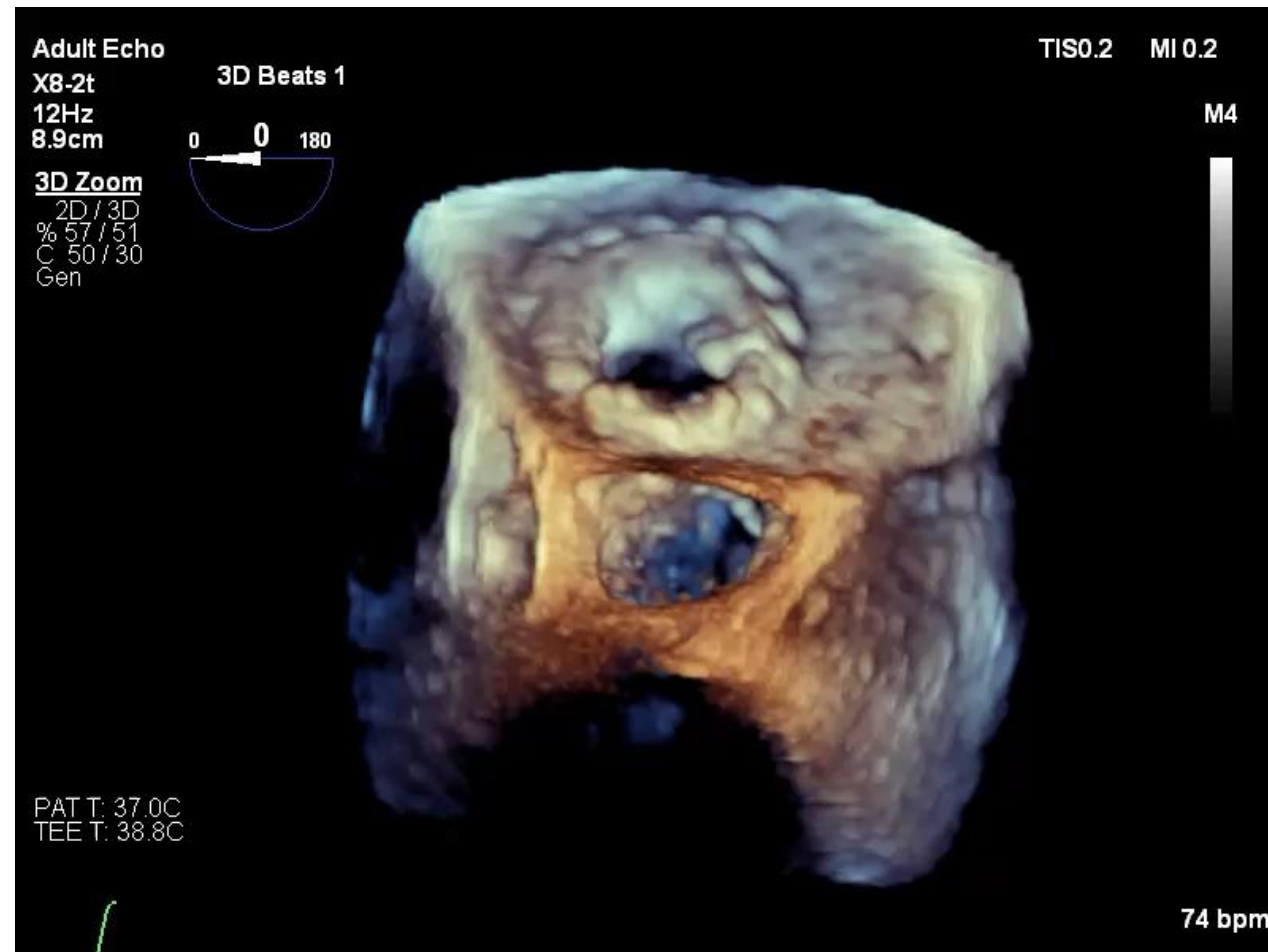
Med

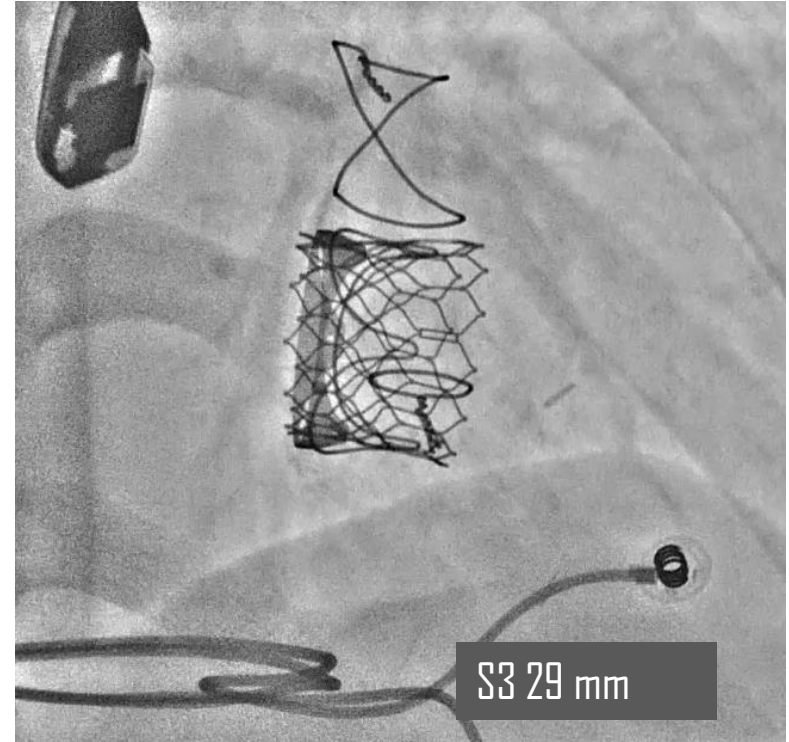
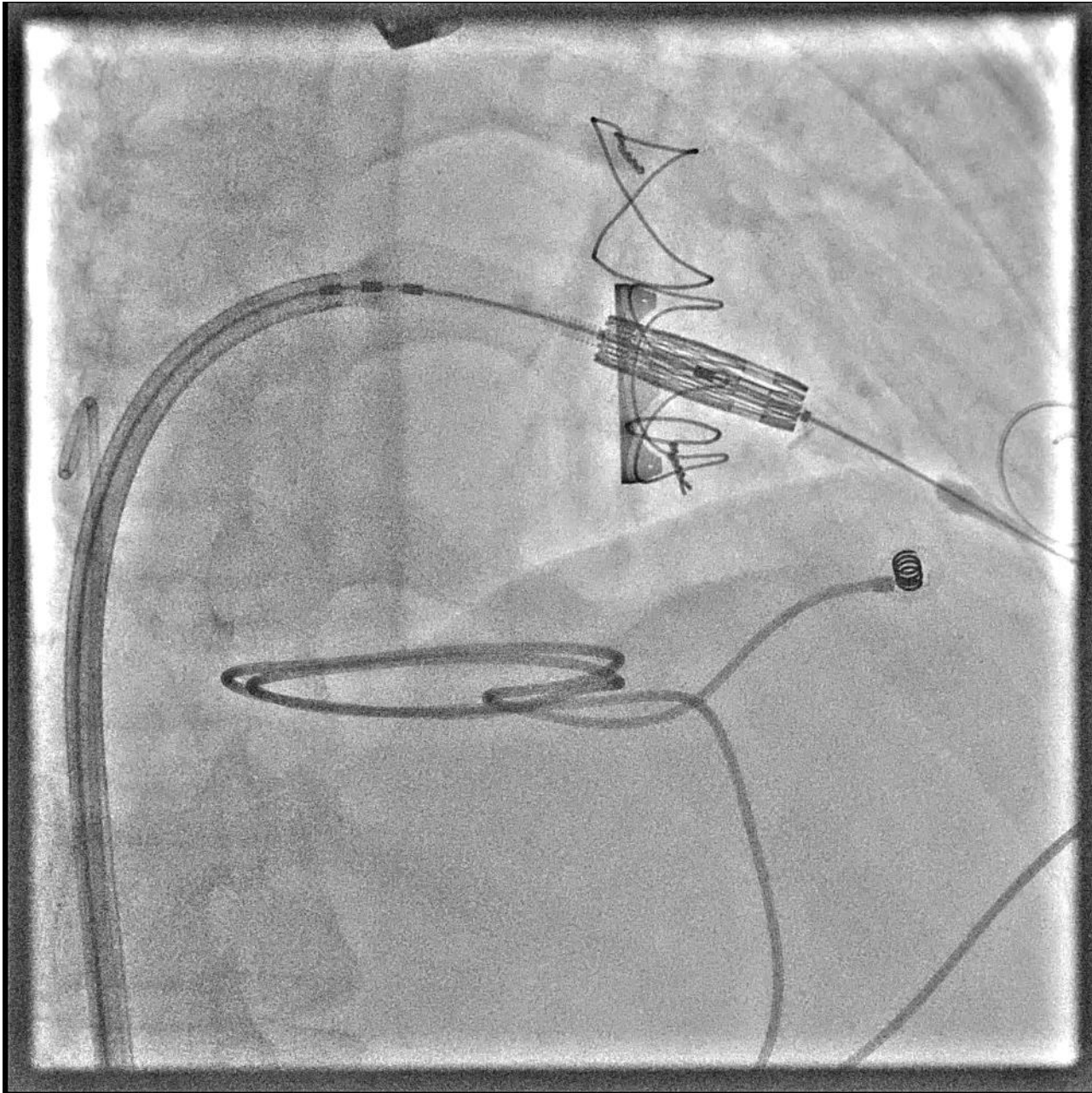


✕ Vmax 175 cm/s
Vmean 126 cm/s
Max PG 12 mmHg
Mean PG 7 mmHg
VTI 49.2 cm

+ Vmax 146 cm/s
Vmean 94.2 cm/s
Max PG 9 mmHg
Mean PG 4 mmHg
VTI 49.6 cm







Age 7 yr: BDG, leaving PS

Age 18 yr: TV replacement, atrial septectomy, VVIR

Age 22 yr Atrial flutter s/p RFA

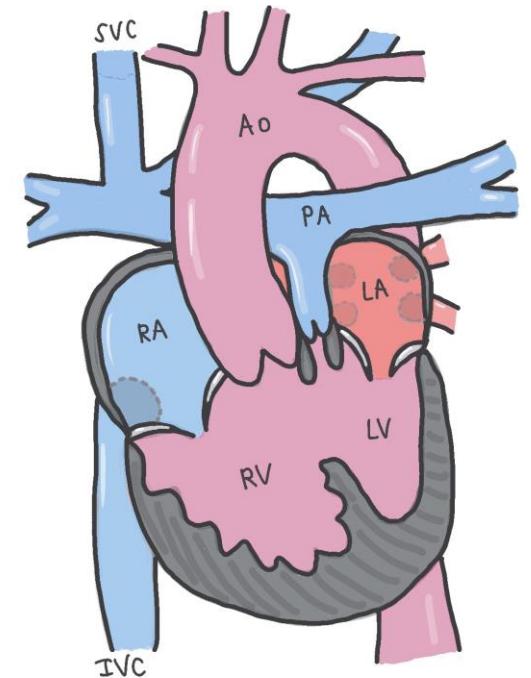


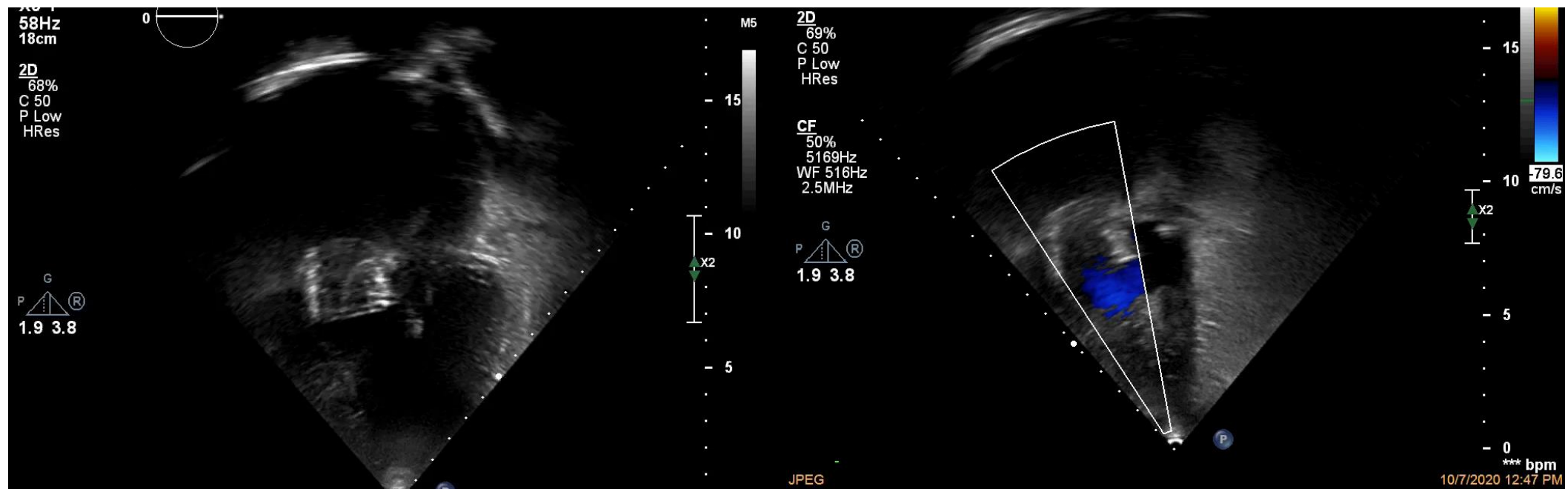
Age 24 yr: Severe TS, moderate MR, moderate MS

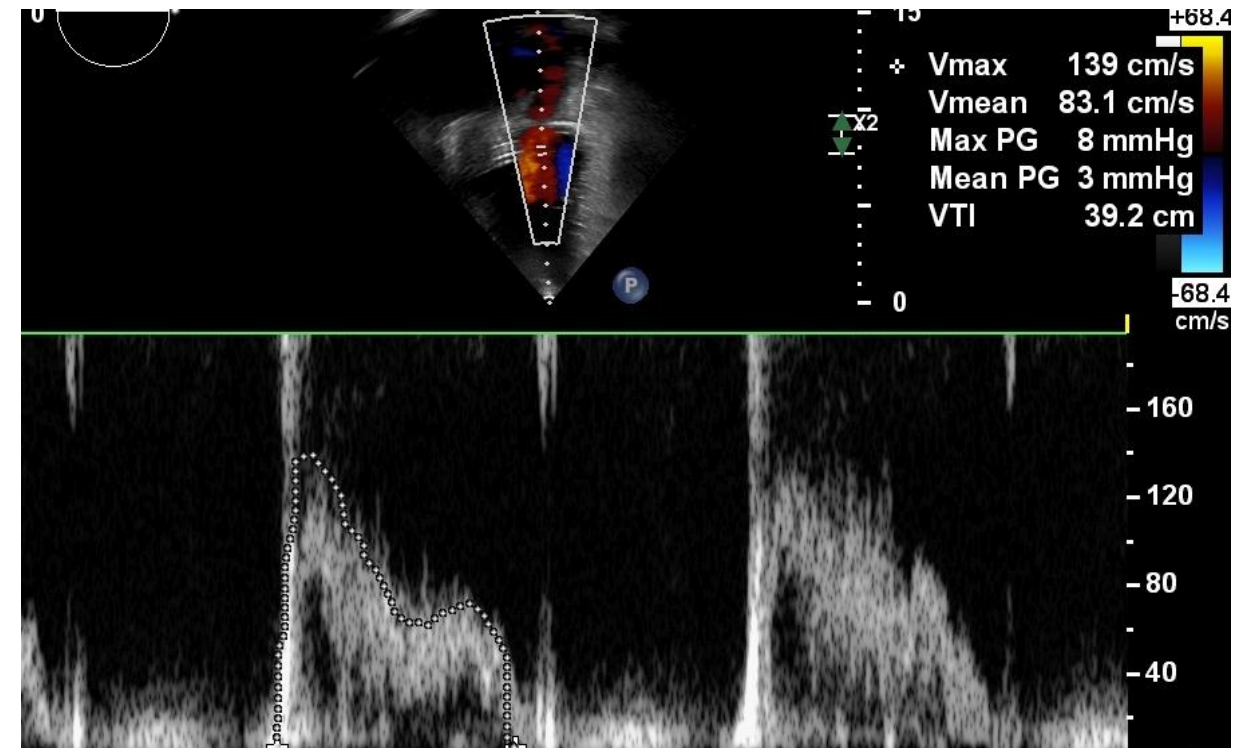
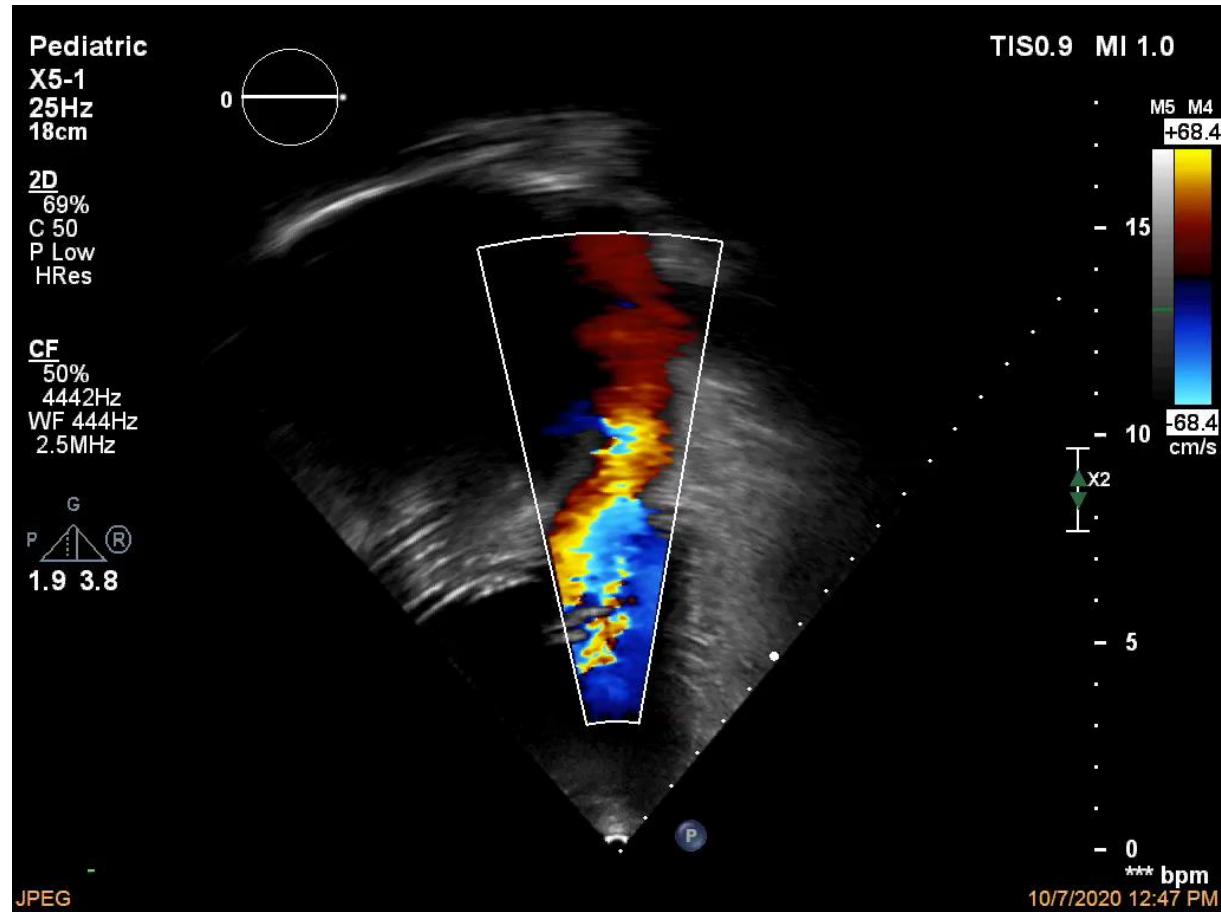
Transcatheter Tricuspid ViV



- Follow up
- O₂sat 79-83%
- FC II
- Plavix, ASA, Warfarin, Diuretics, Enalapril, Thyroxine, Sildenafil



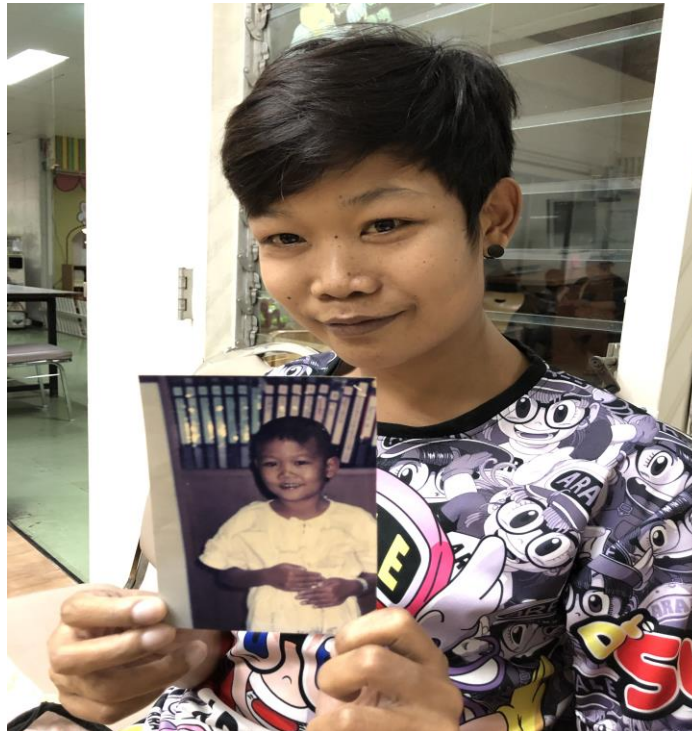






ศูนย์โรคหัวใจ
โรงพยาบาลจุฬาลงกรณ์
สภากาชาดไทย

“FIGHTING TOGETHER”



CASE 4: 16-year-old, male

Progressive dyspnea ~3 months

ToF

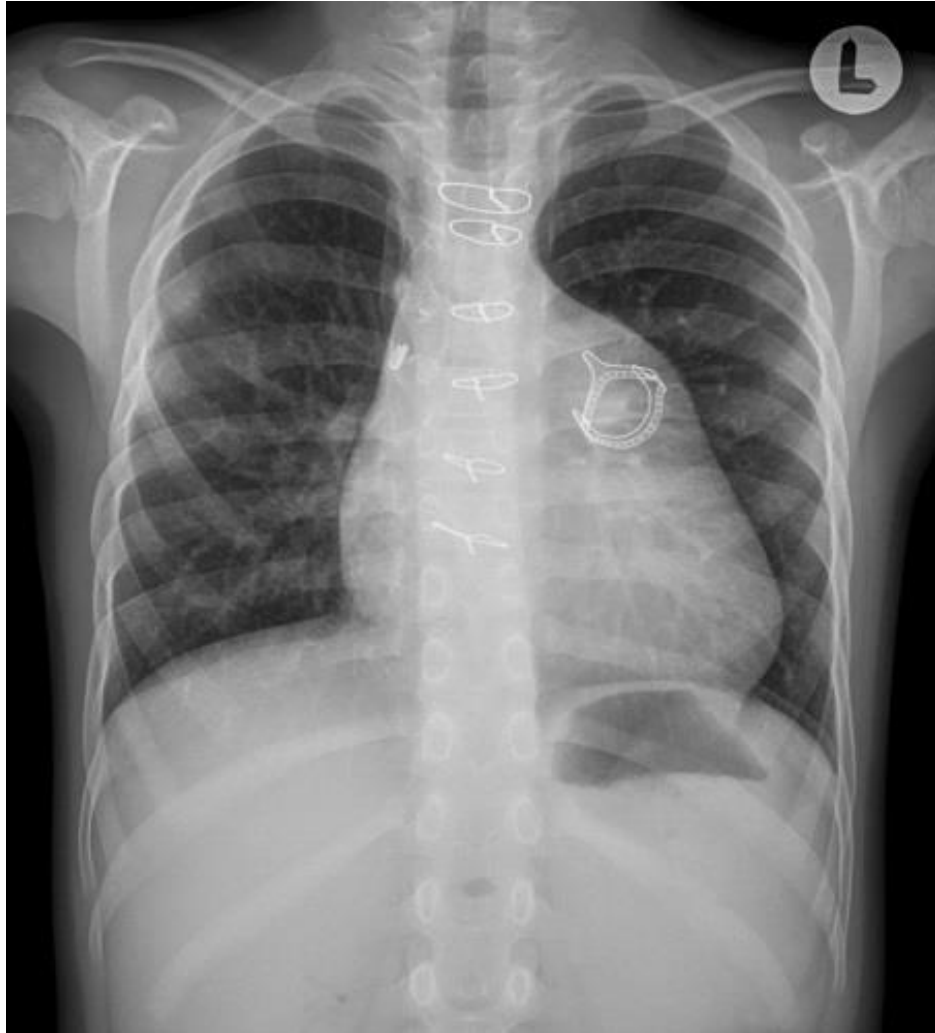
- Age 1½ yrs: RMBT shunt 5 mm
- Age 2 ½ yrs: Total correction (TAP with bovine pericardium monocusp)
- Age 7 yrs: PVR (Bovine 25 mm, Magna), LPA plasty by Gore-Tex, PDA closure and TV repair

LOSS TO F/U

Age 16 yrs:

- Progressive dyspnea, CHF
- CMR: Dilated RVOT, Severe PR, MPA 29.3 mm, RVEDVi 329.9 mL/m², RVEF 25%, LVEF 15.7%
- Rx CHF → Refer

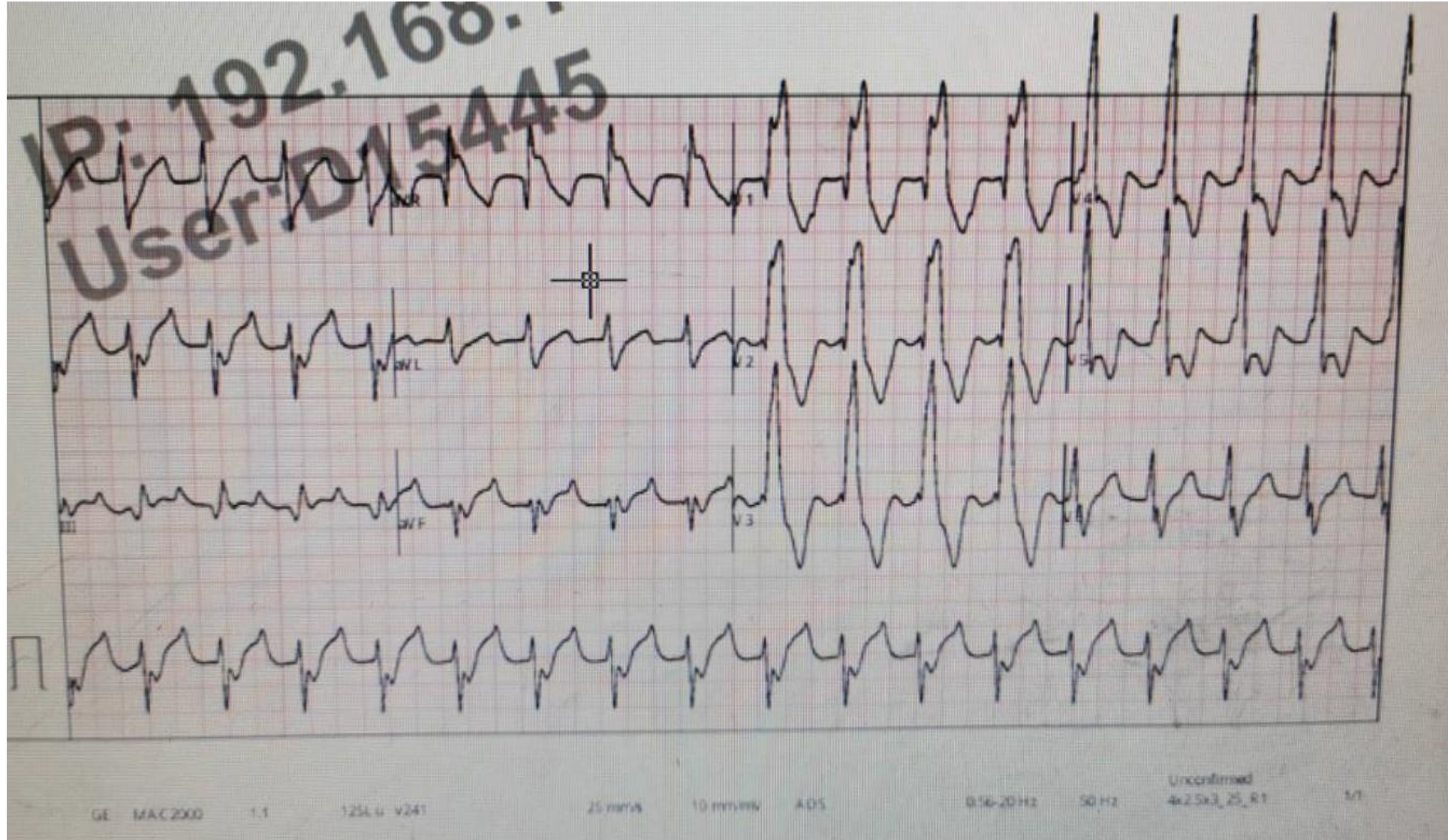
Age 9 years

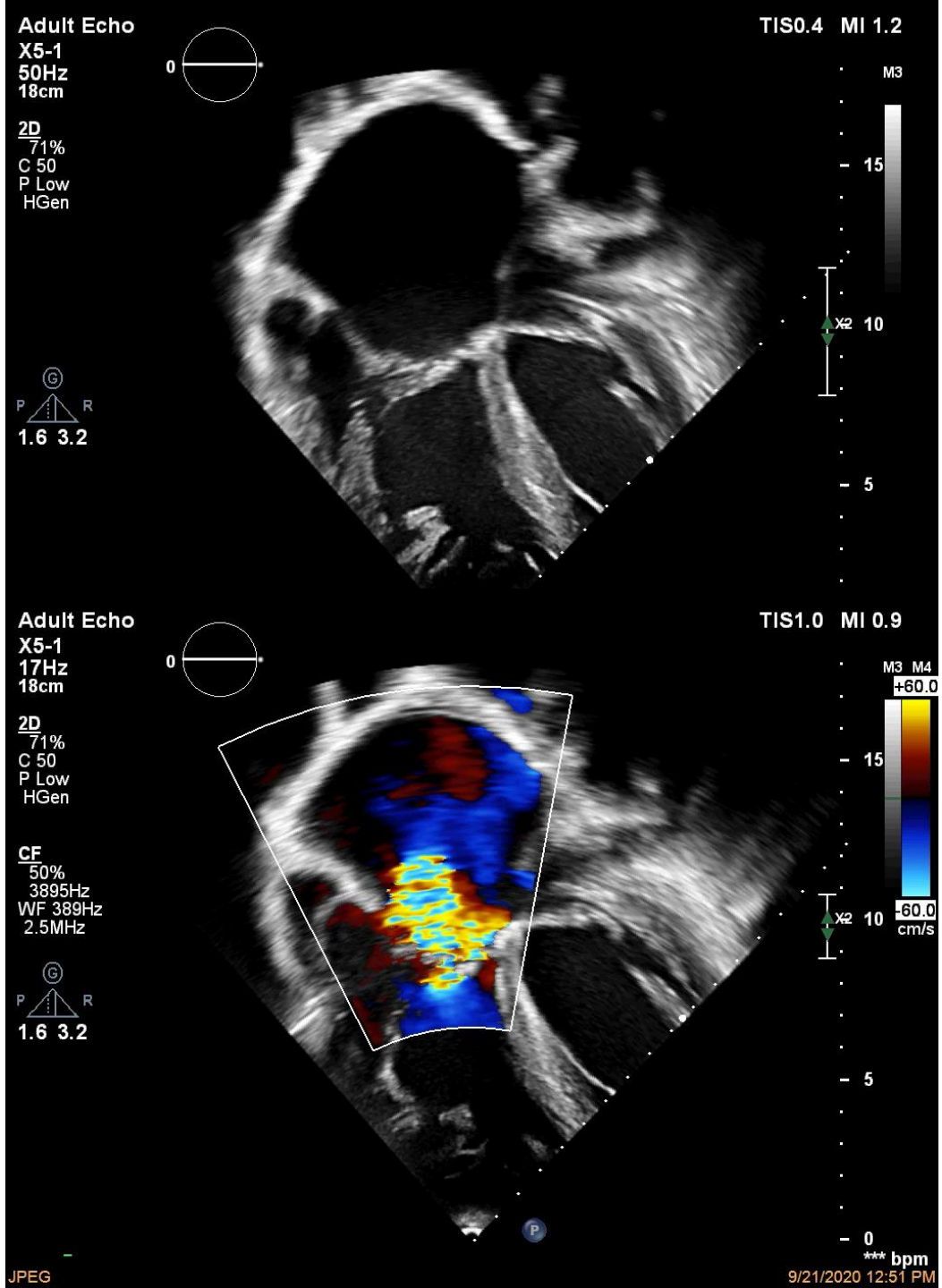


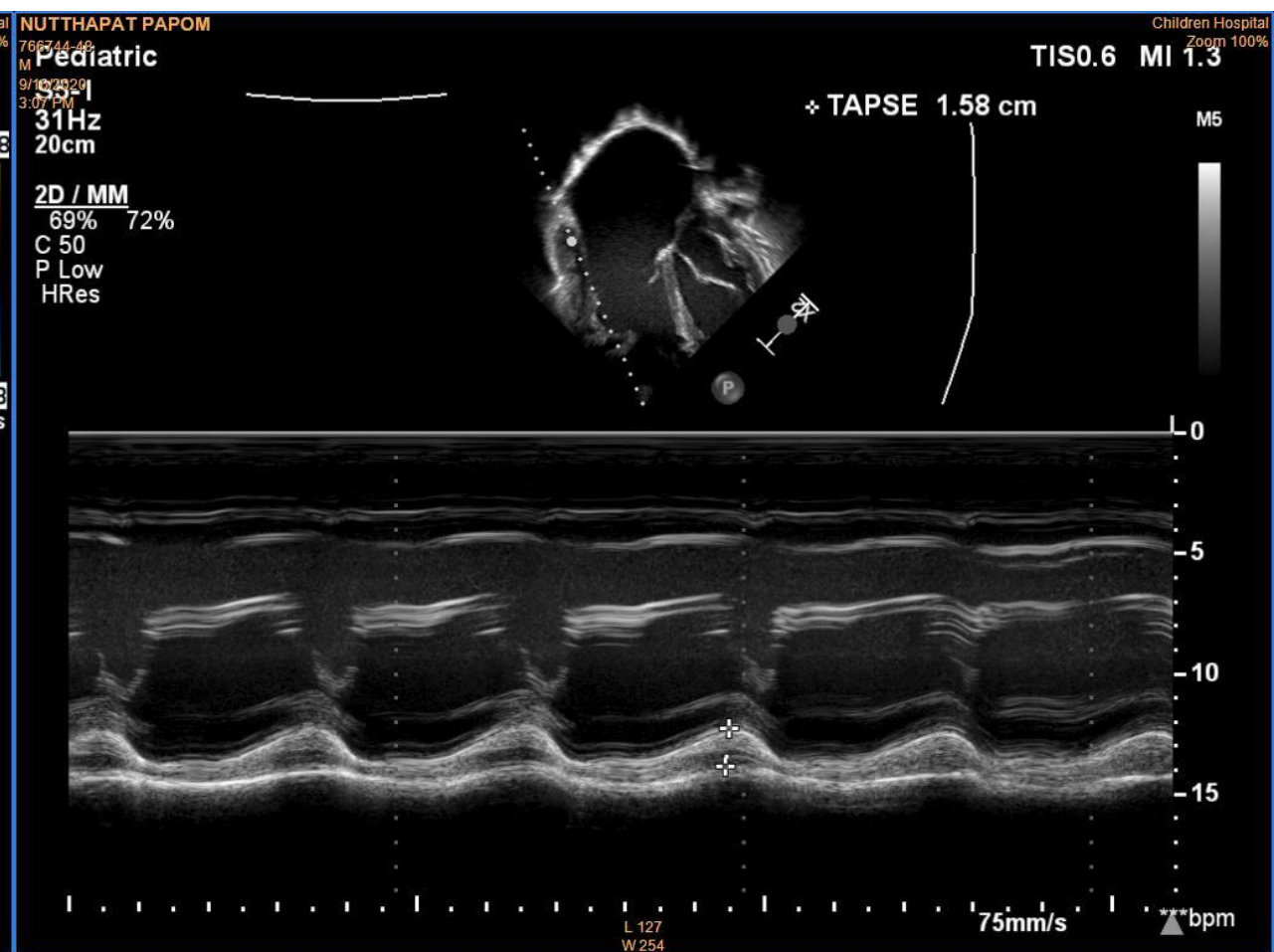
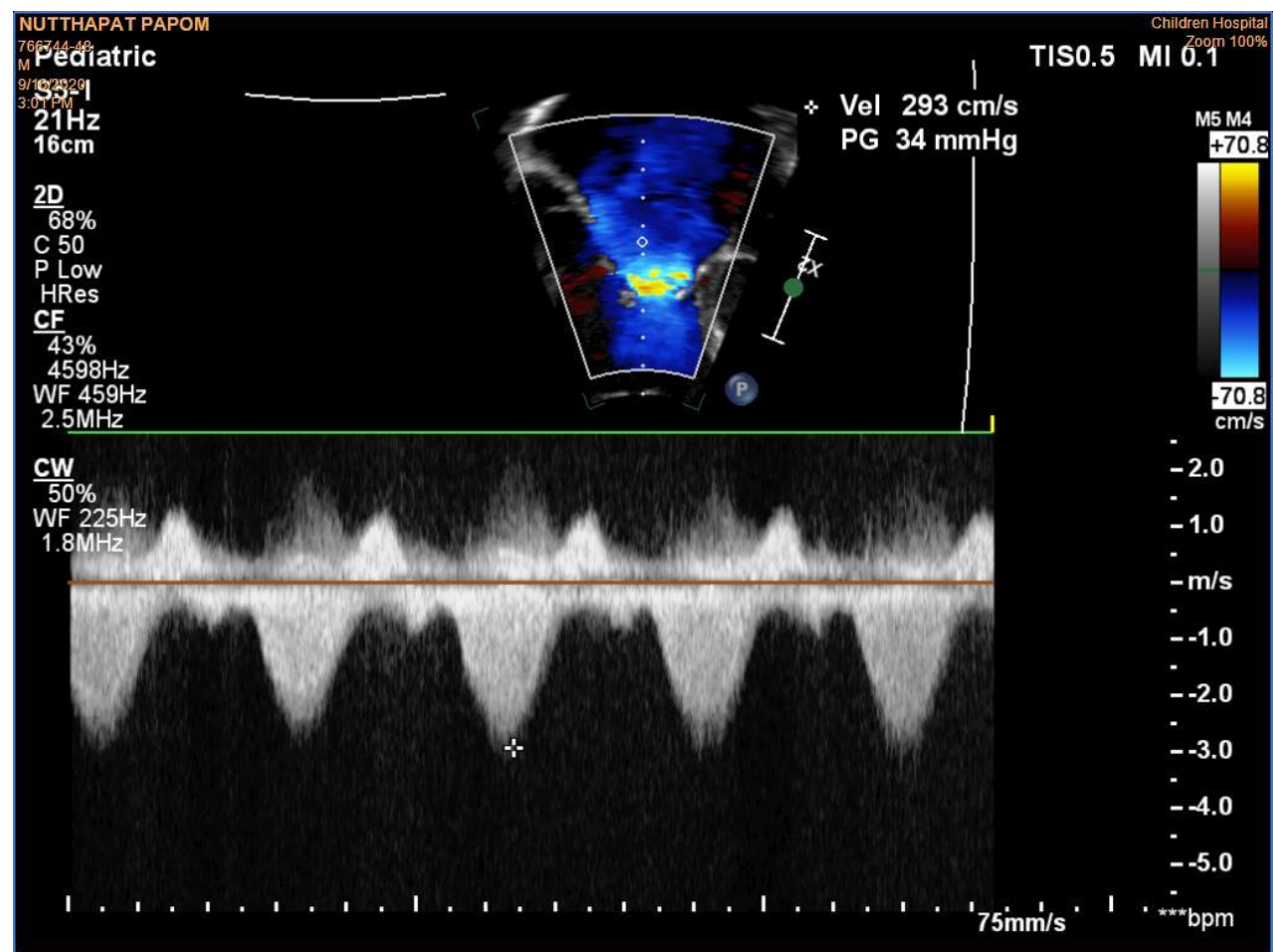
This visit: Age 16 years

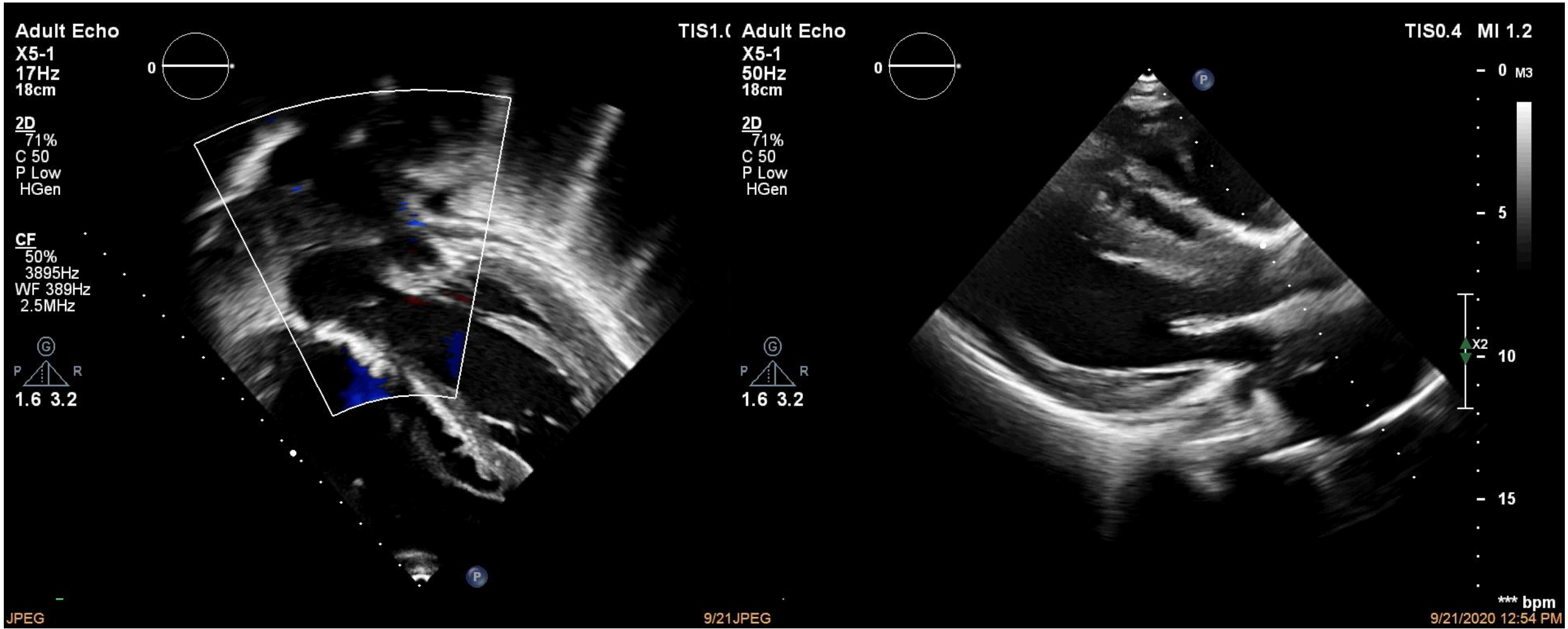


EKG: QRS duration 210 msec









Adult Echo

X5-1
50Hz
18cm

2D
71%
C 50
P Low
HGen

①
P R
1.6 3.2



P

TIS0.4 MI 1.2

- 0 M3

- 5

- 10

- 15

*** bpm

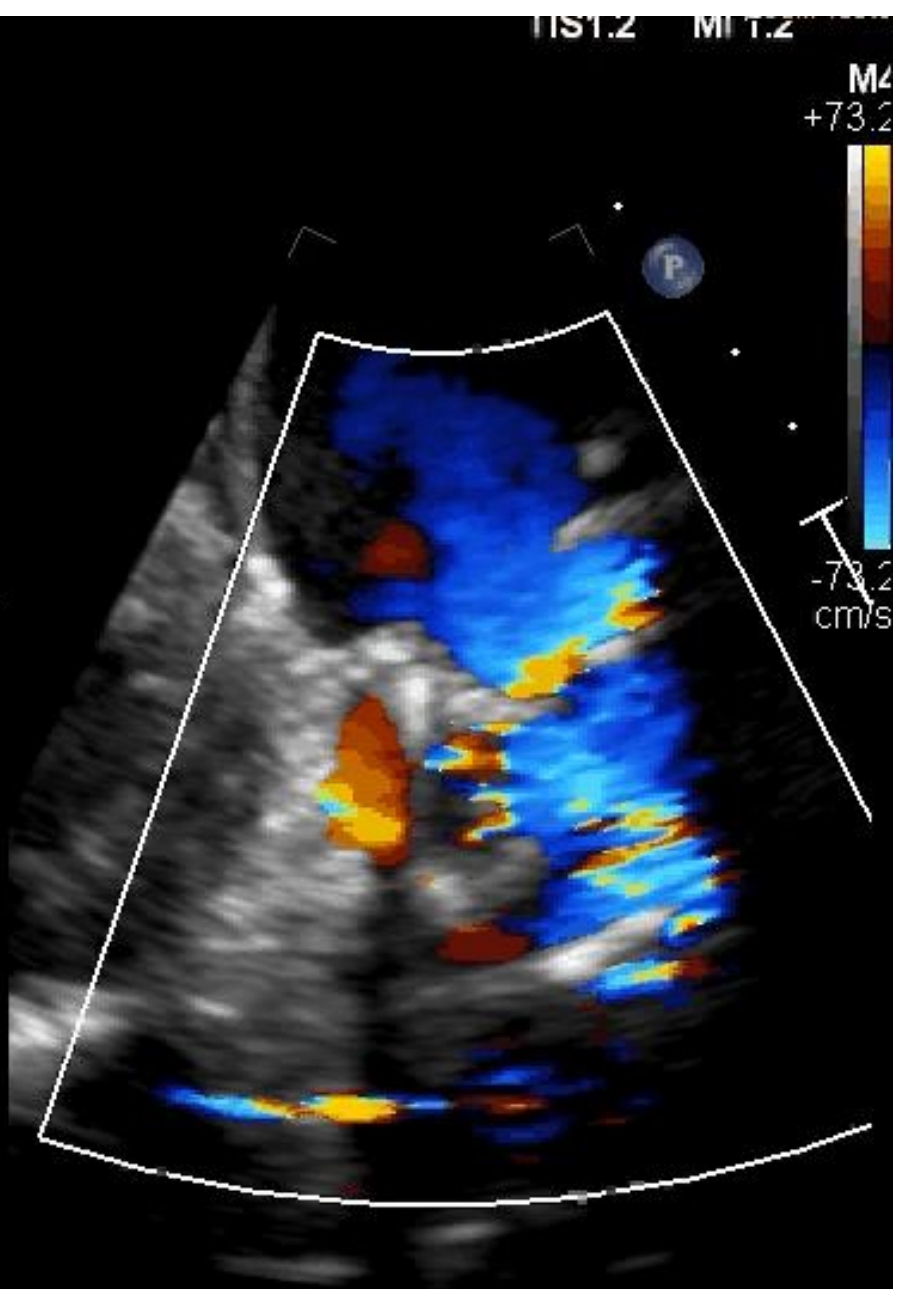
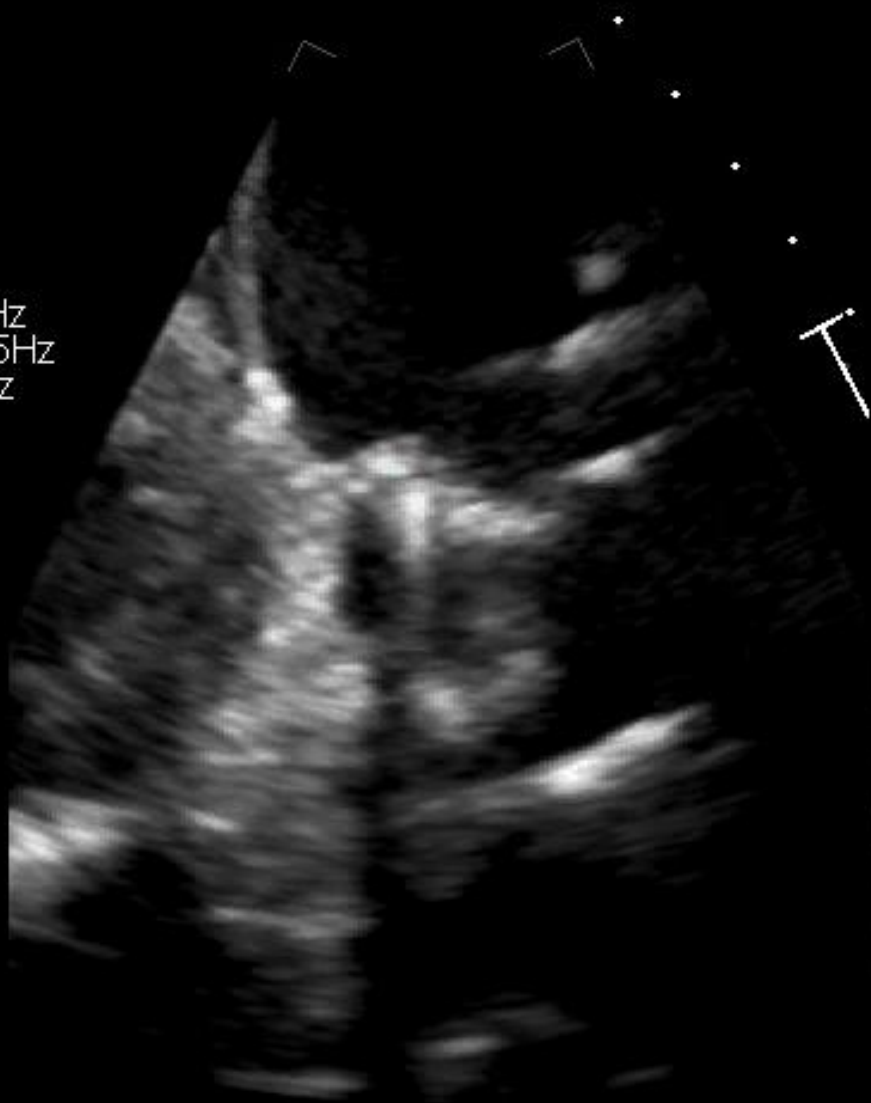
JPEG

9/21/2020 12:52 PM

Pediatric
M
05-1
9/10/2020
3:17 PM
18Hz
14cm
Image 1/37

2D
69%
C 50
P Low
HRes
CF
43%
4750Hz
WF 475Hz
2.5MHz

0 BPM

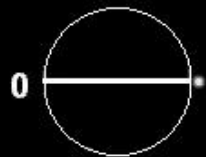


*** bpm

Adult Echo

X5-1
50Hz
18cm

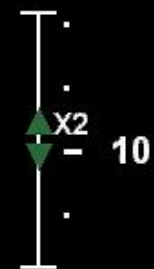
2D
71%
C 50
P Low
HGen



TIS0.4 MI 1.2

- 0 M3

- 5



- 15

*** bpm

Ⓒ
P R
1.6 3.2

JPEG

9/21/2020 12:53 PM

Adult Echo

X5-1
45Hz
18cm



2D / MM

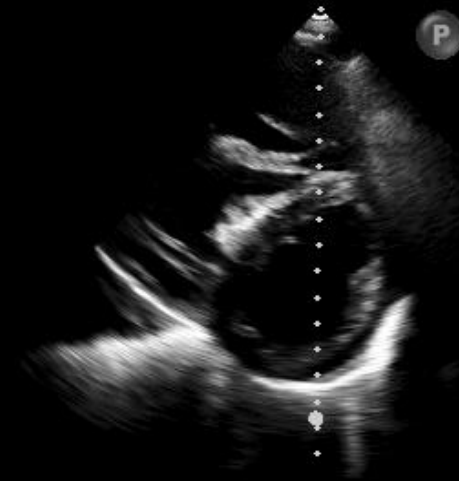
74% 68%

C 50

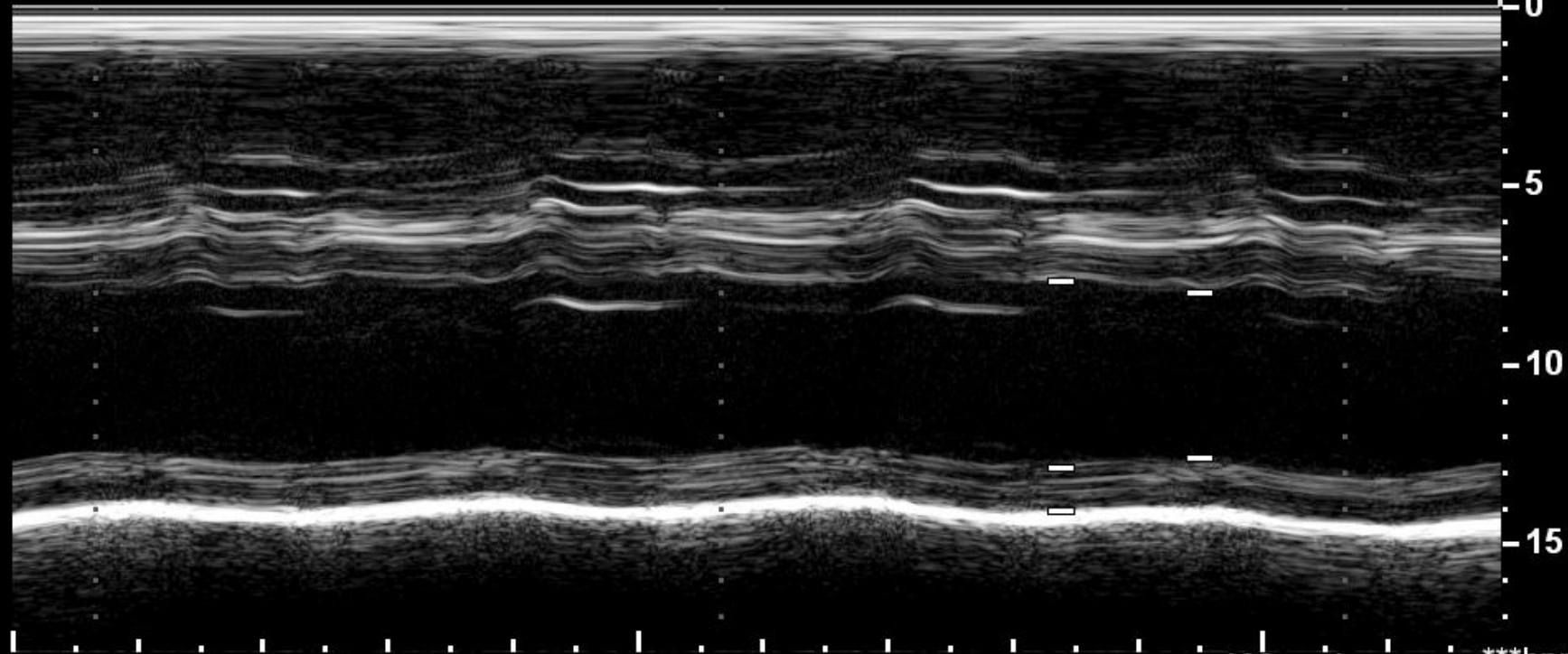
P Low

HGen

TIS0.4 MI 1.0



0	M3
- LVIDd	5.21 cm
- LVPWd	1.20 cm
- LVIDs	4.61 cm
EDV (MM-Teich)	130 ml
FS (MM-Teich)	11.5 %
ESV (MM-Teich)	97.8 ml
EF (MM-Teich)	24.8 %



100mm/s

***bpm

21/9/2563 12:55

TTE

- Dilated hepatic vein and IVC, RAE
- Very poor systolic function both ventricles, LVEF 24%
- Severe TR, atrial septum bulging to the left
- No MR
- Free PR

IMPRESSION

- ToF
- s/p Total correction with TAP at age 2½ years
- s/p Redo-PVR (Bovine 25 mm, Magna), LPA plasty by Gore-Tex, PDA closure and TV repair at age 7 year
- CHF, severe TR, severe PR, poor both ventricular function

What is the next plan?

Recommendations for intervention after repair of tetralogy of Fallot (1)

Recommendations	Class	Level
Pulmonary valve replacement is recommended in symptomatic patients with severe PR ^a and/or at least moderate RVOTO. ^b	I	C
In patients with no native outflow tract, ^c catheter intervention (TPVI) should be preferred if anatomically feasible.	I	C
<p>Pulmonary valve replacement should be considered in asymptomatic patients with severe PR and/or RVOTO when one of the following criteria is present:</p> <ul style="list-style-type: none"> • Decrease in objective exercise capacity • Progressive RV dilation to RVESVi ≥ 80 mL/m² and/or RVEDVi ≥ 160 mL/m^{2d} and/or progression of TR to at least moderate • Progressive RV systolic dysfunction • RVOTO with RVSP >80 mmHg. 	IIa	C

^aRegurgitant fraction by CMR >30–40%. - ^bPeak velocity >3 m/s - ^cPatients with previous RVOT surgery using homografts, bovine jugular vein grafts, bioprostheses/conduits - ^dConfirmed by repeated measurements.

Recommendations for intervention after repair of tetralogy of Fallot (2)

Recommendations	Class	Level
VSD closure should be considered in patients with residual VSD and significant LV volume overload or if the patient is undergoing pulmonary valve surgery.	Ila	C
In patients with sustained VT who are undergoing surgical pulmonary valve replacement or transcatheter valve insertion, pre-operative catheter mapping and transection of VT-related anatomical isthmuses before or during the intervention should be considered.	Ila	C
Electrophysiologic evaluation, including programmed electrical stimulation, should be considered for risk stratification for SCD in patients with additional risk factors (LV/RV dysfunction; non-sustained, symptomatic VT; QRS duration ≥ 180 ms, extensive RV scarring on CMR).	Ila	C

PLAN

- Intervention
 - IABP
 - Transcatheter pulmonic valve-in-valve



NATAPAT, PAPROM
766744-48
8/11/2004 M
9/22/2020
2:22 PM
Series 5
Run 5A - Frame 1 / 81
Frontal
22/9/2008

Rajawithi Hospital
Perf. Phys.: WORAKAN, PROMPHAN
70.8kV, - mAs, 559mA, 42.7ms
Zoom 100%



RAO 21.0°
Caudal 18.2°

L 128
W 256

NATAPAT, PAPROM
766744-48
8/11/2004 M
9/22/2020
3:40 PM
Series 28
Run 28 - Frame 1 / 187
22/9/2563

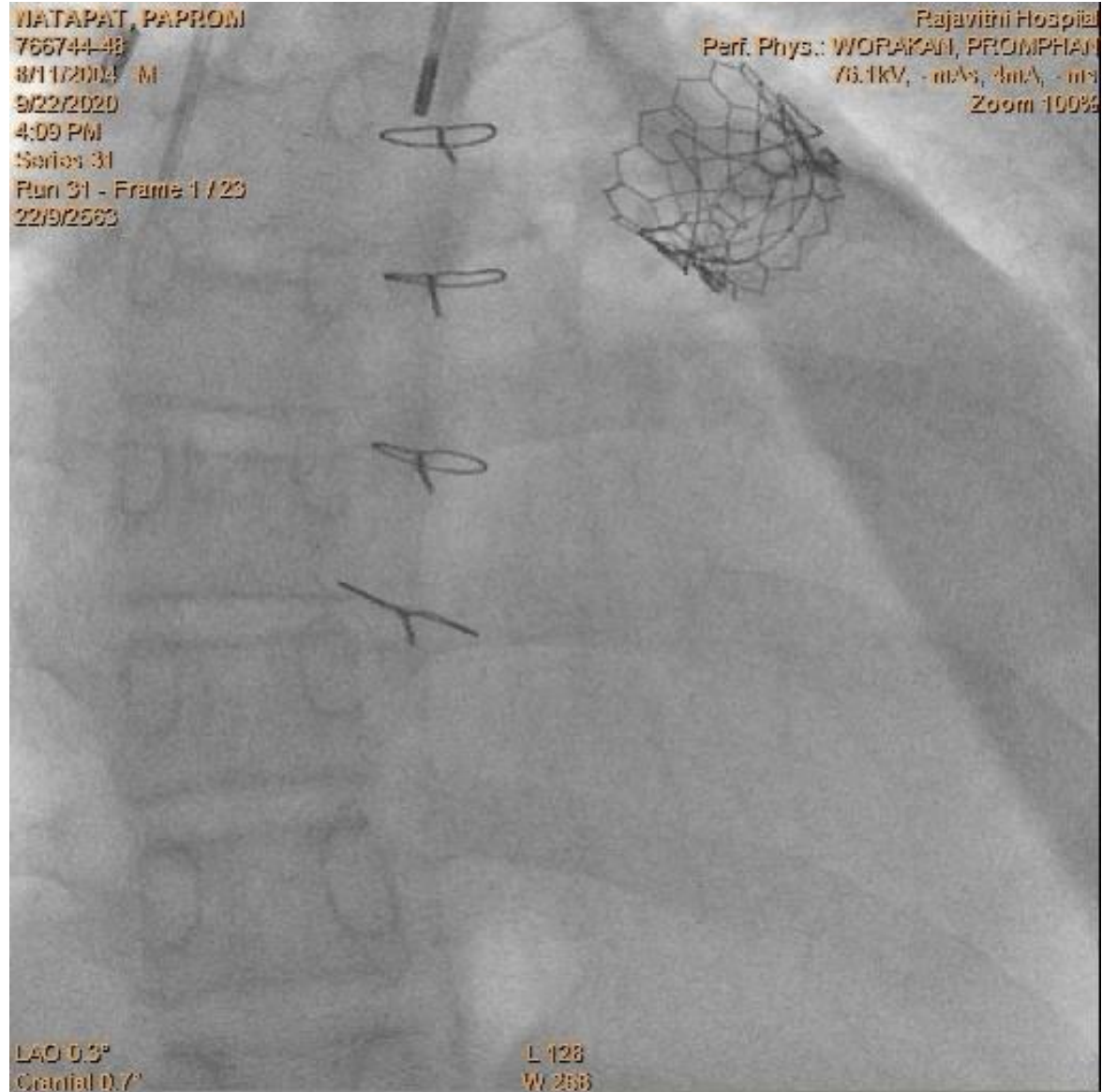
Rajavithi Hospital
Ref: Phys: WOPAKAN, PROMPHAN
87.2kV, - mAs, 8mA, - ms
Zoom 100%



RAO 47.3°
Caudal 26.4°

L 128
W 256

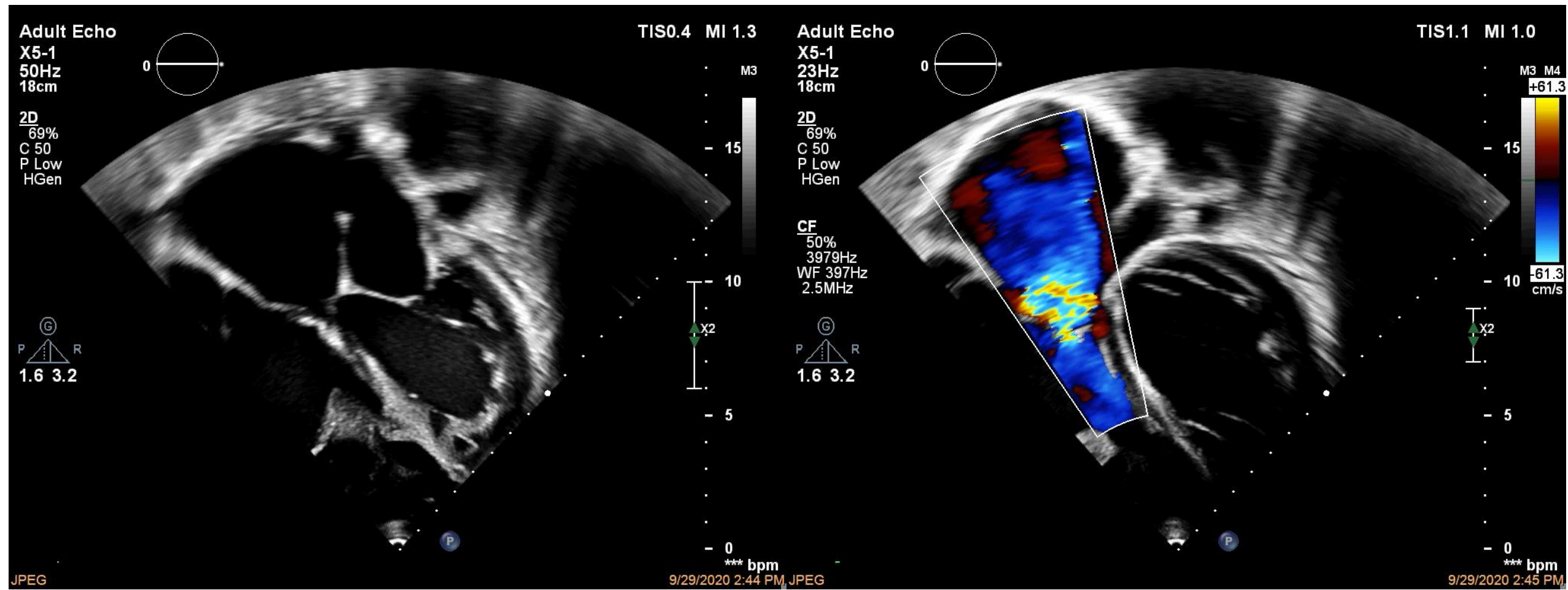
S3 26 mm



Post Pulmonic Valve-in-Valve (PViV)

- Off IABP
- On low dose inotrope 2 days
- Medications: Plavix, ASA, Diuretics, ARB, Lanoxin





Adult Echo

X5-1
23Hz
18cm

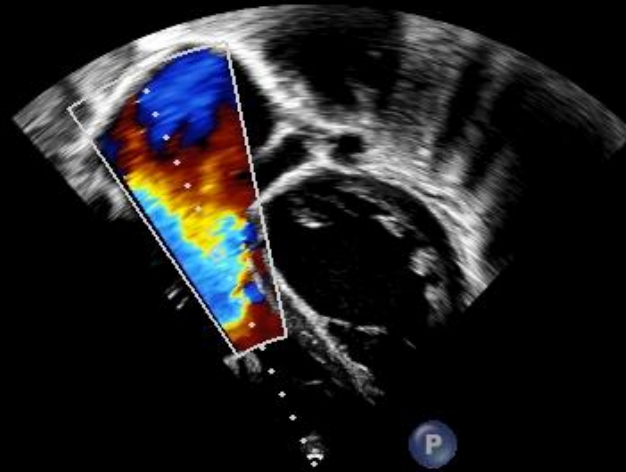


2D

69%
C 50
P Low
HGen

CF

50%
3979Hz
WF 397Hz
2.5MHz



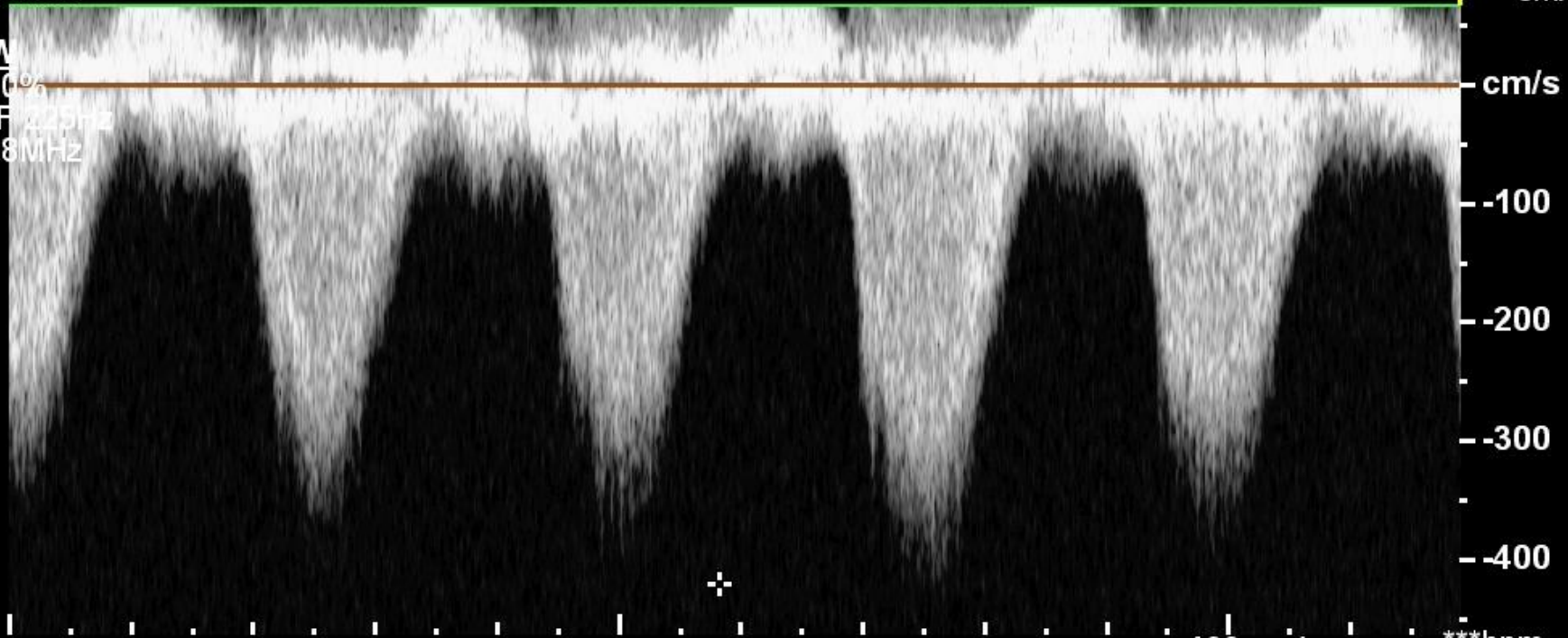
15
+
5
0

Vel 421 cm/s
PG 71 mmHg

M3 M4
+61.3
-61.3
cm/s

CW

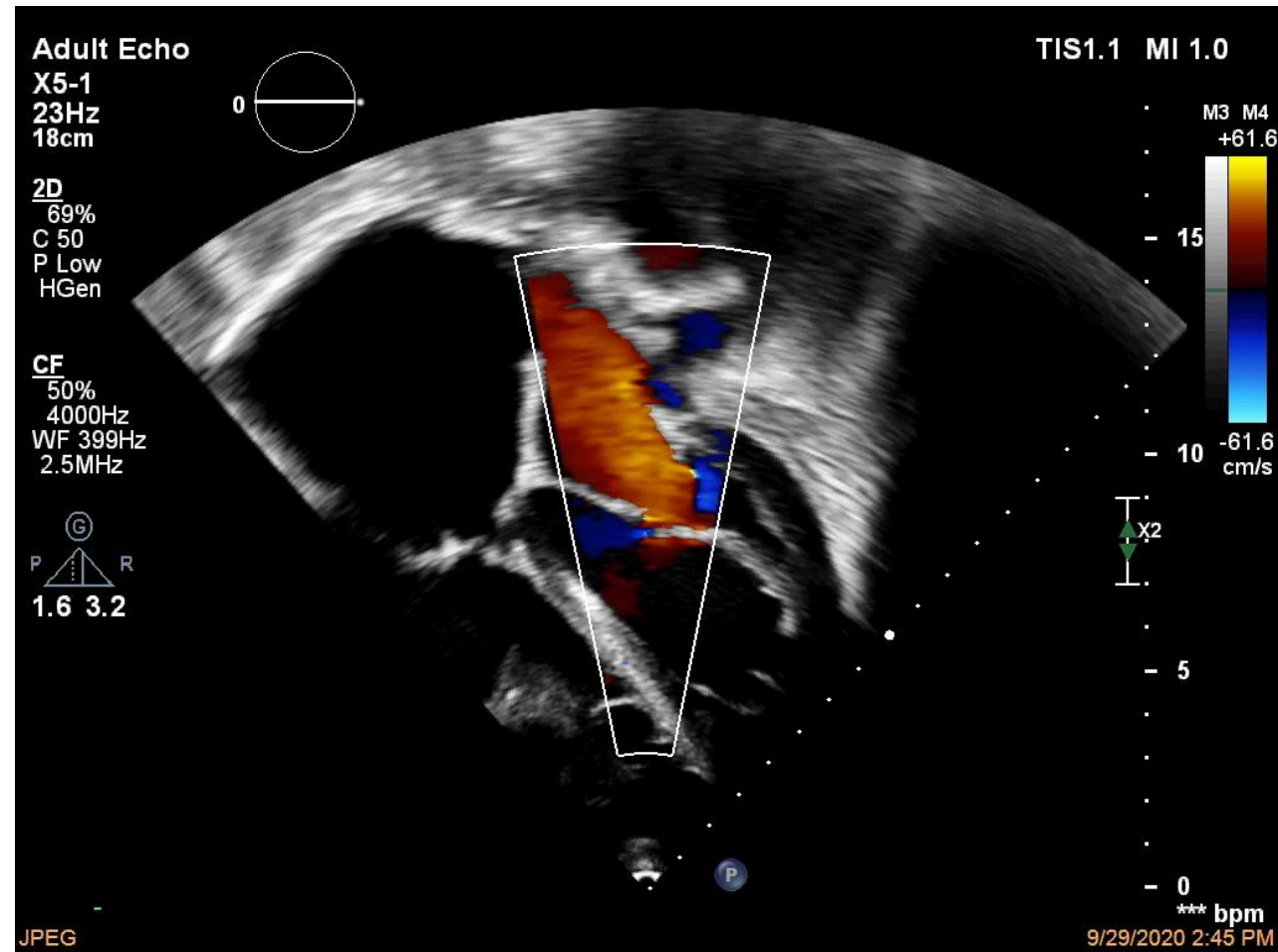
50%
WF 225Hz
1.8MHz

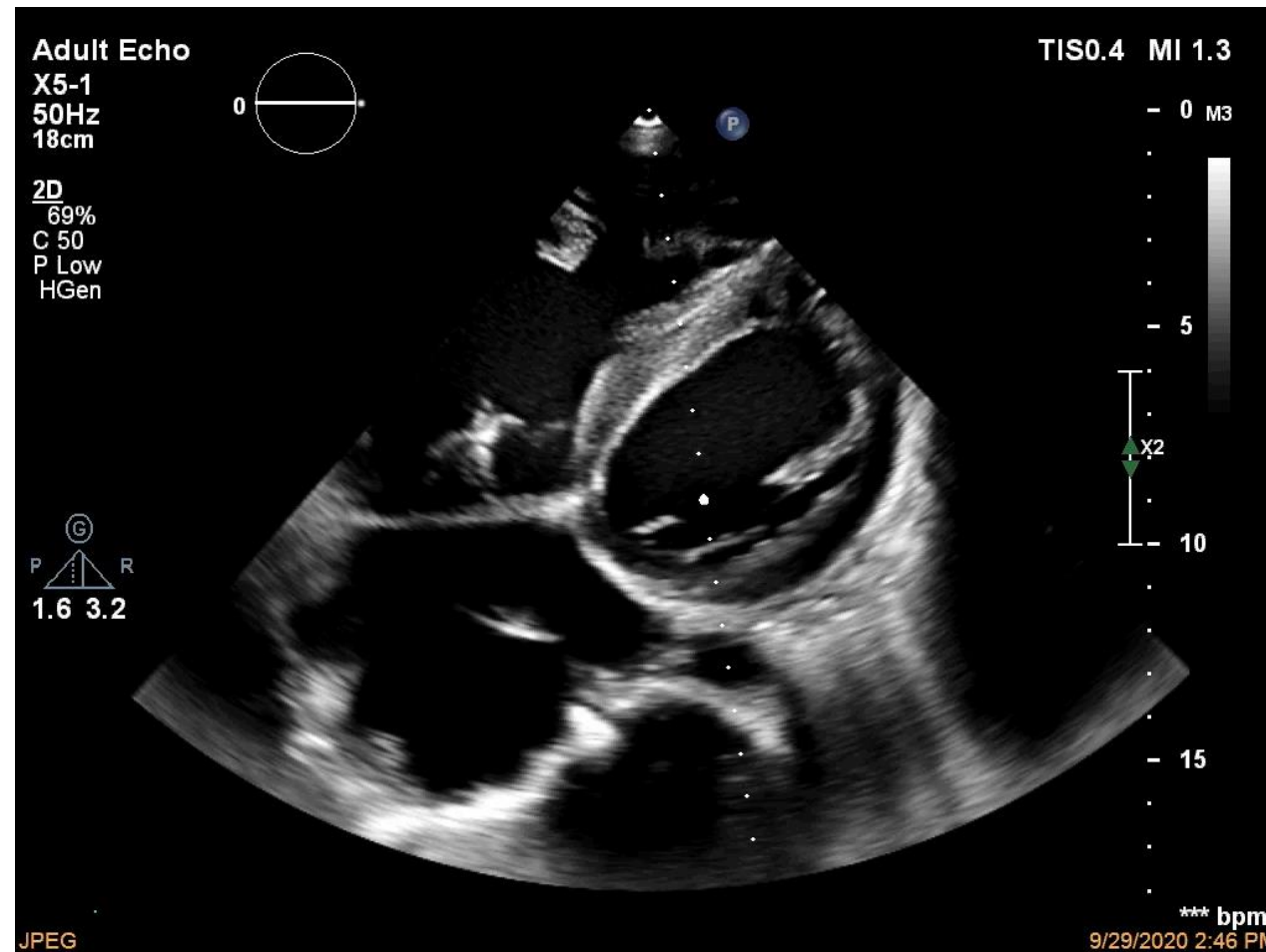


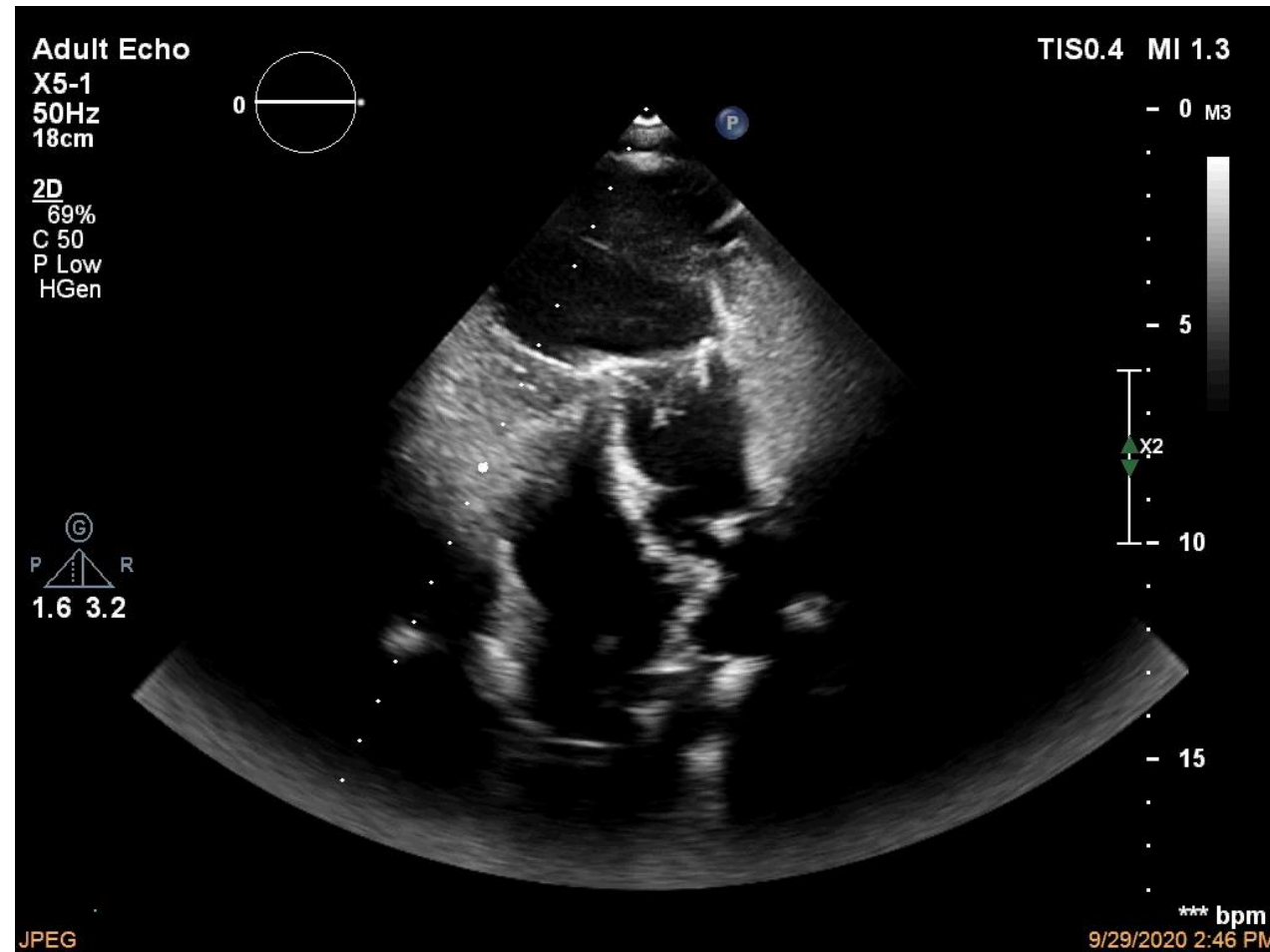
100mm/s

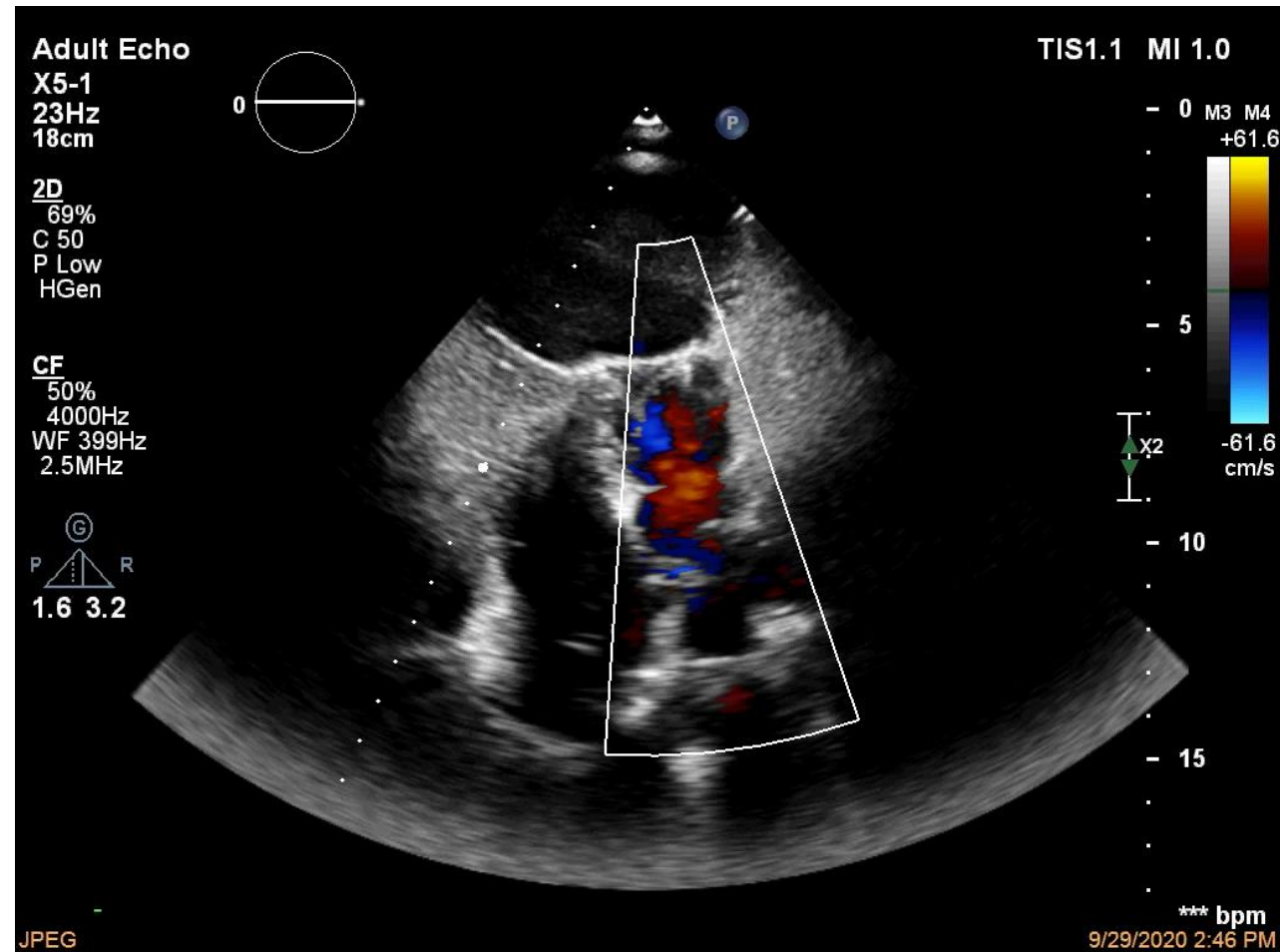
***bpm

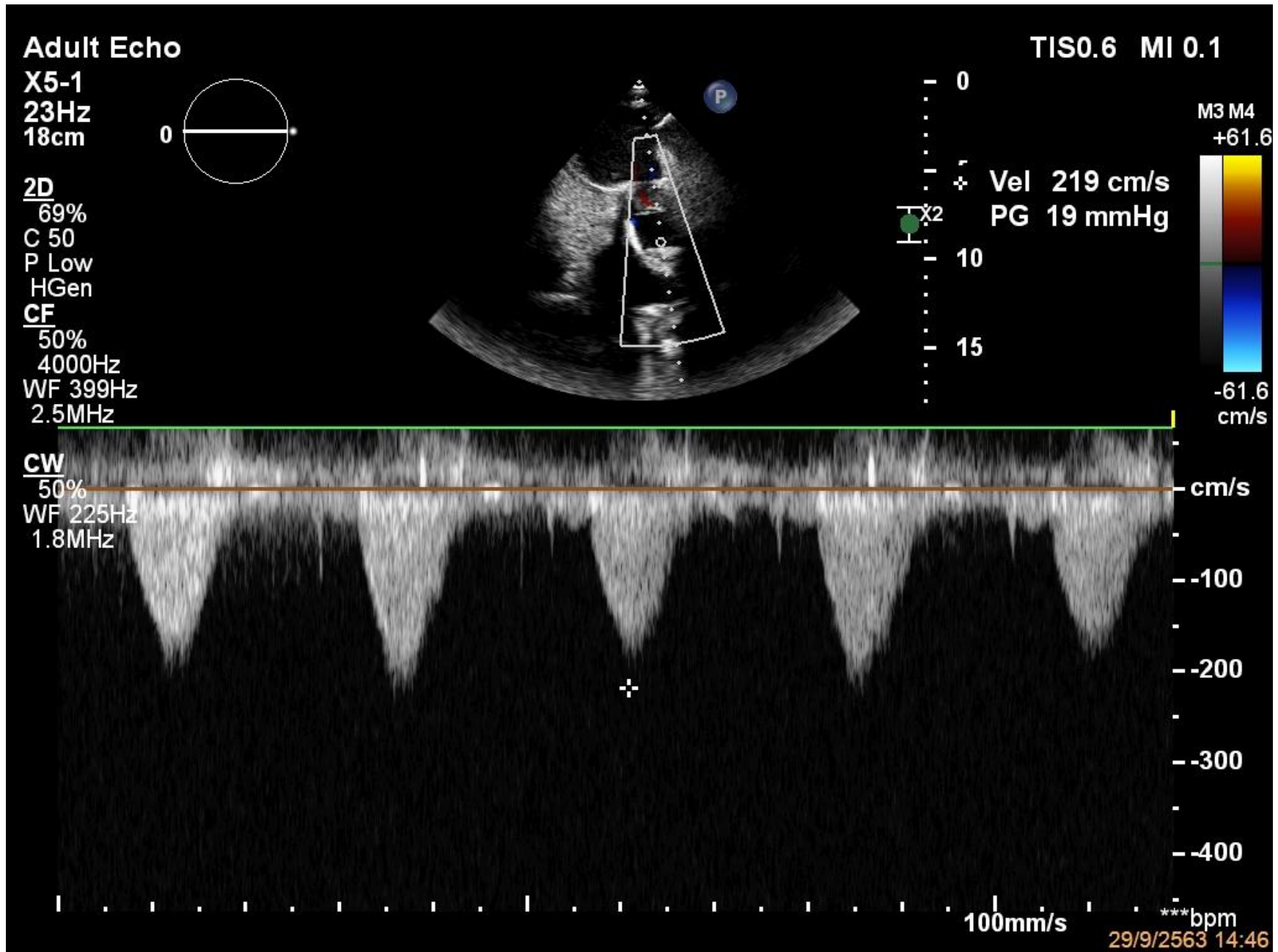
29/9/2563 14:45









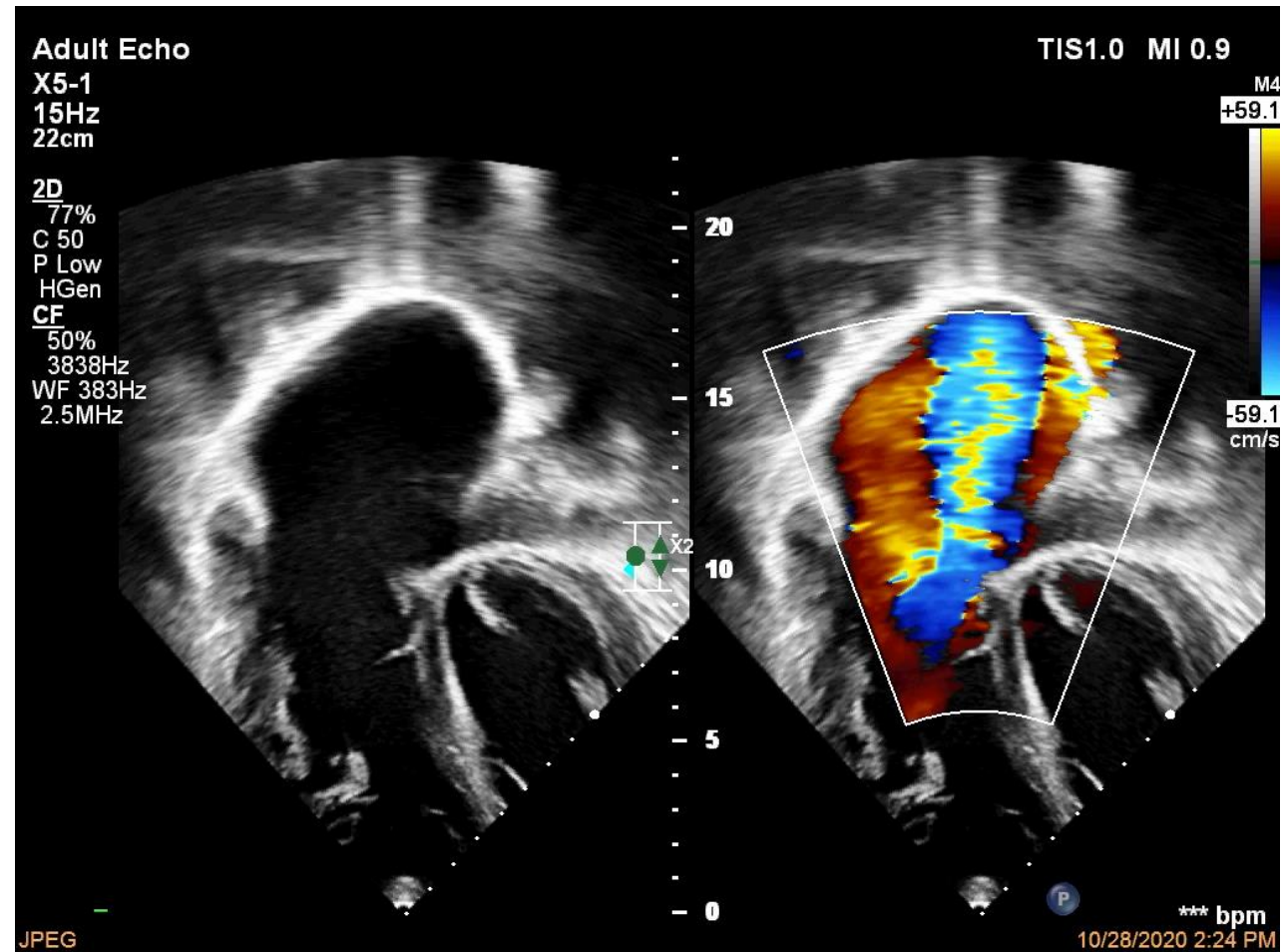


Immediate post PViV



1 month post PViV





Age 2 1/2 yr: Total correction with TAP

**Age 7 yr: Redo-PVR (Bovine 25 mm, Magna),
LPA plasty by Gortex, PDA closure , TV repair**

**Age 16 yr Transcatheter Pulmonic ViV,
SAPIEN3 SIZE 26 MM**

Severe TR, poor both cardiac function

WHAT'S NEXT???

What is the next plan?



ศูนย์โรคหัวใจ
โรงพยาบาลจุฬาลงกรณ์
สภากาชาดไทย

“FIGHTING  TOGETHER”

THANK YOU