

## Levo-Transposition of Great Arteries Presented with Complete Heart Block in Young Age: A Case Report

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### Abstract

Levo-transposition of great arteries (l-TGA) is an atypical congenital malformation of heart characterized by inversion of ventricles along with associated atrioventricular valves. In this article, a case of 38 years old male, with no known comorbidities, is described who presented with complete heart block. No other associated cardiac incongruity was found. Transthoracic echocardiography revealed tricuspid valve regurgitation. A permanent pacemaker was implanted which resulted in alleviation of patient's symptoms. No complications were developed and patient was discharged after observation.

**Keywords:** Levo-Transposition of Great Arteries; Complete Heart Block; Permanent Pacemaker; Tricuspid Regurgitation

### Introduction

Congenitally corrected transposition of great arteries (CCTGA), also known as levo-transposition of great arteries (l-TGA) is an uncommon congenital cardiac malformation. L-TGA constitutes 0.5 - 1% of all congenital cardiac lesions with prevalence of 0.03 per 1000 live births [1,2]. L-TGA is an acyanotic heart disease characterized by double discordance i.e. atrioventricular and ventriculoarterial discordance. Deoxygenated systemic blood enters the anatomically correct right atrium from pulmonary artery and then to discordant left ventricle (functional right ventricle) via mitral valve [3] whereby, the deoxygenated blood returns to the systemic circulation without passing through the lungs. After oxygenation, blood empties into anatomically correct left atrium through pulmonary veins and then to discordant right ventricle (functional LV) via tricuspid valve. This oxygenated blood returns to the lungs instead of entering the systemic circulation. Blood from discordant right ventricle then enters the systemic circulation via discordant aorta in a different route as compared to the normal blood circulation through the lungs or from the lungs. Despite the adequate circulations, both systemic and pulmonary, there is an increased propensity of complete Atrioventricular (AV) block and recurrent reentry tachycardia attributed to abnormal location and function of AV node and Bundle of His [4]. This report presents anomalous case of patient with l-TGA, presented as complete heart block in exceptionally young age and its consequent management. Furthermore, it also highlights the difference in age of presentation of l-TGA from existing literature which shows the condition to come to attention in old age.

### Case Report

A 38 years old male, unknown comorbidities, presented to our hospital with complaint of 'sinking of heart feeling' along with dizziness for 1 day. Patient had occasional similar complaints since childhood however no medical help was sought due to mild symptoms. Past

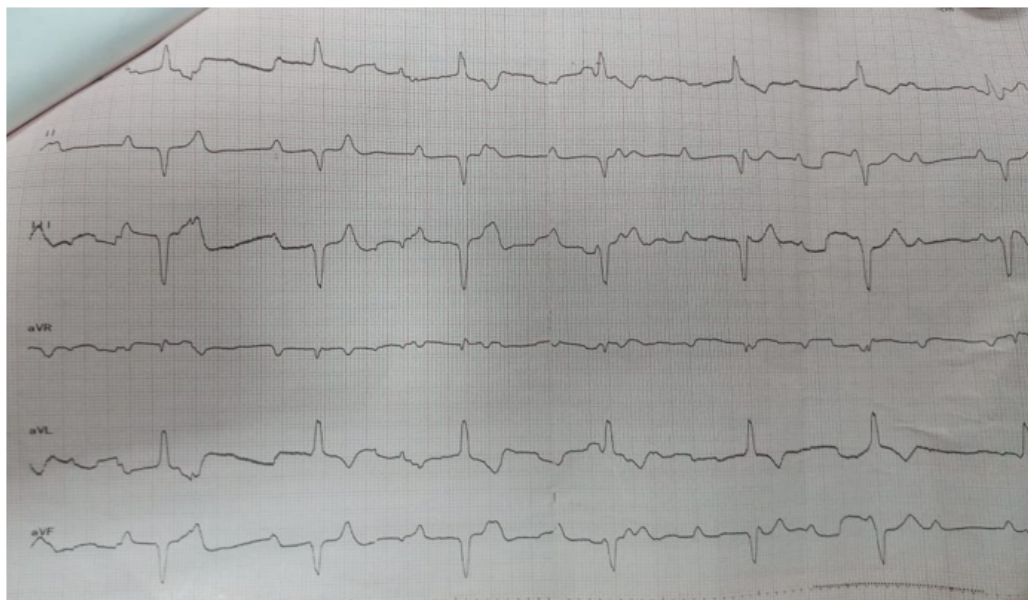
medical and surgical histories were not significant. According to the patient, he had no previous episodes of syncope, intermittent dizziness, or chest pain. There was no history of analogous case in patient’s family.

On examination done after admission in hospital, blood pressure was found to be within normal range. Cardiovascular examination was unremarkable (Table 1). No murmur was found on auscultation.

Cyanosis	Nil
Shortness of breath	Nil
Pallor	Nil
Clubbing	Nil
S1 sound	Normal
S2 sound	Normal
Jugular Venous Pressure (JVP)	Normal
Thrill	Not found
Murmurs	None

**Table 1:** Findings of cardiovascular examination.

Electrocardiogram (ECG) suggested bradycardia with 45 beats per minute. QRS complexes were found to be independent of P waves suggesting complete heart block (Figure 1 and 2).



**Figure 1:** OCT scan. Cystic macular is determined.

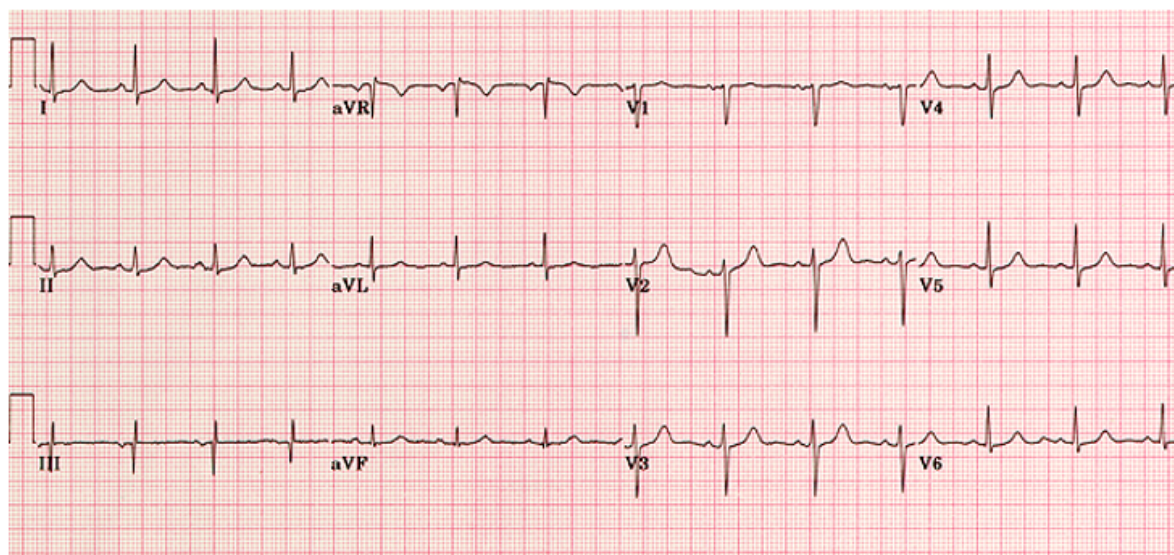


Figure 2: Normal ECG findings.

Hematologic profile was unremarkable. High-sensitivity cardiac troponin I (hs-c TnI) was 0.0037 ng/dl, indicating no ischemic event. Serum electrolytes were within normal range.

2-dimensional transthoracic echocardiography revealed levocardia with ventricles inversion. Left ventricle, both structurally adequate and fully functional, was located at the right side. Morphological right ventricle was located on left side with severely depressed function. Ejection fraction was normal. Aorta was located anteriorly. Pulmonary artery was detected posteriorly on right side of aorta. No septal defects or any other associated cardiac anomaly was found. Color flow mapping via Doppler revealed moderate to severe left-sided tricuspid regurgitation. Pulmonary valve, aortic valve and right-sided mitral valves were found to be functionally sufficient.

Patient’s symptoms and presentation with complete heart block, ECG findings, ventricular inversion and moderate to severe tricuspid regurgitation were suggestive of congenital heart lesion. All of the features were found to be indicative of levo-transposition of great arteries. O<sub>2</sub>/CO<sub>2</sub> concentration in systemic circulation was not recorded. Cardiac computed topography and MRI were not carried out due to socioeconomic constraints.

A permanent pacemaker (PPM) was implanted after which patient’s symptoms were gradually resolved. A chest x-ray was carried out post pacemaker implantation (Figure 3). Under observation, patient developed no complications and therefore, he was discharged afterwards.

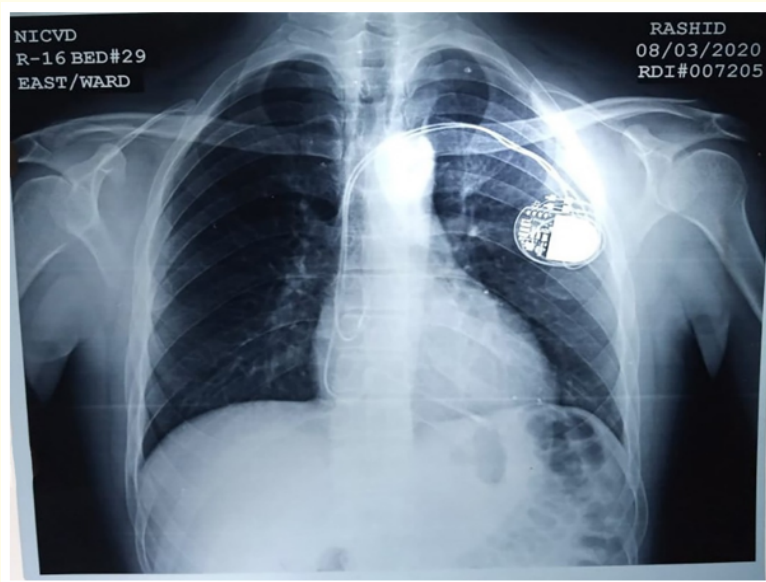


Figure 3: Chest radiograph.

### Discussion

Levo-transposition of great arteries is an atypical congenital cardiac lesion which typically remains asymptomatic until adulthood due to adequate function of discordant ventricles [5]. There is an evidence of strong association between l-TGA and heterotaxy [6]. However, in this case, normal position of heart and other organs was found. In the long run, patients with l-TGA have increased risk of heart failure, complete heart block and sudden cardiac death (SCD) [7,8].

About 55% patients with l-TGA have many other associated congenital malformations such as pulmonary stenosis, ventricular septal defects, valvular dysfunction and supraventricular tachycardias, as evident by a study [7]. Among many complications of l-TGA, congestive heart failure, tricuspid regurgitations and arrhythmias are most common [9].

Anatomically, left ventricle has thick muscular walls, strong enough to push the blood into systemic circulation. In l-TGA, inversion of ventricles results in replacement of left ventricle with discordant right ventricle along with tricuspid valve, that results in initial appropriate function of ventricle, hence causing asymptomatic disorder till adulthood, unless complicated by other associated cardiac lesions. With increasing age, due to prolonged exposure to volume overload, ventricles undergo fibrosis, which explains increased propensity of heart failure and AV block [10].

Due to presenting complaints, l-TGA may initially be misdiagnosed as inferior myocardial infarction [3]. However, this differential is excluded by serum biomarker levels. The dizziness, as complained by patient, could be explained by inadequate functioning of discordant right ventricle along with incompetent tricuspid valve resulting in regurgitation and hence compromised blood supply to brain [11].

The diagnosis of l-TGA could be based on clinical features aided by various invasive and noninvasive diagnostic modalities. Cardiac magnetic resonance imaging (MRI) is considered to be the gold standard technique in diagnosis of double discordance disease of heart [10]. Associated coronary artery malformations may be detected by coronary angiography, however, it is largely replaced by advanced techniques such as MRI, computed topography, and nuclear imaging [3].

Choice of management i.e. medical versus surgical, is multifactorial. Patient's age, severity of symptoms, ejection fraction, valvular dysfunction, associated congenital anomalies and degree of rhythm disturbances should be taken into account [12].

In our case, patient had complaints of dizziness and subjective 'sinking of heart' feeling since childhood. However, he sought medical help when the symptoms exacerbated. Findings of ECG were found to be consistent with complete heart block. Tricuspid regurgitation was detected via 2D-echocardiography. No peripheral edema or murmurs were found. Ejection fraction was normal. Considering otherwise healthy state of patient, a permanent pacemaker was inserted to counter AV block. ECG was carried out to determine the rhythm after pacemaker implantation, which revealed normal function. Patient developed no complications therefore consequently discharged after observation.

Levo-transposition of great arteries has good prognosis. Though asymptomatic, it should always be suspected in case of heart block in early age. The choice of management should be based on patient's comorbidities, ventricular function and associated congenital cardiac anomalies.

### Conclusion

This report outlined an atypical case of levo-Transposition of Great Arteries in which patient presented with heart block in aberrantly early age. L-TGA is a congenital cardiac anomaly with good prognosis if identified in early age. Timely diagnosis and management are imperative in prevention of complications associated with l-TGA. Medical practitioners should always suspect l-TGA if a young to middle aged

individual presents with complete heart block. Provided there were no associated cardiac lesions in our patient, a permanent pacemaker insertion resulted in improvement in patient's condition. Choice of management should solely depend on individual's condition and other comorbidities.

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