

Case of Recurrent Syncope in young Girl

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Abstract: Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital heart defect associated with multiple cardiac morphological abnormalities and conduction defects. It occurs in approximately 0.5 to 1.4% of all congenital heart diseases (CHD).¹ The prognosis rests on clinical presentation, progression of disease and the effect of systemic pressure on the functional Ventricle. We report a case of 15yr girl who was admitted with complaints of recurrent syncope since 1 month. After further investigation, she was diagnosed as a case of Congenitally corrected transposition of the great arteries (CCTGA) with complete heart block (CHB).

Keywords: Congenitally corrected transposition of the great arteries (CCTGA), Transthoracic Echocardiography (TTE), Congenital Heart Disease (CHD), Electrocardiogram (ECG), Complete heart block (CHB).

Introduction

CCTGA, is a complex heart defect. It may be present with or without associated anomalies. Merely 10% of the patients with CCTGA do not have any associated anomalies like ventricular septal defect, pulmonary artery stenosis, tricuspid valve abnormalities, and mitral valve abnormalities.^(2,3) The abnormal positioning of the conduction system increases the risk for fibrosis of the conduction system with a progressive incidence of complete atrioventricular (AV) block of roughly 2% per year.⁴ A 15yr girl presented with complaints of recurrent syncope of 1 month duration. ECG showed complete heart block, absence of q waves in lead v5 & v6, Q wave in lead v1 suggesting reversal of septal depolarization. TTE study showed AV & VA discordance suggesting congenitally corrected Transposition of Great Arteries.

Case

A 15yr girl was admitted with complaints of recurrent syncope since 1 month. On examination her weight was 50kg, Height of 150cm. Pulse -40/min regular, BP-100/70mmHg, spo2-98%. JVP showed cannon A waves. Cardiovascular examination revealed loud second heart sound. No murmur. Other system examination were unremarkable.

Electrocardiogram (Fig.1) showed heart rate of 40, Narrow QRS complexes, AV dissociation suggesting complete heart block, absence of q waves in lead v5 & v6, Q wave in lead v1 suggesting reversal of septal depolarization. Chest Xray PA view (Fig.2) showed situs solitus, levocardia, cardiomegaly and left heart border to appear more vertical than usual. Transthoracic echocardiography showed -Situs Solitus (Fig.3), Levocardia (Fig.4), L looped Ventricles & AV discordance (Fig.5) & VA discordance suggesting Congenitally corrected Transposition of Great Arteries. Morphological Left Ventricle to right and morphological Right Ventricle to the left. There were L-transposed great arteries (Fig.6). There was no Ventricular Septal Defect, no pulmonary stenosis (Fig.7). There was small jet of tricuspid regurgitation (Fig.8). Good Right Ventricular function (Fig.9{a & b}) and Good Left Ventricular function.

Electrocardiogram

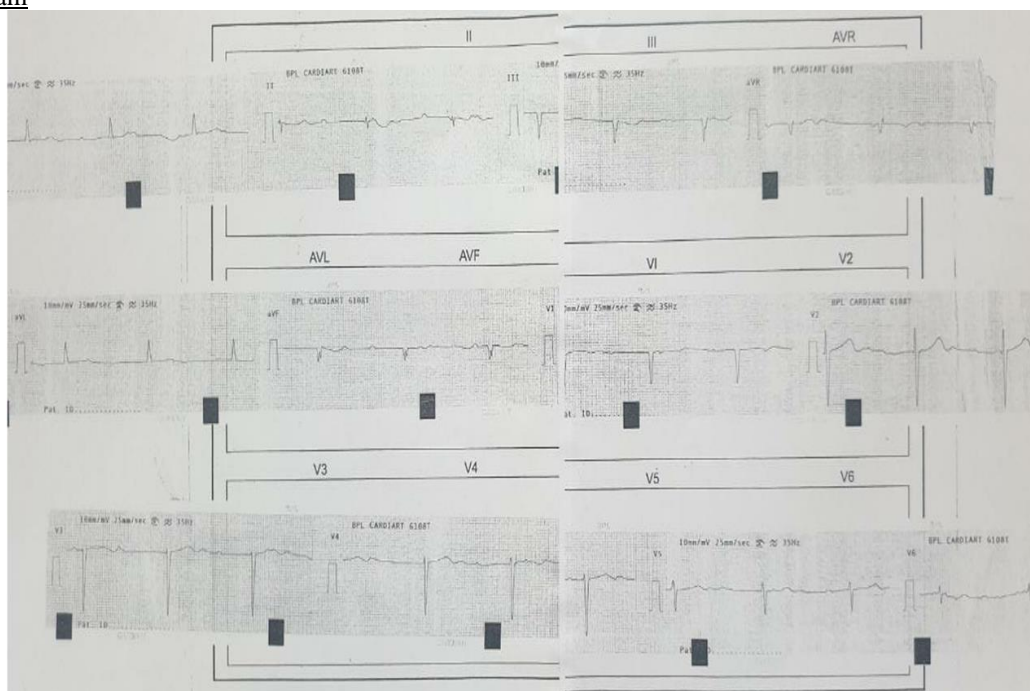


Fig 1:Heart rate of 40, Narrow QRS complexes, AV dissociation suggesting complete heart block, absence of q waves in lead v5 & v6, Q wave in lead v1 suggesting reversal of septal depolarization.

Chest X ray PA View

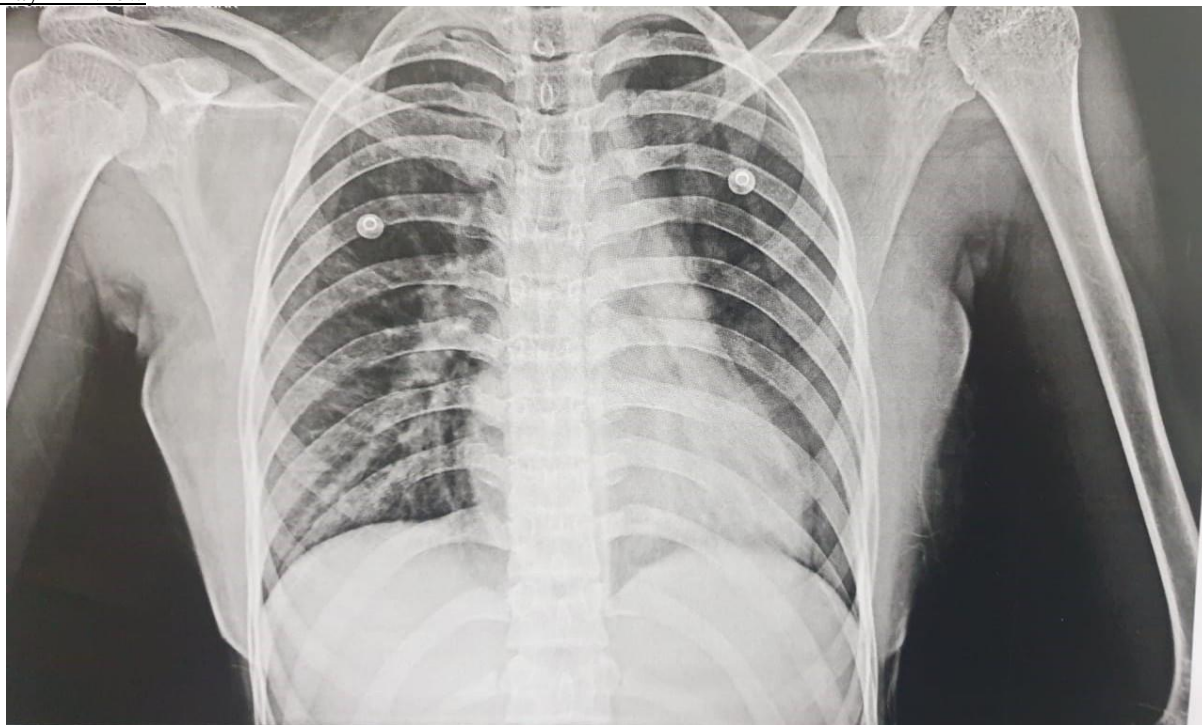


Fig 2:Situs solitus, levocardia, Cardiomegaly and left heart border to appear more vertical than usual.

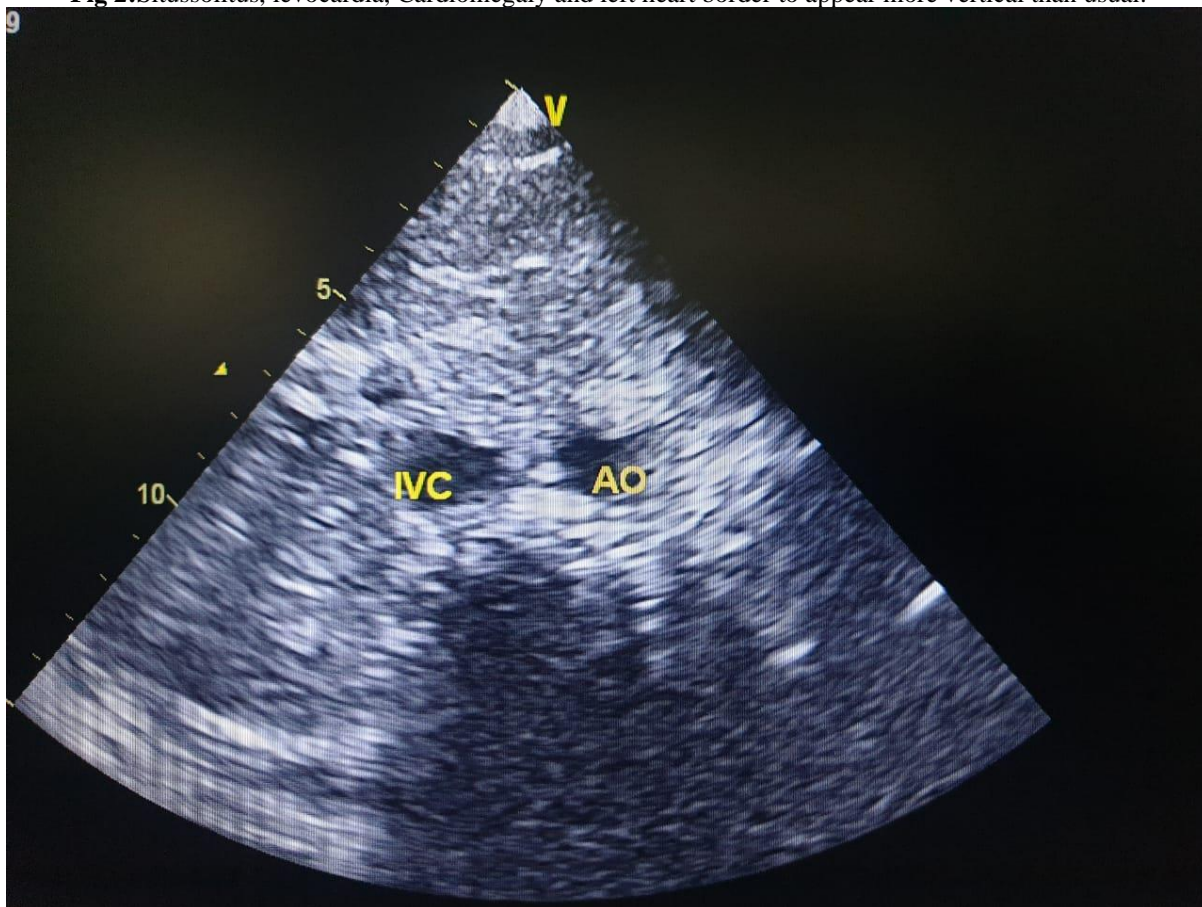


Fig 3:TTE subcostal view showing Situs solitus. Inferior vena cava on right & Descending aorta on left of spine.

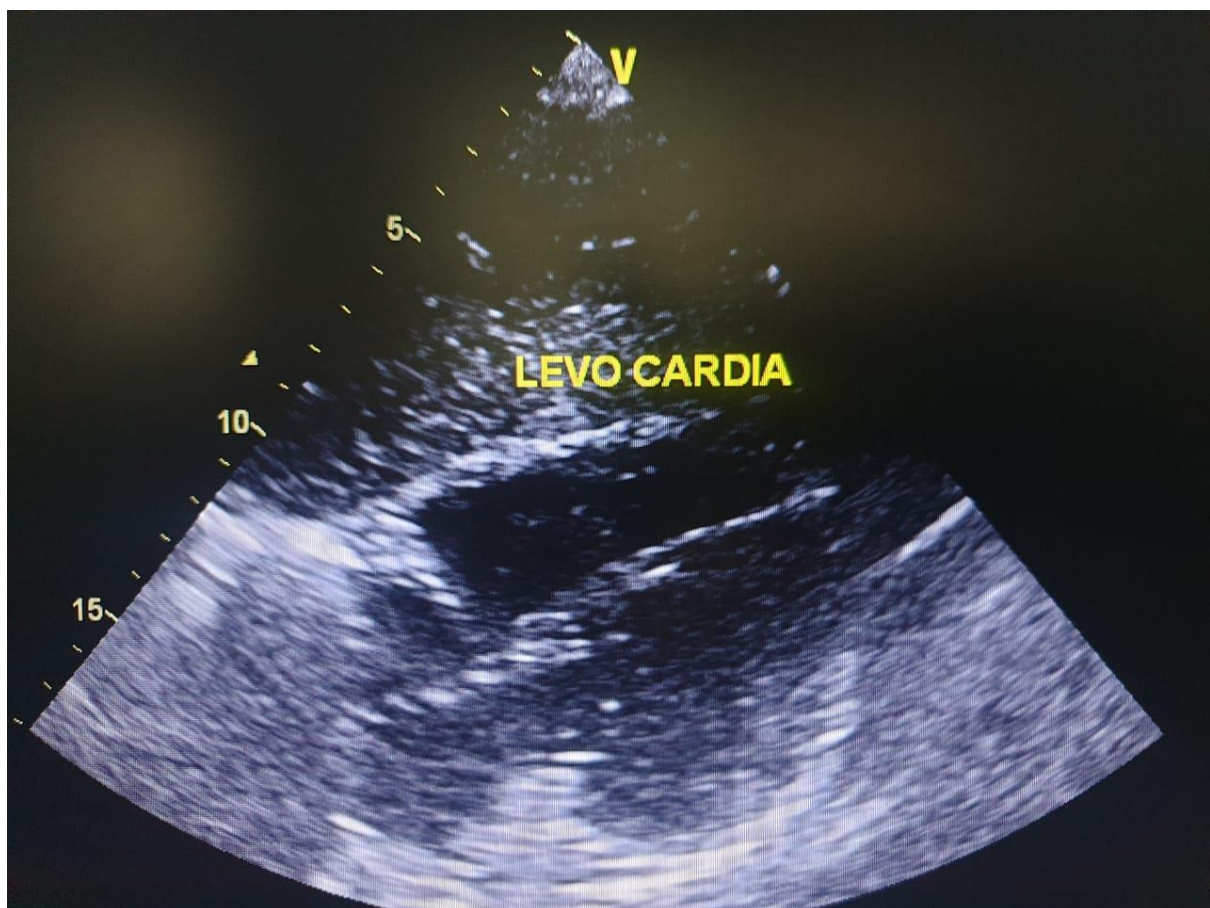


Fig 4:TTE Subcostal view showing Levocardia.

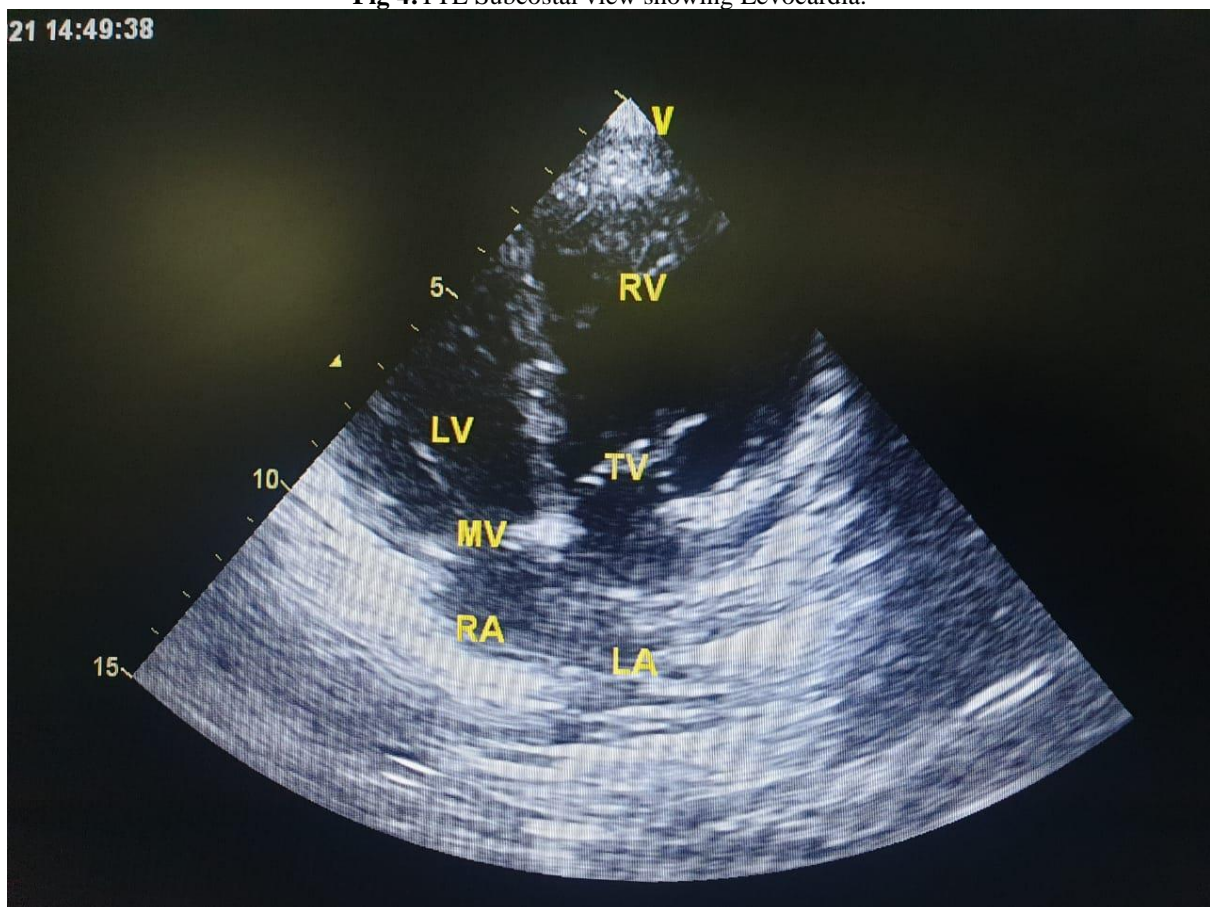


Fig 5:TTE apical 4chamber view showing AV discordance.



Fig 6:TTE short axis view showing L posed Aorta.

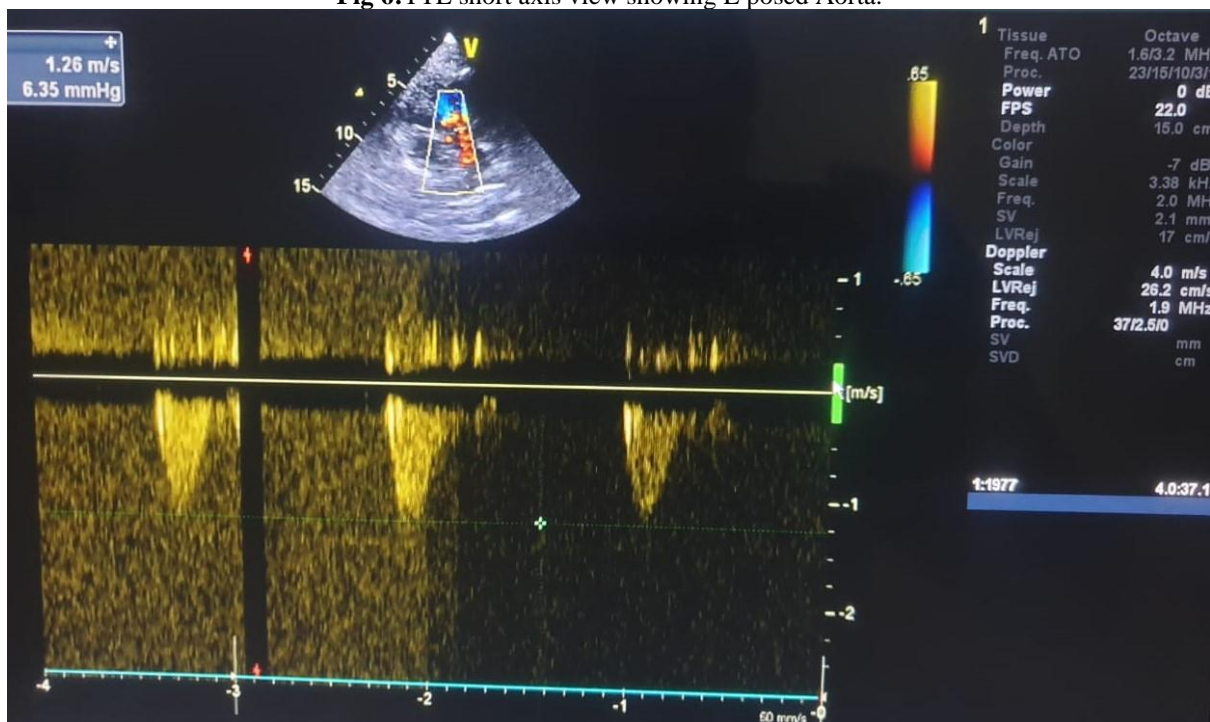


Fig 7:TTE showing No evidence of Pulmonary stenosis.



Fig 8:TTE Apical 4 chamber view showing small Tricuspid Regurgitation

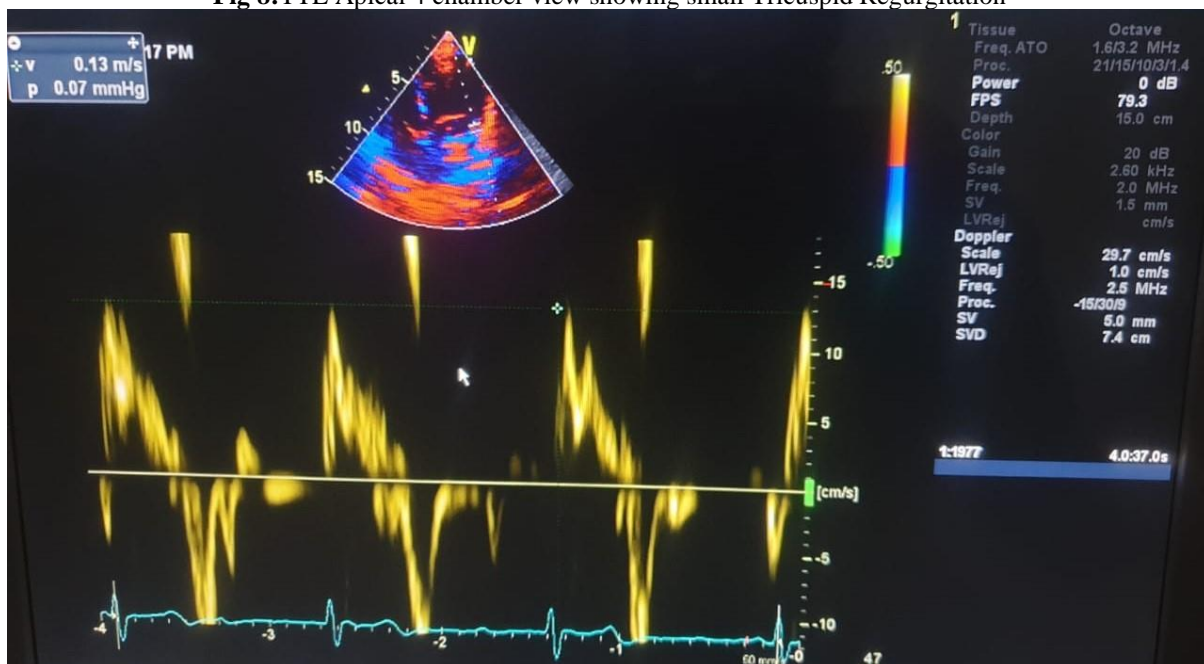


Fig 9a:TTE Apical 4 chamber view showing RV S¹ of 13cm suggesting Normal RV function.

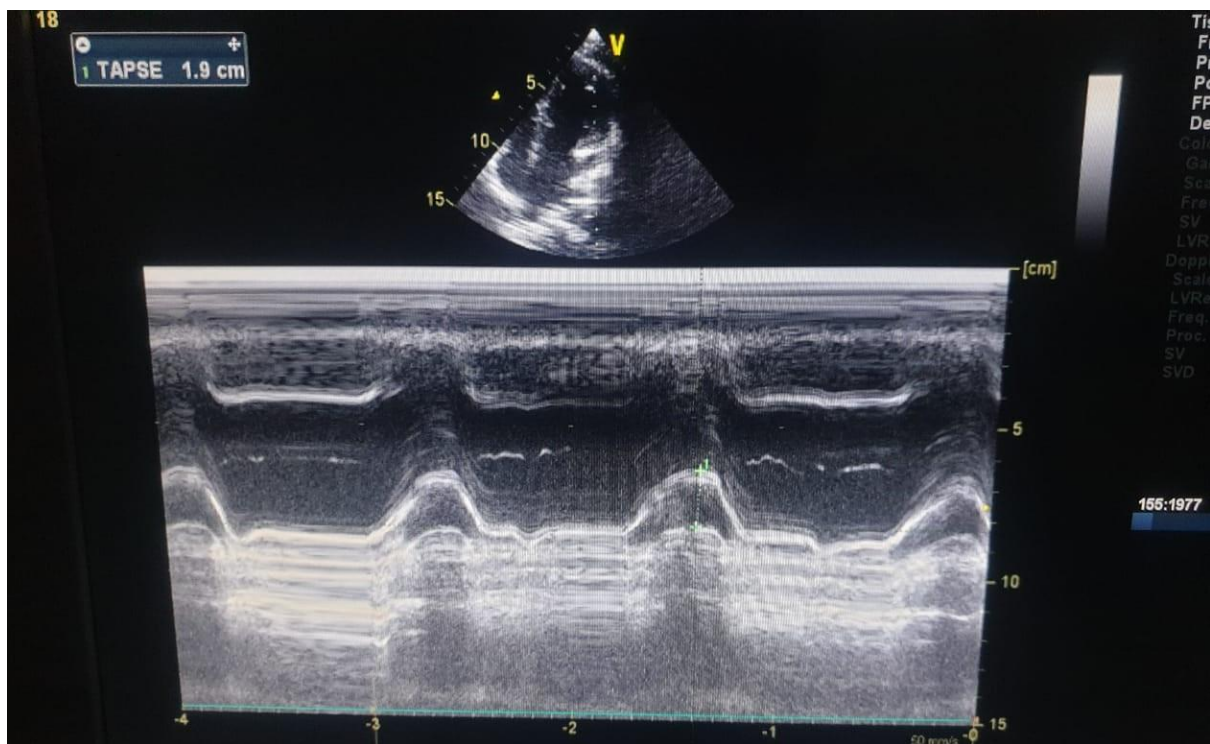


Fig 9b:TTE Apical 4 chamber view showing TAPSE of 19mm suggesting Normal RV Function.

Discussion

CCTGA is characterized by AV and ventriculoatrial discordance. The aorta is located closer to the anterior and more to the left than the pulmonary artery. The AV valves follow their respective ventricles. Because of the displacement of the AV node and the abnormal course of conduction tissue, there is an increased risk of spontaneous complete AV block.^[5,6] Our patient was asymptomatic till 15 years of age until 1 month back when she started to have recurrent syncope. Cause of recurrent syncope was CHB. Meticulously performing segmental analysis protocol lead us to the diagnosis of CCTGA.

Conclusion

CCTGA is a rare cardiac anomaly, difficult to diagnose unless structured segmental analysis protocol is followed. Echo diagnosis rests on demonstrating abnormal Atrioventricular & Ventriculoarterial connections. We should always look meticulously for trio of major associated abnormalities. (VSD, PS & Left AV Valve Regurgitation). Forgotten ventricle is key for survival, success of therapy & quality of life hence thorough Assessment of systemic RV performance is key. Lifelong follow up of patient is required as risk of CHB is 2% every year.

References

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