An Unusual Presentation of Rare Congenital Heart disease

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5yrs old male child resident of Osmanabad was brought by parents to hospital , mother being informant with chief complaints of

1. Exertional Breathlessness since 2 yrs

2. Chest Pain on exertion since 6 months

Patient was apparently alright 2 yrs back when the mother noticed that patient had breathlessness while playing with other kids of his age he had to stop while playing take rest . During this period mother noticed that he had increased movements on the precordium. Child complained of pain which was present of playing or exertion , non radiating , associated with sweating , relived on rest .

No H/O PND, orthopnoea, leg swelling, facial puffiness

No H/O palpitations, Cyanotic spells, Fever,

- Past History:

No history of recurrent Hospitalization for fever, LRTI, URTI

No history of any surgery

Birth history

Patient was born out of Full Term Normal Vaginal Delivery with normal birth weight and no NICU stay.

He is completely immunized for his age

Developmental Milestones: All social, Motor and cognitive milestones achieved at proper age till 5yrs

- General Exam

Pt was lying in mothers lap in supine postion Spo2-98% on room air

Height- 1.11m weight- 18.6kg, BSA- 0.757m2, BMI- 14.09kg/m2

he is thin bulit and moderately nourished

Pulse- 102 beats/ min regular Normal volume and suptroke

BP- 102/68 mm Hg rt Arm Supine Position

No Pallor cyanosis clubbing lymphadenopathy

JVP – Not raised

No Pedal edema

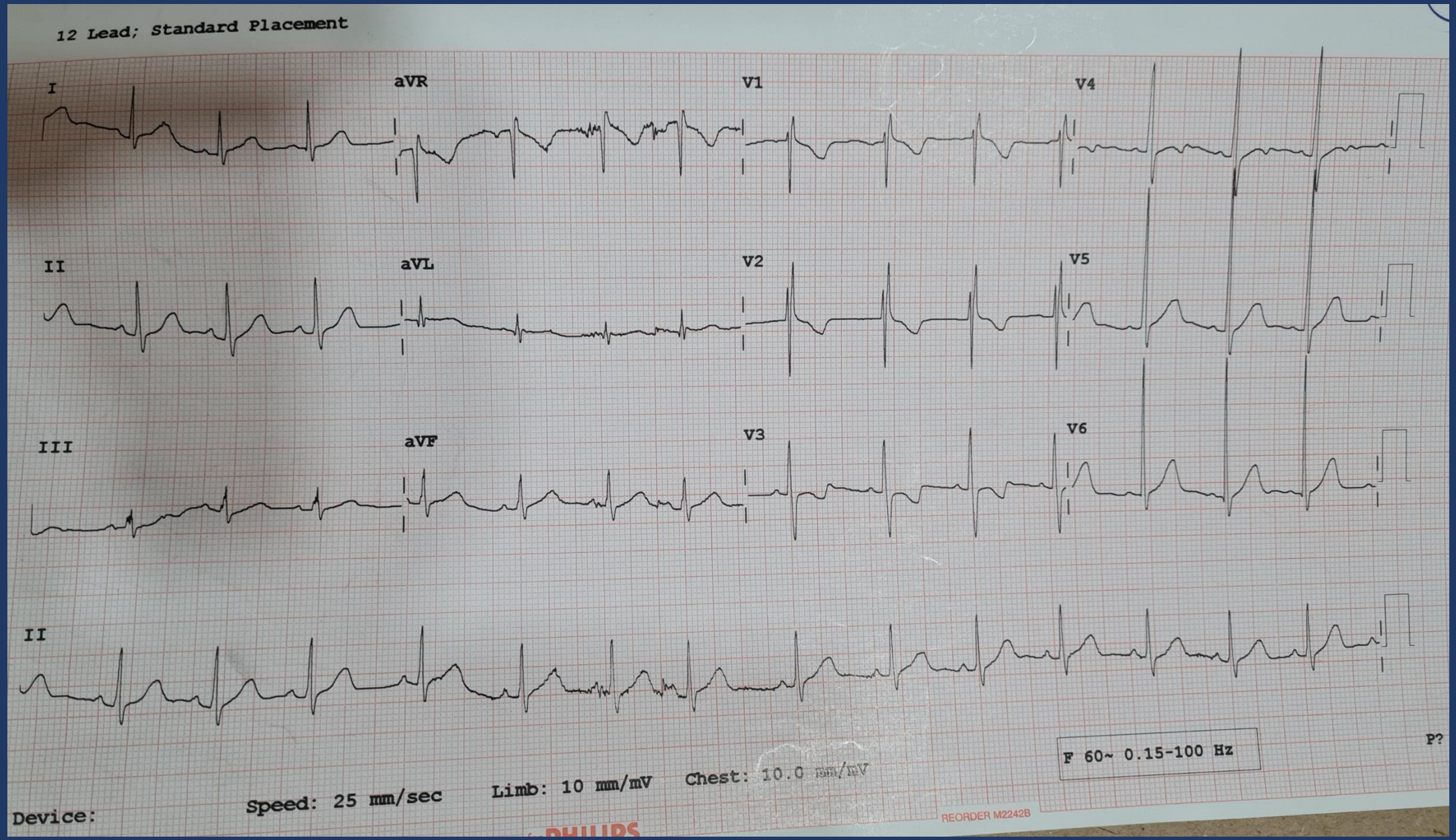
Spine and Joint Exam- Normal

- CVS:
- Shape of precodium- buldging noted in left sternal border with active precordium
- No pulsations over precordium
- Apical Impulse- diffuse (RV Type)
- Thrill- Absent
- Grade I Parasternal Heave / No epigastric pulsations
- continuous murmur at the right 2nd and 3rd intercostal spaces.

RS- NAD

P/A- No organomegaly

- D/Ds
- 1. Congenital acyanotic Heart disease
  - ALCAPA
  - Coronary Cameral Fistula
  - RSOV


12 Lead; Standard Placement
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II
F 60~ 0.15-100 Hz
P?
Device:
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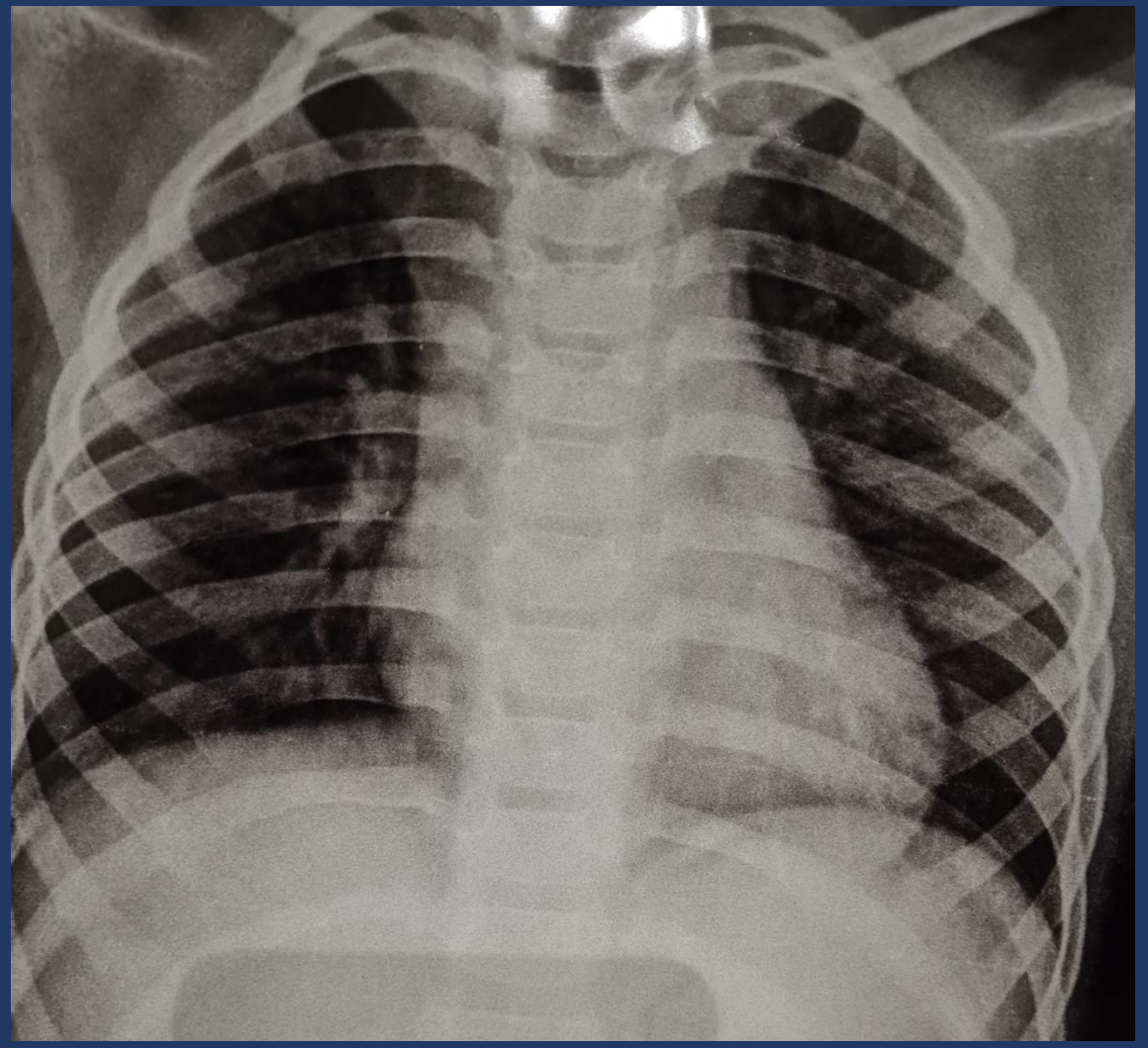
Limb: 10 mm/mV
Chest: 10.0 mm/mV
REORDER M2242B

Xray Chest P/A



2 D Echo

Viscero atrial situs solitus – (S, D, S)

IVC and SVC drain into- RA

3 pul. Vein drain into LA

A-V & V-A Concordance

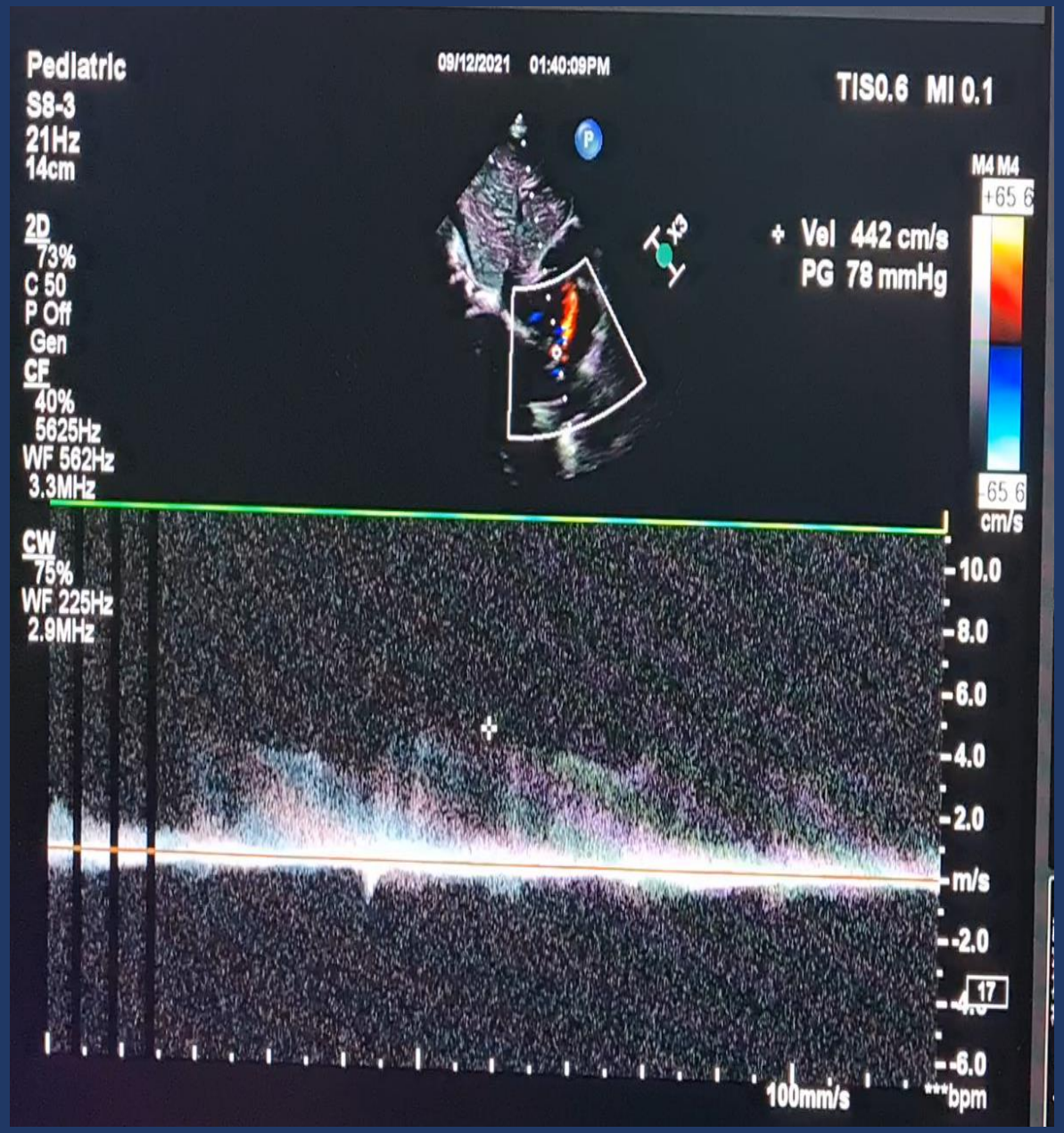
IAS- RA Side od IAS Shows Fistulous tract arising from aorta with left to right shunt with retro aortic course and opening into RA

A-V and Semilunar Valves Normal

Mild RAVA & RVVO

Mild PAH

No PDA , Good biventricular function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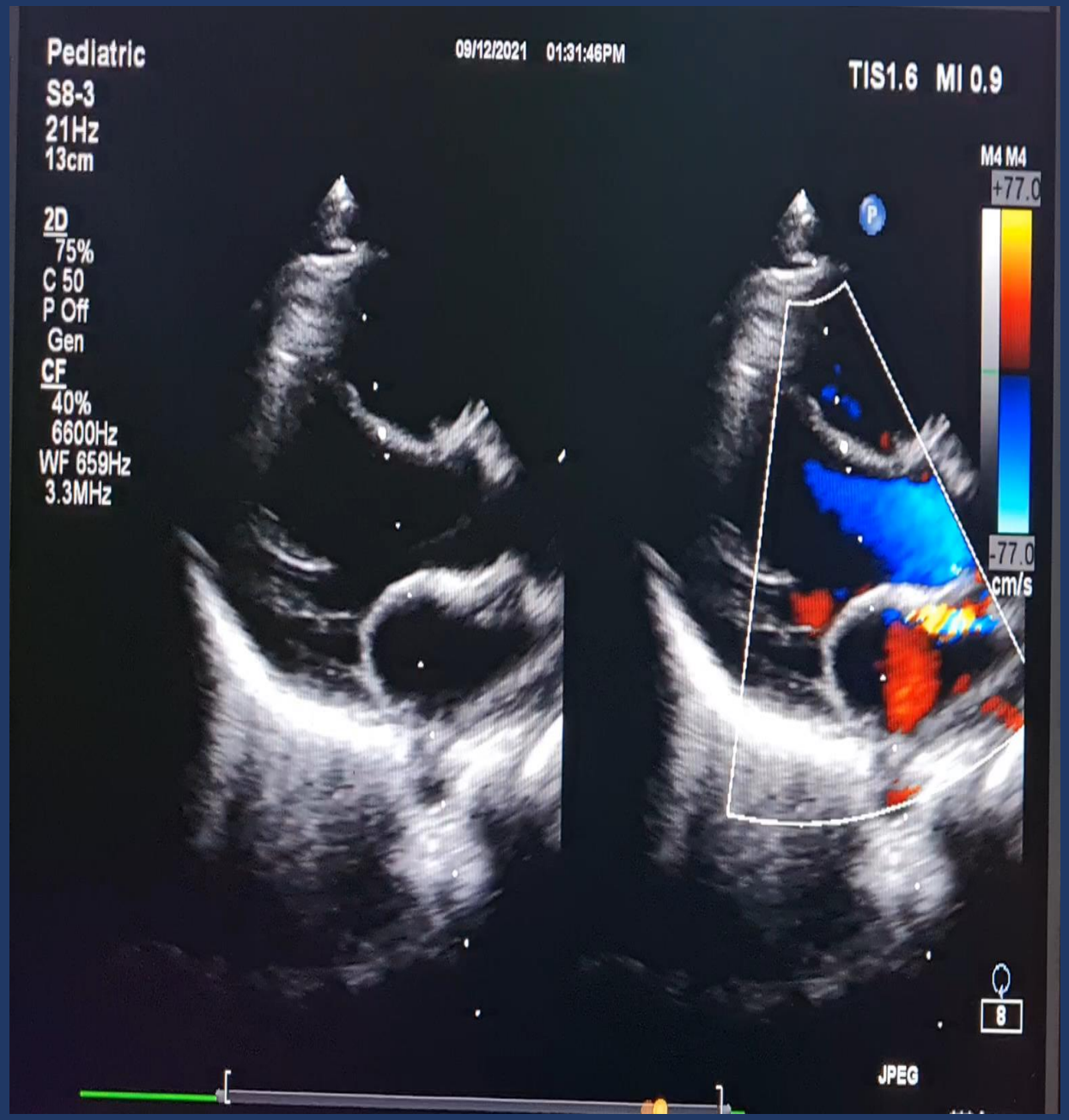
Pediatric
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TIS1.6 MI 0.9
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C 50
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Gen
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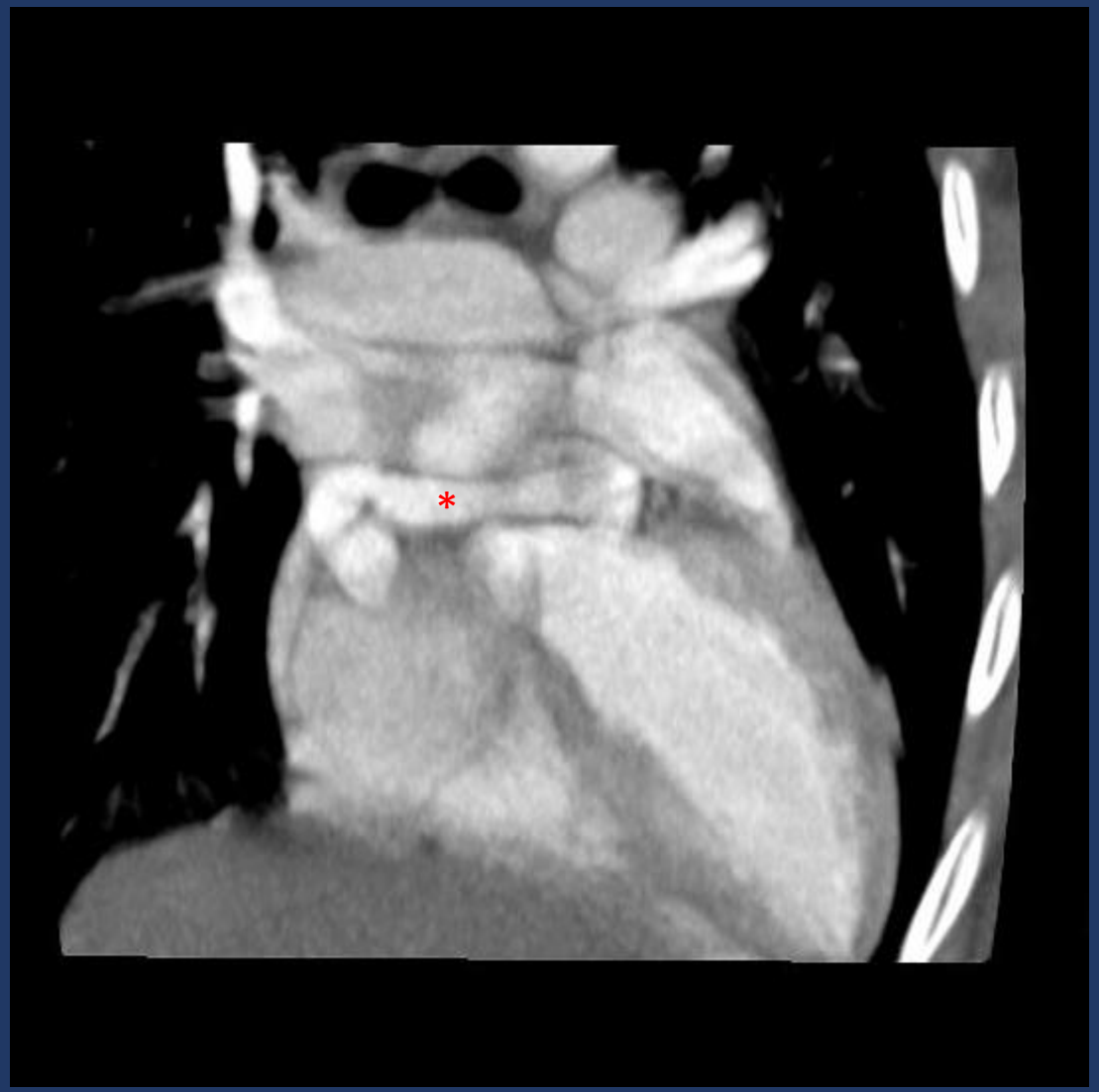
3.3MHz
M4 M4
+77.0
-77.0
cm/s
8
JPEG

CT Coronary Angiography

- Viscero-atrial situs- solitus. Cardiac situs- Levo-cardia with mild levorotation . SVC , IVC and coronary sinus draining into morphological right atrium. Superior and inferior pulmonary veins joining morphological left atrium

  The right ventricle located to right side of left ventricle .Normal atrio-ventricular concordance with tricuspid and mitral valve form morphological right and left AV connection. Superior and inferior pulmonary veins joining morphological left atrium the left ventricle.

- Ascending aorta is 1.7 cm, descending aorta is 1.2 cm in caliber. A dilated vessel is noted arising from left main coronary artery just above the left coronary sinus, coursing posteriorly in the aorto- left atrial groove and the inter- atrial groove. It measures 12 mm at ostium and 7mm in mid-portion. It is bifurcating 1.3 cm proximal to termination, with larger channel (6mm ostium) opening into right atrium 6mm below superior venacavo-atrial junction and smaller channel (5 mm ostium) terminating at superior cavo-atrial junction.LAD and LCX appear to originate from this vessel, approx. 1.1 cm from its origin.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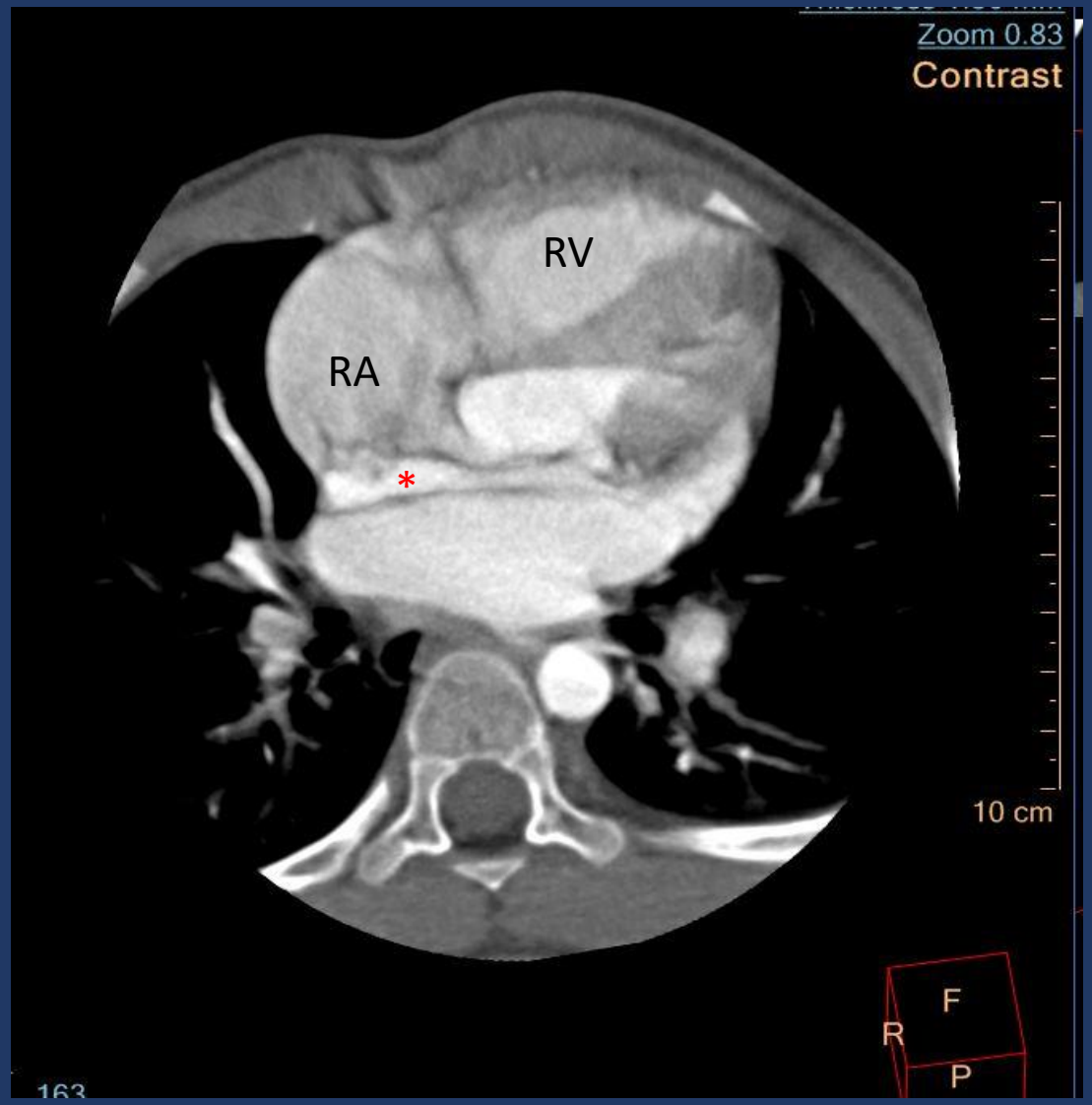
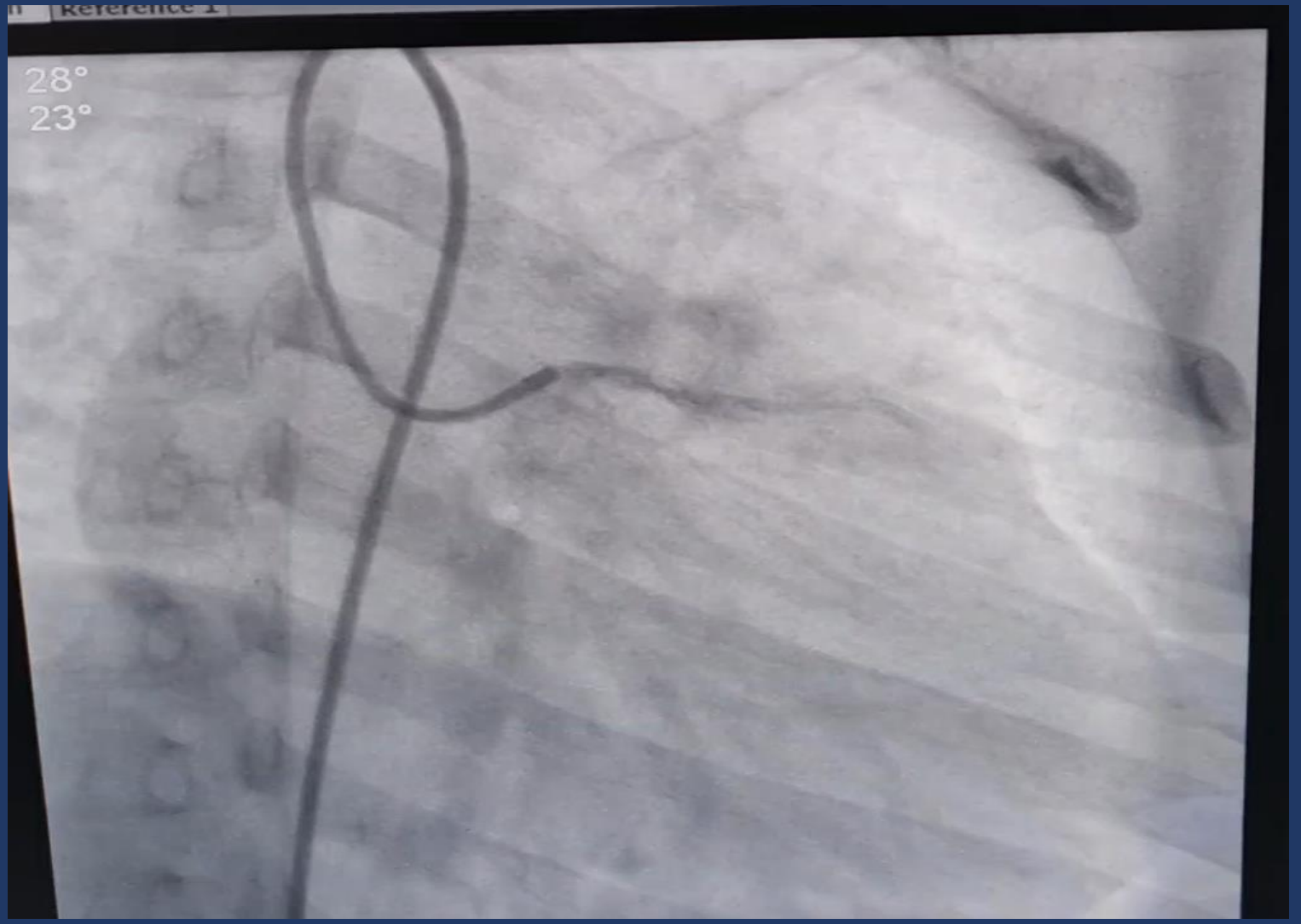

Zoom 0.83
Contrast
RV
RA
*
10 cm
F
R
P
163

Coronary Angiography & Aortography



RT & LT HEART CATHETERIZATION STUDY/OX/METRY/CARDIAC OUTPUT CALCULATIONS

Parameter	Pressure
RA	1 mmHg
RVSP	23 mmHg
RVEDP	-
PA	17/5mmHg(11)
FA	111/47 mmHg(70)
AO	100/58 mmHg (76)
LVEDP	5 mmHg

Parameter	Saturation
SO, (RA)	82 %
SO. (RV)	88 %
SO, (PA)	90 %
SO. (IVC)	80 %
SO, (SVC)	80 %
AO	98%

CALCULATED VALUES

Cardiac Output FICKS METHOD 4.52 lit/min

Cardiac Index FICKS METHOD 5.96 lit/min/m2

Stroke volume 48 ml/beat

Stroke Volume index 64 ml/ beat/M2

SHUNT CALCULATION

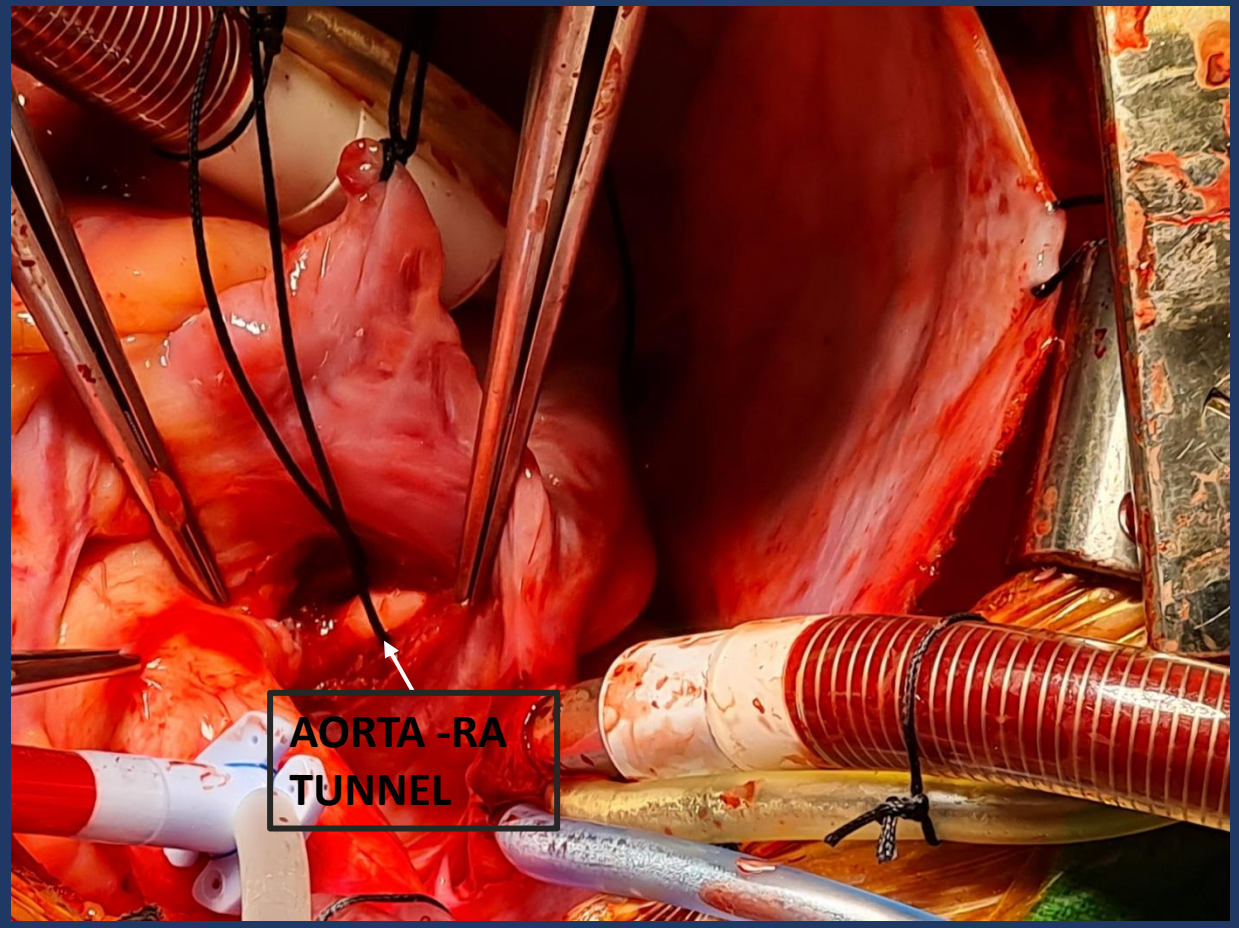
QP/QS= (SAO,- MVO,] / [PVO,- PAVO2]

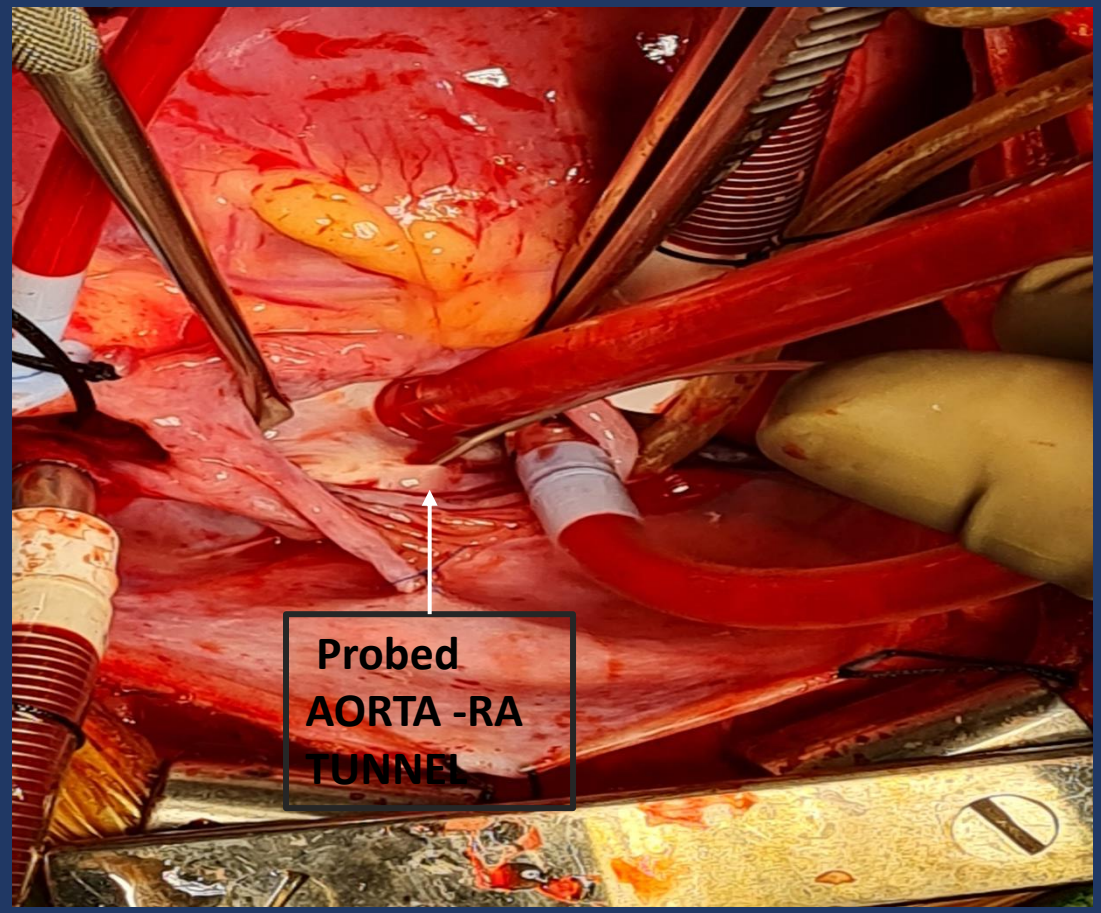
QP/QS=2.2

Intra Operative Management

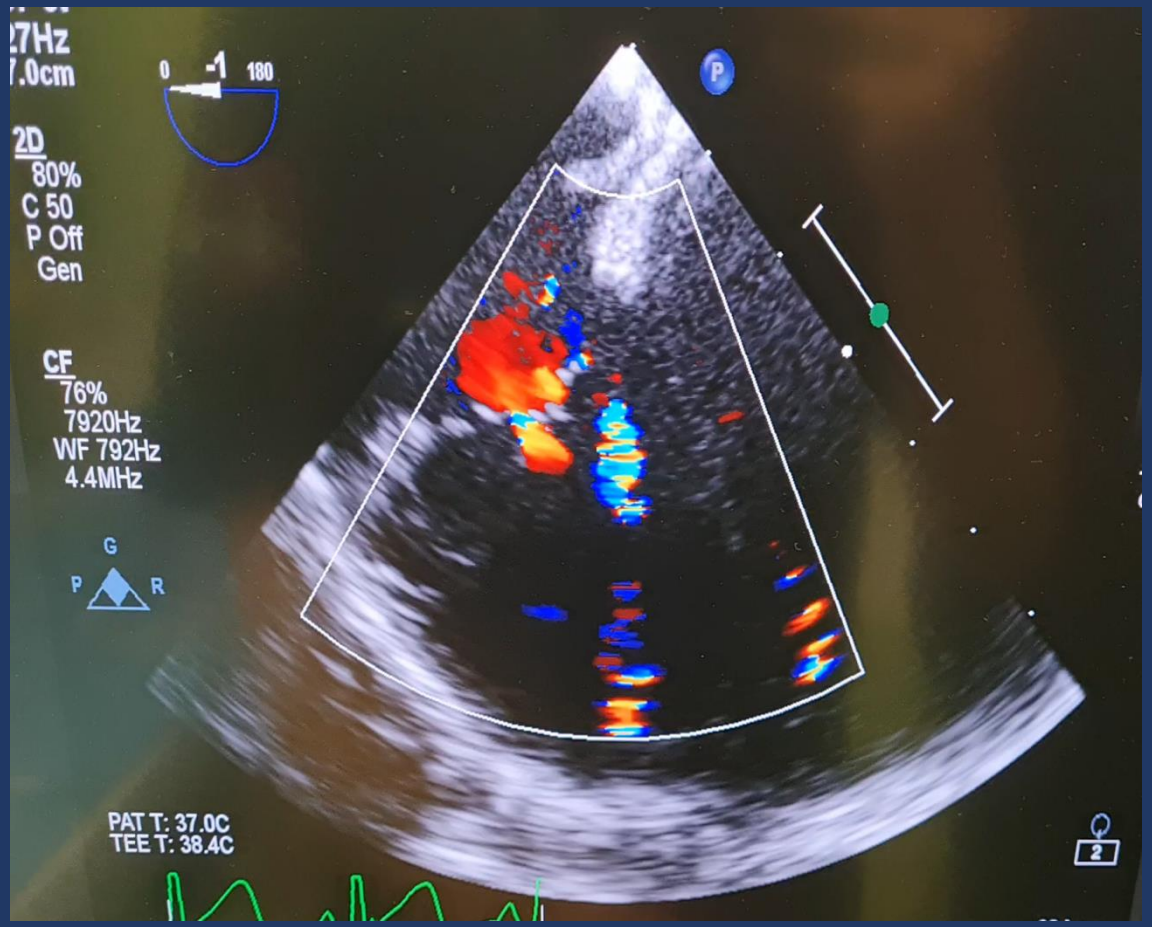
- Intraoperatively, the tunnel was seen to course along the antero-inferior aspect of the right pulmonary artery and to terminate on the posteromedial aspect of the SVC–RA junction.
- Initial cardiac arrest was achieved by means of antegrade cardioplegia. The SVC–RA junction was opened and the internal openings were identified.
- Upon exploration of the aortic root were moderate dilation of the left aortic sinus, a competent trileaflet aortic valve, and close juxtaposition of the aortic ostia of the left main coronary artery and the tunnel.
- To prevent injury to the ostium of the left coronary artery, the aortic orifice of the tunnel was left undisturbed.
- The RA opening was obliterated through direct suture approximation and the tunnel was ligated.

Intra Operative Findings





Pre OP TEE



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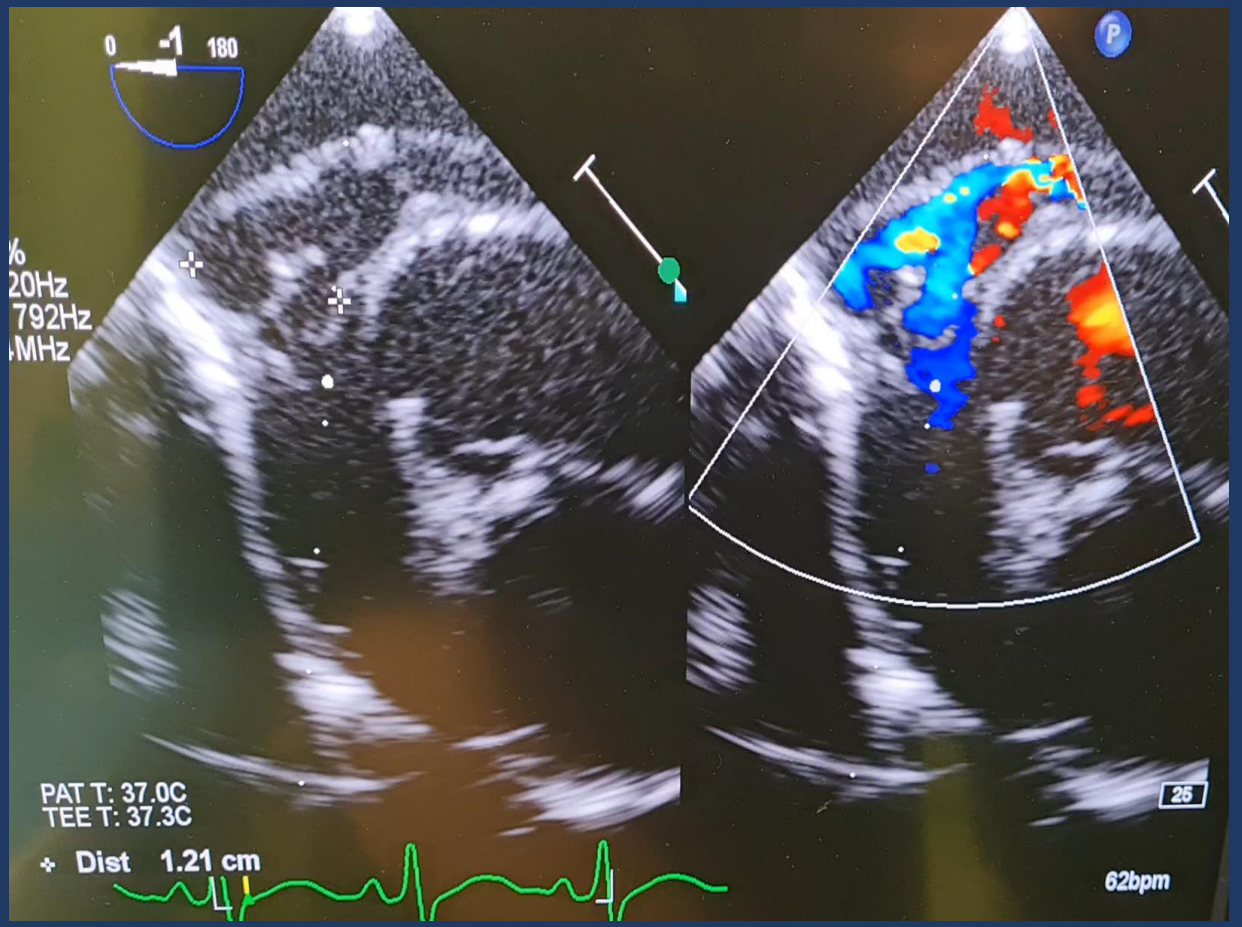

- Congenital aorta–RA tunnel, first described by Coto and colleagues in 1980 1, is an extracardiac vascular communication between one of the aortic sinuses and the RA.

- During periods of increased myocardial oxygen demand, a relatively greater fall in the resistance and subsequent dilation of the coronary arteries prevents coronary steal phenomenon through the tunnel

- Etiology
- 1)congenital deficiency of the elastic lamina in the aortic media, which gradually enlarges under the influence of high aortic pressure to form an extracardiac tunnel.
- 3) mesocardiac cystic structure arising from 5th aortic arch during early intrauterine cardiac development
- 4) supravalvular aneurysmal dilatation of primitive aorta, or an intrauterine rupture of sinus valsalva aneurysm.

- Types:
- The posterior type which originates from the left sinus valsalva is more common 4. The anterior type which has an origination from right sinus Valsalva is less common. It can also have an origination from non-coronary sinus.
- Close Differentials: Coronary-Cameral Fistula/ Sinus of Valsalva Aneurysm
- The absence of myocardial branches differentiates aorta–RA tunnel from a coronary–cameral fistula. The tunnel's origin above the supra-aortic ridge differentiates it from a sinus of Valsalva aneurysm.5
- Patients with an aorta–RA tunnel may be asymptomatic, or they may present with exertional breathlessness, palpitations, or recurrent respiratory tract infections.4

- Complications:

The continued patency of the tunnel poses these risks:

- calcification of its wall
- aortic regurgitation
- biventricular volume overload or aneurysmal expansion
- congestive heart failure
- pulmonary vascular disease
- infective endocarditis

- Treatment options

catheter-based intervention - Coil embolization.3

Surgery-ligation under controlled hypotension, or repair with the patient under CPB.

- External ligation of the tunnel close to the aorta should be performed only after accurate evaluation of the external anatomy and of the relationship between the coronary ostia and the orifice of the tunnel at the aortic end.2
- The aortic orifice may be left undisturbed in order to prevent inadvertent injury to the coronary ostia. If the coronary artery arises from the tunnel, an alternative is to reimplant the artery as a button into the aortic sinus.2.

Learning Points

- Aorta–RA tunnel is a rare lesion often detected during the evaluation of a heart murmur in an asymptomatic patient.
- Echocardiography is adequate tool for diagnosis but tomography, angiography, and aortographyis needed to rule out differential diagnosis from ruptured sinus Valsalva aneurysm or coronary cameral artery fistula.
- Multi-Modality Imaging is necessary for determination of other accompanying anomalies, exact localization of lesion, and interaction with coronary arteries which is pivotal for line of treatment selection.
- The surgical technique is usually planned taking into account the Aorta –RA Tunnel's localization, size, and interaction with coronary ostia.
- Surgery is aimed at removal of the connection between right atria, primary repair of right atrium, closing of the aortic side with patch, and if necessary reconstructing coronary ostia

References

1.Coto EO, Caffarena JM, Such M, Marques JL. Aorta—right atrial communication. Report of an unusual case. J Thorac Cardiovasc Surg 1980;80(6):941-4.

2. Rosenberg H, Williams WG, Trusler GA, Smallhorn J, Rowe RD, Moes CA, Freedom RM. Congenital aortico-right atrial communications. The dilemma of differentiation from coronary- cameral fistula. J Thorac Cardiovasc Surg 1986;91(6): 841-7.

3. Moraes F, Santos CL, Moraes CR. Aortic-right atrial tunnel. Cardiology Young 2004;14(1):86-8.

4. Gajjar T, Voleti C, Matta R, Iyer R, Dash PK, Desai N. Aortaright atrial tunnel: clinical presentation, diagnostic criteria, and surgical options. J Thorac Cardiovasc Surg 2005;130(5): 1287-92.

5. Sreedharan M, Baruah B, Dash PK. Aorta-right atrial tunnel-a novel therapeutic option. Int J Cardiol 2006;107(3):410-2.

THANK YOU