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PG Clinic

Clinical Grand Round

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Knowledge is having the right answers. Intelligence is asking the right questions. Wisdom is Knowing when to ask the right questions.
Asking 10 questions to solve a clinical conundrum

Case

A 49 year old female presented with complains of insidious onset gradually progressive dyspnea on exertion (NYHA class II) for last 2 years. She had no history of orthopnea, PND (paroxysmal nocturnal dyspnea), cough or hemoptysis. She denied having any angina on exertion, palpitation, syncope, recurrent lower respiratory tract infections in childhood or previous acute rheumatic fever. There was no history of cyanotic spells or squatting episodes in childhood.

The pulse rate was 88 beats/minute, regular, normal in volume & character, blood pressure was 134/78 mm Hg in right upper limb, supine position, respiratory rate was 18/min, thoraco-abdomial. The JVP (jugular venous pressure) was not raised, with normal wave pattern and abdominojugular reflex was absent. Cyanosis & clubbing were absent.

Cardiovascular examination revealed normal shaped precordium with no obvious bulge or pulsation. Apex beat was present in left 5th ICS @ mid-clavicluar line, was right ventricular (RV) type and a grade II left parasternal heave was noted without any thrill. On auscultation, S1 was normal, S2 was wide with fixed splitting with loud P2 component. No S3, S4 or murmur was present.

A clinical diagnosis of acyanotic congenital heart disease (ACHD), pretricuspid shunt, Ostium secundum atrial septal defect (ASD) with severe pulmonary artery hypertension (PAH) was made based on the clinical findings.

Q.1. Is it a case of Eisenmenger syndrome?
- Less likely, as the patient had no clinically evident cyanosis or clubbing. There was no history of hemoptysis or symptoms suggestive of hyper-viscosity were reported.
- However in all such cases, it is important to take resting oxygen saturation and repeat it after exercise or 6minute walk test. In the present case, the resting SpO2 on room air was 92% while after 6-minute walk test it dropped to 87%.

A 12 lead ECG and X ray chest was done (Figure 1 & Figure 2 respectively).

Q.2. What is the impression after ECG analysis?
- The ECG does not reveal any significant right ventricular hypertrophy (RVH), no qR pattern in V1 or RV strain. Hence RV pressures are likely to be sub-systemic.

Q.3. What are the voltage criteria for diagnosis of RVH in presence of RBBB in ECG?
- Voltage criteria for RVH in presence of RBBB: R of >15 mm in lead V1 in presence of complete RBBB & > 10 mm in presence of incomplete RBBB.
- Neither of these criteria are fulfilled in the current case.

Q.4. Does the chest X-ray favor Eisenmenger's syndrome?
- No; The X ray reveals increased pulmonary blood flow without any pruning, suggesting that there is still L-R shunt across the ASD. Findings favor the presence of hyperkinetic

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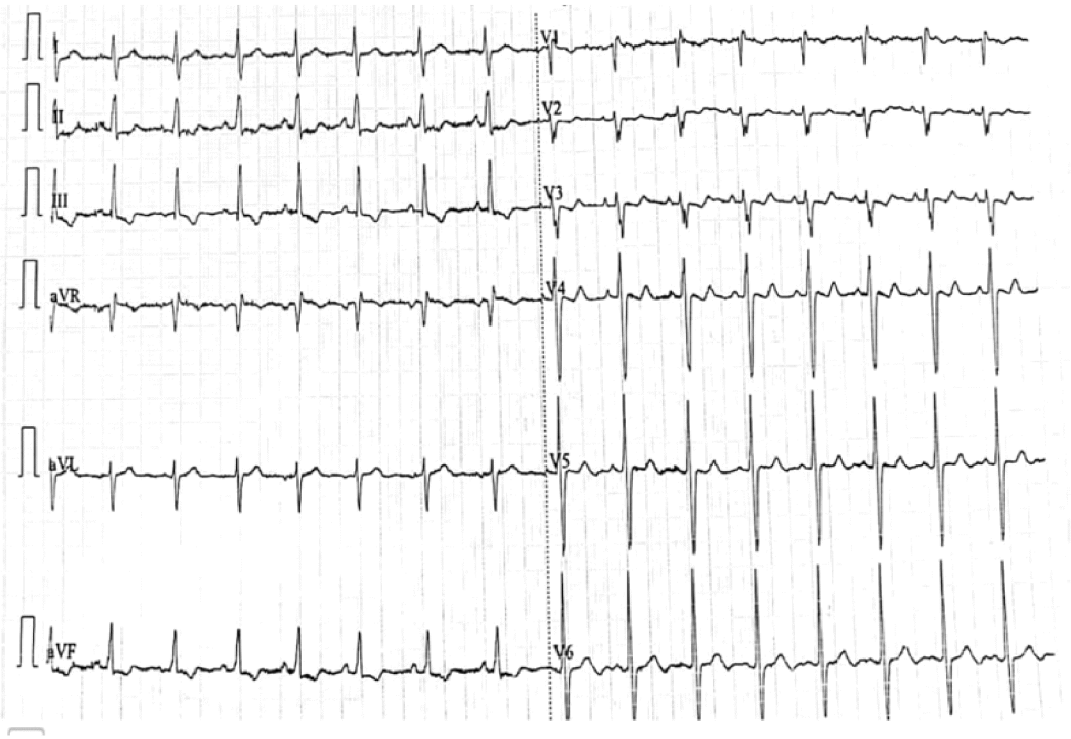

Fig. 1 : ECG showing RBBB(rsR pattern) in ead V1 with right axis deviation

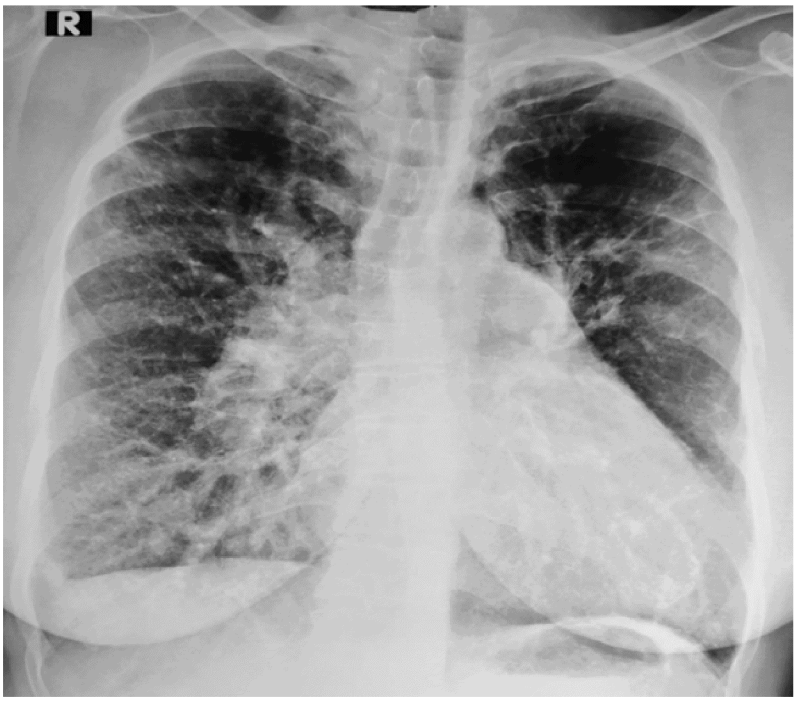

Fig. 2 : Chest X ray showing moderate card iomegaly with dilated main pu Imonary artery & right descend ing pulmonary artery, with increased pulmonary blood flow

PAH, suggesting reversibility.

A transthoracic echocardiography (Figure 3, Video 1) and agitated saline (bubble) contrast echocardiography (Figure 4, Video 2)

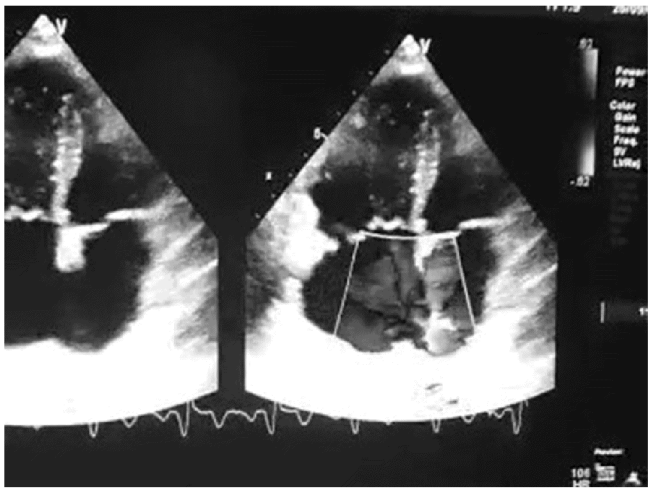

Fig. 3 : Echocardiography (apical 4 chamber view) showing large OS ASD (29 mm) with bidirectionalshuntwithdilated RA& RV.

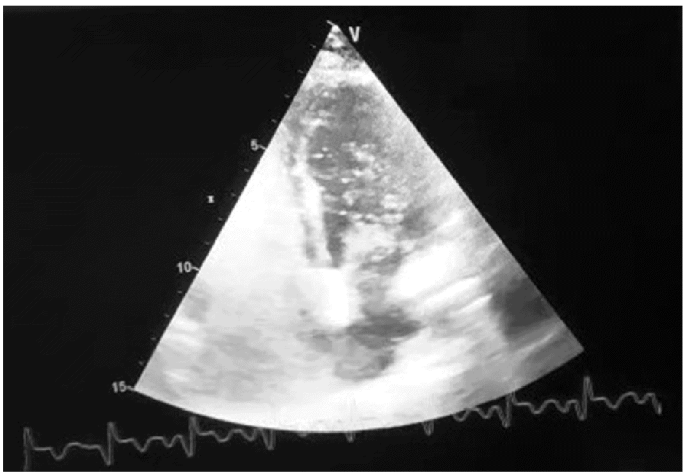

Fig. 4 : Agitated saline (bubble) contrast echocardiography- The appearance of bubble in left heart chambers immediately after the opacification of right heart chambers within 3 cardiac cycles suggest the presence right to left shunt at atrial level.

- There is usually only a relatively weak correlation between pulmonary artery systolic pressure estimated from TR velocity by Doppler Echocardiography & pulmonary artery pressure measured by right heart catheterization (RHC) in presence of ASD. Doppler Echocardiography can overestimate pulmonary artery systolic pressure determined from TR velocity in presence of ASD due to RV volume overload & pulmonary over circulation. In this case echocardiography showed moderate TR with predicted RVSP of 72 mm Hg.

Q.6. Is it true that appearance of bubbles in left heart chambers on agitated saline (bubble) contrast echocardiography is always due to intracardiac right to left shunt?

- No; the timing of appearance of bubbles in left heart chambers is important
- Appearance of bubbles within 3 cardiac cycles after the opacification of right heart chambers on agitated saline (bubble) contrast echocardiography is suggestive of intracardiac right to left shunt. Delayed appearance of bubbles in likely due to intrapulmonary shunt (Pulmonary AV malformations or fistula).

Q.5. Could there be a discrepancy between RV pressure as measured by echo and that on cath in such cases?

Q.7. How does one approach further?

- Since this patient had mild resting hypoxemia which further worsened on exercise, R-L shunt needs to be rule out in this case. The echocardiography clearly confirms a large ostium secundum ASD with bidirectional shunt. Agitated contrast saline is also a useful maneuver in patients where one suspects a R-L shunt. In view of the X ray findings of increased pulmonary blood flow and echo suggesting a bidirectional shunt, it is advisable

	Saturation (%) (Pre O2)	Pressure (mmHg) (Pre O2)	Saturation (%) (Post O2)	Pressure (mmHg) (Post O2
SVC	74		85	
IVC	78		88	
RA	87	10/12/8	90	9/11/8
RV	86	46/0/9	94.7	49/0/10
PA	81	47/21/29	95.3	43/18/26
PCWP	94		95.3	
PV	94		99.4	
LA	91.5	11/12/9	98.8	9/12/8
LV	93	140/0/3	99	139/0/7
AO	92.4	142/72/97	98.6	140/70/96

Fig. 5 : Oximetry and pressure data

to perform a cardiac catheterization study to assess RV pressures, net left to right or right to left shunt and operability.

A Cardiac catheterization study was done and the data is summarized in Figure 5 and 6. This revealed a step up of 12% at RA with a systemic desaturation (Ao saturation 92.5). The RV pressure was 46 mm Hg with mild PAH and no gradient across RVOT.

Q.8. What are the possible causes of systemic arterial desaturation in this case?

- The most obvious cause for systemic arterial desaturation in the present case could be ASD with bidirectional shunt due to pulmonary artery hypertension.
- However a closer look at the saturation data reveals that LV, LA and PV are all desaturated to an equal extent.

Q 9. How does this data help us?

- This tells us that desaturation is actually occurring further upstream and the possible causes could be
  - Associated persistent left superior vena cava (LSVC) draining into LA
  - Presence of unroofed coronary sinus & associated persistent LSVC
  - Prominent eustachian valve with selective streaming of IVC blood to LA
  - TAPVC (total anomalous pulmonary venous connection)

  All the above causes would lead to desaturation in LA with normal saturation in PV. Since even the PV has desaturation, cause of this would be even further upstream ie. due to
  - Presence of Pulmonary AV malformations or fistula

Parameters		Pre O2	Post O2
Qp (L/min)	$\frac{O_2 \text{ consumption (mL/min)}}{PV\ O_2 - PA\ O_2 \text{ (content in ml/L)}}$	7.24	22.95
Qs (L/min)	$\frac{O_2 \text{ consumption (mL/min)}}{SA\ O_2 - MV\ O_2 \text{ (content in ml/L)}}$	5.42	7.35
Qp/Qs		1.34	3.12
PVR (WU)	$\frac{(PA_{mean}) - (LA_{mean})}{Qp}$	2.76	0.78
PVRI	PVR X BSA	4.33	1.23
SVR (WU)	$\frac{(Ao_{mean}) - (RA_{mean})}{Qs}$	16.79	11.97
SVRI	SVR X BSA	26.36	18.79
PVR/SVR		0.164	0.065

Fig. 6 : Shunt Calculations, PVRI & PVR/SVR ratio

  - Lung diseases like COPD (chronic obstructive pulmonary disease) or emphysema, Bronchial asthma, Bronchiolitis etc.
  - Diagnosis of lung disease as the cause of systemic desaturation is confirmed by the fact that pulmonary vein & all chambers distal to it (LA, LV) and Aorta are all desaturated almost equally, and this gets corrected post oxygenation. (Figure 6)

Q10. What further investigation is needed?

- HRCT chest & PFT (pulmonary function test) with DLCO (Diffusion lung capacity with carbon monoxide) should be done.
- A HRCT chest was done which demonstrated fibroatelectatic changes in bilateral upper lobes (left>right) (Fig 7)

The final diagnosis was large ostium secundum ASD, mild PAH. Since there was a net left to right shunt of >1.5 :1, with PVRI < 5 WU m^2, & PVR/SVR ratio of <0.3, this patient is considered operable.

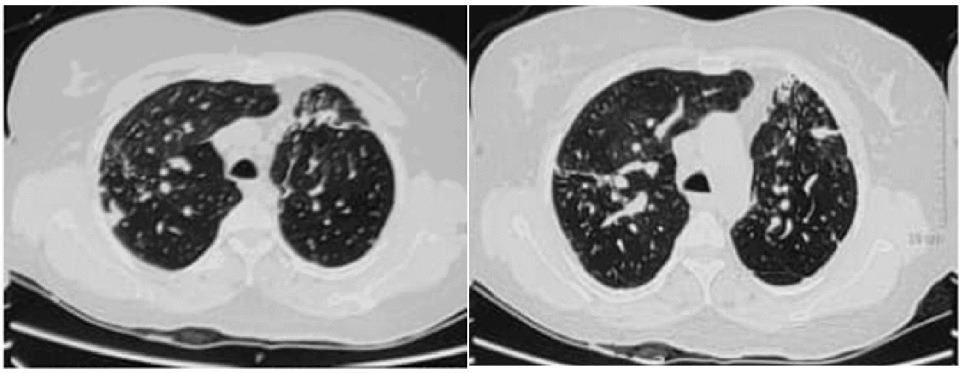

Fig. 7 : A HRCT chest demonstrated fibroatelectatic changes (arrows) in bilateral upper lobes (left>right)