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Case Study

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CASE REPORT: A CASE OF CONGENITALLY CORRECTED TRANSPOSITION OF GREAT ARTERIES (CC-TGA) WITH EBSTEIN'S ANOMALY IN A CASE OF ATRIAL FIBRILLATION

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INTRODUCTION

Congenitally corrected Transposition of Great Arteries (cc-TGA), also known as looped-TGA (Transposition of Great Arteries) is an extremely rare clinical manifestation of cyanotic heart disease, accounting for 0.5-1% of all the cases of congenital heart diseases while Ebstein's Anomaly accounts for <1% of all congenital heart diseases worldwide. Ebstein's Anomaly with cc-TGA is seen in about <0.5% of cases of all congenital heart diseases in the world. The incompetent morphological right ventricle is faced with an enormous workload and eventually, heart failure occurs in these patients if ccTGA remains undiagnosed and untreated.

CASE DESCRIPTION

A 45-year-old Indian female presented with chief complaints of shortness of breath, palpitations since 5 months, and frequent syncopal attacks for the past one month.

The patient was apparently alright 6 months back when she complained of breathlessness which was more on exertion (NYHA grade II), with no history of orthopnea or PND, or palpitations. She also complained of frequent syncopal attacks which were more whenever she used to exert.

She does not have any comorbidities like DM/HTN/IHD/CVA etc.

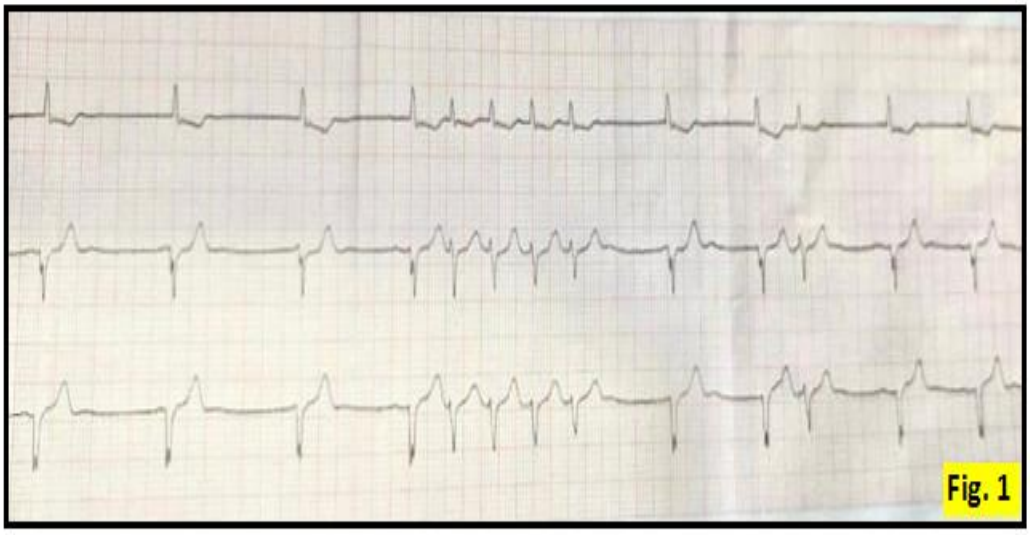
On examination, the patient was conscious, cooperative, and oriented to time, place, and person. She was hypotensive with a blood pressure of 80/60 mm Hg and a pulse rate of 64 beats per minute which was irregularly irregular in rhythm maintaining saturation at 98% on room air.

The patient is a known case of Atrial Fibrillation diagnosed 2 years back and treated for the same with Inj. Amiodarone.

INVESTIGATIONS

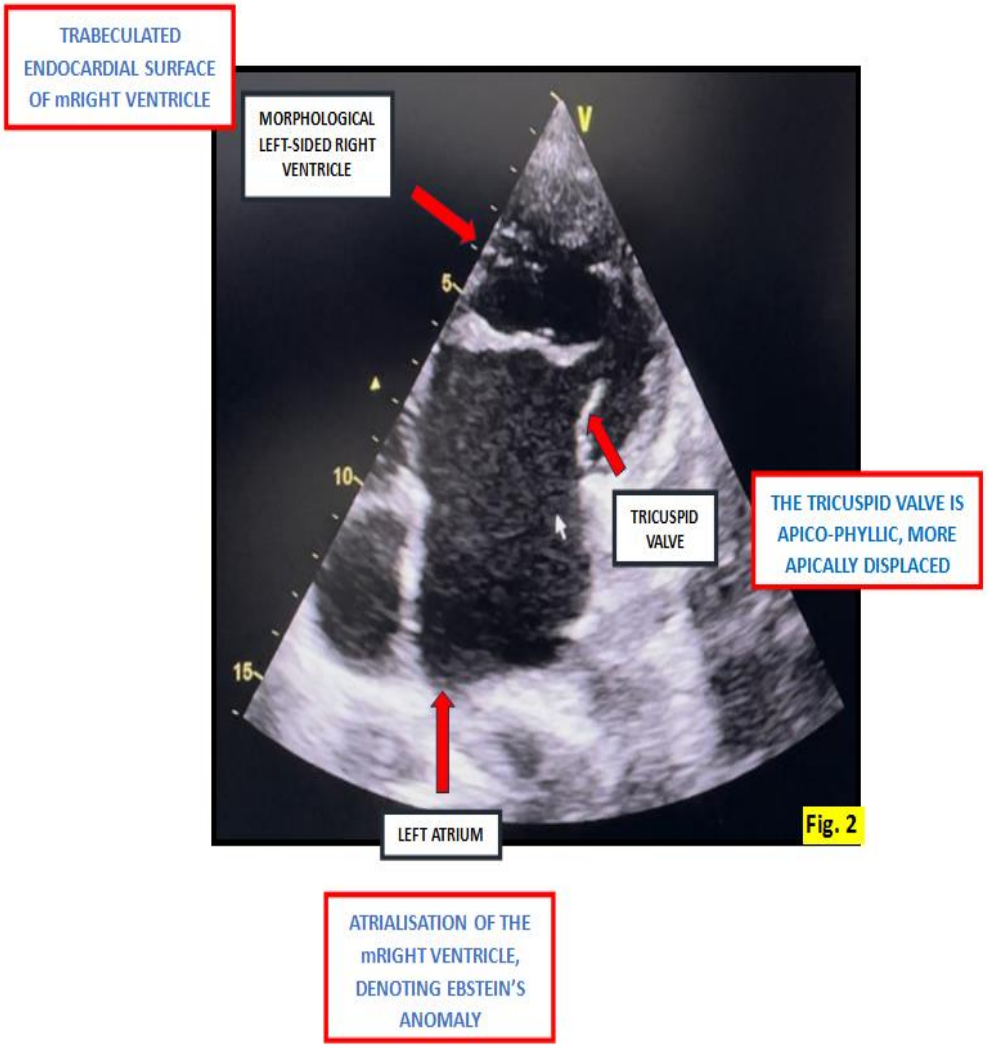
Investigation Modality	**FINDINGS**
Electrocardiography	Atrial fibrillation with Sick Sinus syndrome
2 Dimensional Echocardiography	• Atrio-ventricular valve (A-V) discordance and Ventriculo-arterial(V-A) discordance with situs solitus • Ventricular inversion with Ebstein's anomaly of left atrio-ventricular valve with right ventricle as systemic ventricle • Mild Tricuspid regurgitation was noted.
Chest X-Ray (P-A View)	Egg on a string appearance

1. ELECTROCARDIOGRAPHY (ECG) FINDINGS

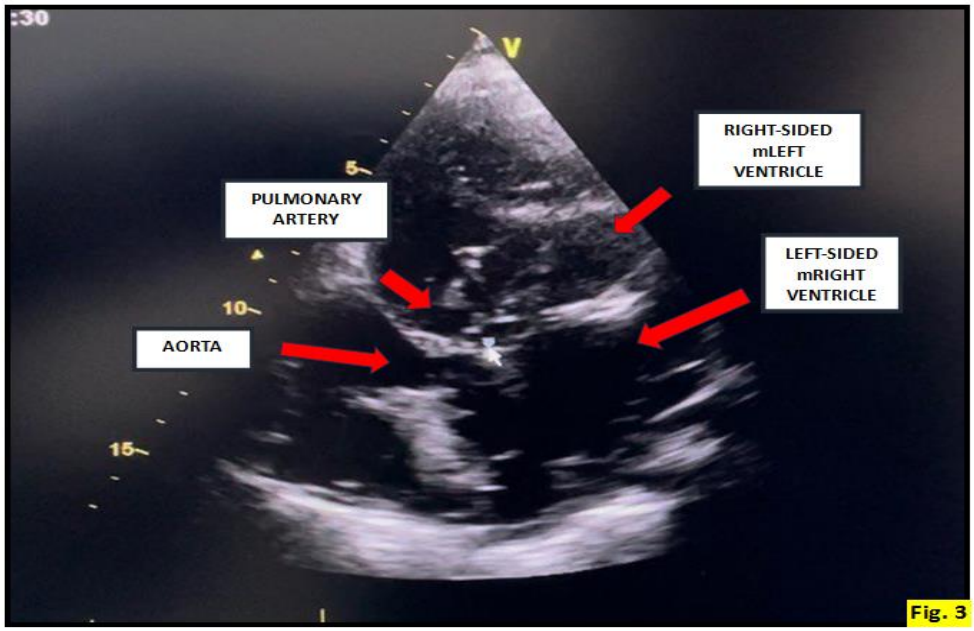


The ECG strip shows an irregularly irregular rhythm of Atrial fibrillation alternating with bradycardia denoting Tachy-Brady Syndrome which is also known as Sick-Sinus Syndrome.

2-DIMENSIONAL ECHOCARDIOGRAPHY LONG AXIS 4-CHAMBER VIEW

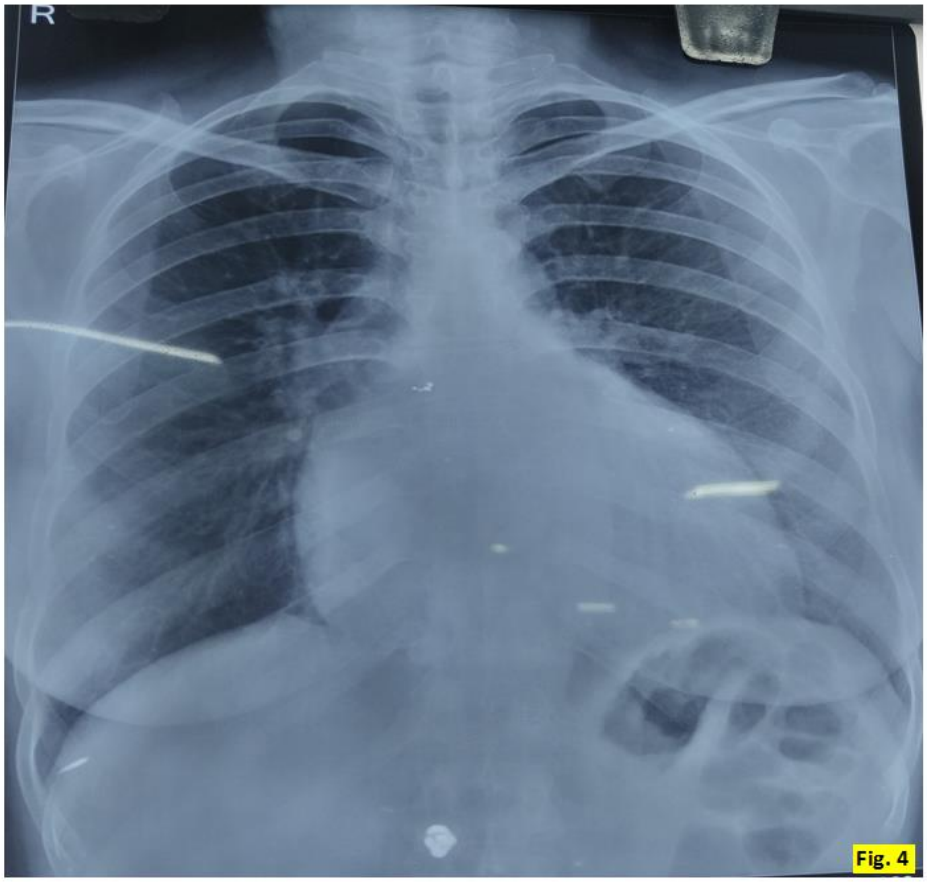


APICAL 5 CHAMBER VIEW



- PARALLEL ARRANGEMENT OF THE GREAT ARTERIES
- AORTA ORIGINATING FROM LEFT-SIDED mRIGHT VENTRICLE AND PULMONARY ARTERY ORIGINATING FROM RIGHT-SIDED mLEFT VENTRICLE – DENOTING VENTRICULO-ARTERIAL(VA) DISCORDANCE

CHEST X-RAY (P-A VIEW)



Classical EGG ON A STRING APPEARANCE suggestive of Transposition of Great Arteries.

DISCUSSION

Congenitally corrected transposition of Great arteries(cc-TGA) also called levotransposition of great arteries was first reported by Rokitansky in 1875.[1] cc-TGA accounts for 0.5-1% of all the cases of congenital heart diseases while Ebstein's Anomaly accounts for <1% of all congenital heart diseases all over the world. Ebstein's Anomaly with cc-TGA is seen in about <0.5% of cases of all congenital heart diseases worldwide.[2] Since the identification of the abnormal position of ventricles and associated AV valves can be missed, diagnosis of cc-TGA becomes a challenge in cardiology.[3] In cc-TGA, the ventricles are L-looped. This means there is abnormal leftward movement of the primitive heart resulting in abnormal positioning of the ventricles, the morphological right ventricle is placed on the left, and the morphological left ventricle is placed on the right. AV-discordance means abnormal connection between the atria and the ventricles. The morphological right atrium is connected to the morphologic right-sided left ventricle and the morphological left atrium is connected to the morphologic left-sided right ventricle. VA discordance is defined as the pulmonary artery (RIGHT-SIDED GREAT ARTERY) being connected to the right-sided morphologic left ventricle and the aorta (LEFT-SIDED GREAT ARTERY) being connected to the morphologic left sided right ventricle. Because there are discordant connections at both levels, the circulation of blood flow remains hemodynamically correct, hence the term Congenitally Corrected -TGA. cc-TGA can manifest alone or in combination with other forms of congenital heart disease. The anomalous location of the atrioventricular (AV) node along with the AV bundle leads to an abnormal cardiac conduction system and hence arrhythmias in these patients occur. In our case, cc-TGA was associated with Ebstein's Anomaly. When the tricuspid valve is abnormally formed, there is leakage or backflow of blood from the right ventricle into the left atria resulting in Tricuspid Regurgitation. The prognosis is good, however, patients can commonly survive in adulthood with a variety of clinical sequelae. due to morphological R-V and delicate tricuspid valve supporting the systemic circulation as a result of A-V and V-A discordance. The longstanding systemic pressure overload onto the morphological RV leads to mRV hypertrophy as a compensatory mechanism which will eventually fail resulting in cardiac failure. The mainstay of treatment is to reduce the afterload on R-V and improve R-V dysfunction using Angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, and beta blockers, also Digoxin or anticoagulants like warfarin may be used for arrhythmias. The best intervention for cc-TGA is surgical correction with a double-switch operation (DSO) but it eventually leads to heart failure. As for our patient, no decision for surgical intervention has taken place as of now. One of the Holter ECG monitoring reports of our patient resulted in long pauses which is suggestive of Sick Sinus Syndrome. At present, our patient is posted for Dual Chamber PPI(permanent pacemaker implantation).

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