A Case of Congenitally Corrected Transposition of Great Arteries (CCTGA)

Ahmed TU[1], Rahman MM[2], Khan PR[3], Hasan MR[4]

Abstract

Recent years, much scientific attention has been given to congenital heart diseases (CHD) and probable complications. Congenitally corrected transposition of the great arteries (CCTGA) is a rare, complex form of congenital heart defects. CCTGA is characterized by atrioventricular (AV) and ventriculoarterial (VA) discordance and hence by a physiologically normal direction of blood flow sometimes called "double discordance". The development of complete AV block and global ventricular dysfunction has been identified as the cause of cardiac death. This paper presents a case of CCTGA with rhythm disorders and exertional dyspnea.

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Introduction

Congenitally corrected transposition of the great arteries of the heart is a rare form of congenital heart disease that was first described by Von Rokitansky in 1875 [1]. Male female ratio is approximately 1.5:1 while familial recurrence has been reported and no genetic linkage has been identified [2]. The atrioventricular dissociation means that the morphological right atrium discharges blood into the morphological left ventricle, while the morphological left atrium drains into the morphological right ventricle. Thus the left ventricle supplies the pulmonary circulation while the right supports the systemic circulation. Few patients with this anomaly survive beyond 50 years of age [3]. CCTGA has wide spectrum of structural and clinical features. The clinical presentation and prognosis of patients with CCTGA vary depending on the severity of the associated cardiac anomalies, the development of systemic ventricular dysfunction, and the development of arrhythmias. The patients with CCTGA have a progressive risk of spontaneous complete AV block throughout life (2% per year),[4] less common exercise intolerance and fatigability. The incidence of sudden cardiac death (SCD) in CHDs is approximately 1:1000 patients per year, which is 25-100 times greater than in the general population. [5]

Case Report

A 12 years-old boy was admitted into the cardiology department of Community Based Medical college hospital with the complaints of palpitation and chest discomfort 15 days. He was moderately active but fatigue easily, the patient reported no fainting episodes, dyspnea, chest pain, or dependent oedema. The patient had neither family history of congenital heart disease nor any sudden death at a young age.

On examination, the boy weighted 28 kg and was 122 cm tall. There was no cyanosis or clubbing of the fingers, Pulse 52/bpm regular BP/110/70 mmHg. The heart was overactive,

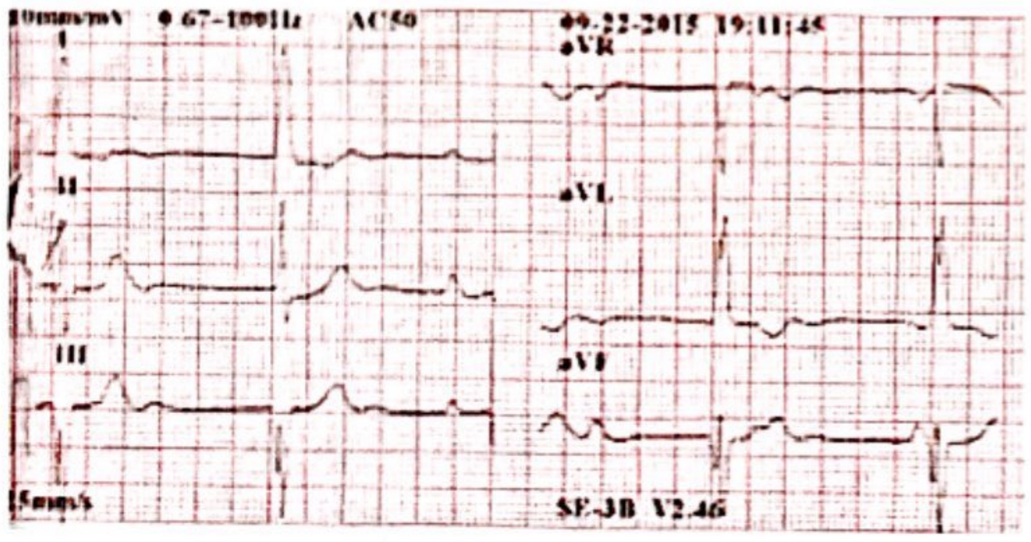
1. * Dr. Tofayel Uddin Ahmed
Assistant Professor, Department of Cardiology
Community Based Medical College, Bangladesh
Winnerpar, Mymensingh.
2. Prof. Dr. M. Mafizur Rahman
Professor & Head of the Department of Cardiology
Community Based Medical College, Bangladesh
Winnerpar, Mymensingh.
3. Dr. Parvez Rahman khan
Registrar, Department of cardiology
Community Based Medical College, Bangladesh
Winnerpar, Mymensingh.
4. Dr. Mohammad Rajibul Hasan
Assistant Professor, Department of Anaesthasiology
Community Based Mediçal College, Bangladesh
Winnerpar, Mymensingh.

***Address of correspondence**
Email : tuahmed@gmail.com
Mobile: +8801711906682

a pan systolic murmur was heard along the left sternal border, and a soft apical mid diastolic murmur was present. The second sound was loud and single. On investigation Hb% 11.8 gm/dl, ESR=59 mm in 1st hour, Neutrophil 58% Lymphocyt-33% Monocyte 03% other biochemical examination was normal.

The ECG showed first degree atrioventricular block with prolong PR interval (280ms),

Heart rate 52/bpm regular (Figure 1a,1b).



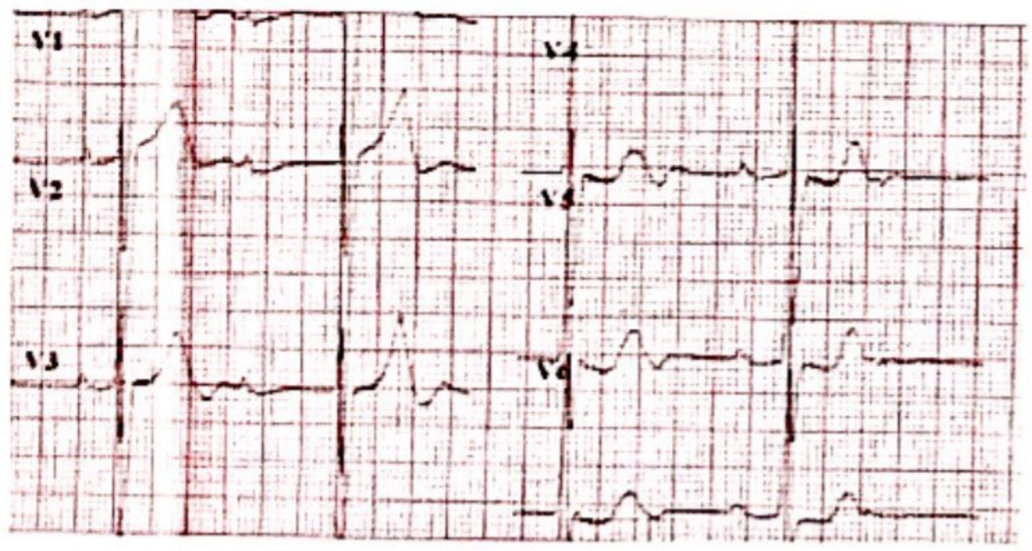


Figure:1a

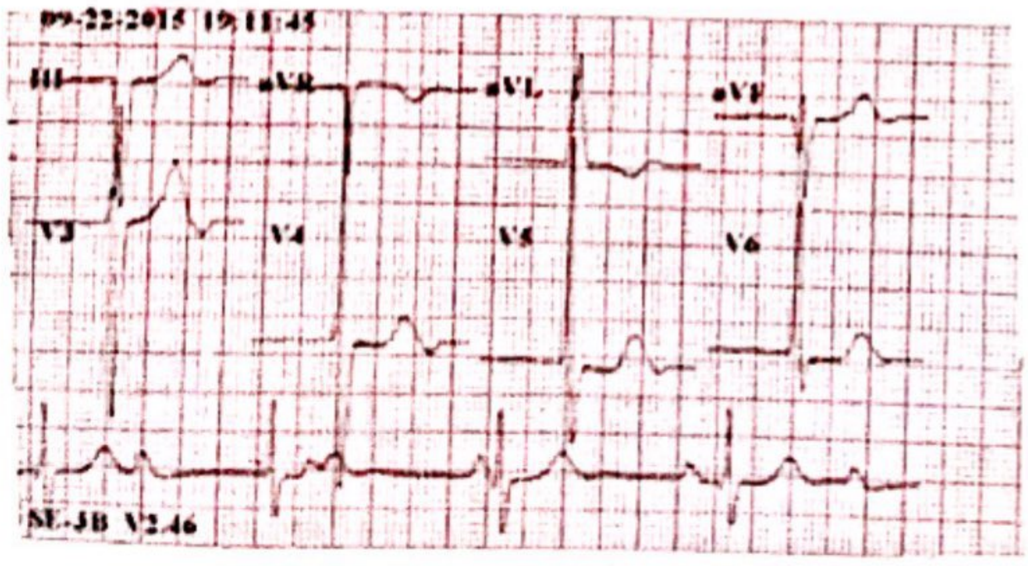


Figure :1b

The chest X-ray showed a slightly elevated cardiac index and narrow aorticopulmonary trunk. More specifically there was no opacification of the asending aortic arch and the main pulmonary arterial trunk with straightening of the left upper cardiac margin and a round lower left cardiac border. (Figure : 2)

Figure : 2

Echocardiography revealed

1. Normal position of the atria.
2. Atrio-ventricular discordance (right atrium is connected to morphologic left ventricle & left atrium is connected to morphologic right ventricle). (Figure-3a)
3. Ventriculo arterial discordance (aorta arises from right ventricle & pulmonary artery arises from left ventricle) (Figure-3a)
4. There is no stenosis at any level apart from mild valvular insufficiency (Figure-3b)
5. The courses of the two great vessels were parallel (Figure: 3c)

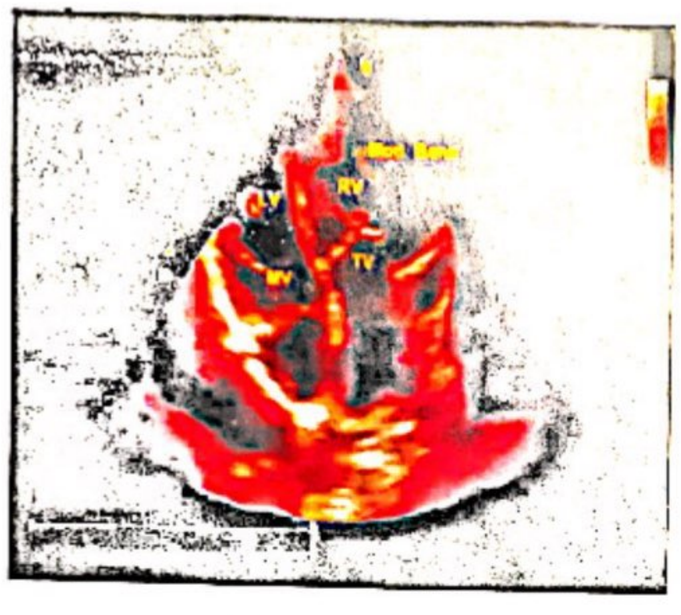


Figure: 3a

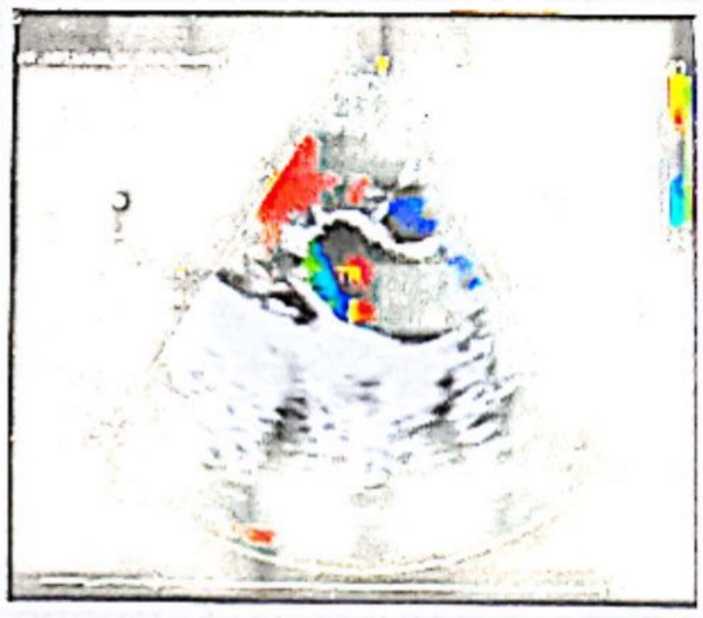


Figure: 3b

Figure: 3c

Figure : 3d

Discussion

In CCTGA the Atrioventricular relationships are discordant: the right atrium is connected to the left ventricle and the left atrium to the right ventricle (also known as ventricular inversion). [6]

Because the two great arteries arise in parallel, there is "figure eight" appearance rather than usual arrangement pulmonary artery in long axis surrounding aortic valve [2]. The two fundamental anatomic abnormalities in CCTGA consist of transposition of the ascending aorta and pulmonary trunk, as well as inversion of the ventricles. This arrangement results in desaturated systemic venous blood passing from the right atrium through the mitral valve to the left ventricle and into the pulmonary trunk, whereas oxygenated pulmonary venous blood flows from the left atrium through the tricuspid valve to the right ventricle and into the aorta. The circulation is thus physiologically "corrected" without other defects, the hemodynamic would be nearly normal.[6] The clinical presentation, course and prognosis of patients with CCTGA vary depending on the nature and severity of any complicating intracardiac anomalies, as well as development of dysfunction of the systemic subaortic right ventricle (RV).Congenitally corrected transposition of the great arteries is a rare entity that represents less than 1% of all clinically diagnosed congenital heart diseases. Recognisable associated defects are seen in 98% of cases.[7] 1-10% of individuals with congenitally corrected transposition of the great vessels have no associated defects.[8] First degree atrioventricular block is found in around 50% of cases, while its progression to complete heart block occurs at a rate of 2% per year[9] Insufficiency of the systemic ventricle is the cause of death in more than 50% of patients.[9] If the insufficiency of the systemic(tricuspid) atrioventricular valve is severe it must be replaced, and this should be done before the appearance of dysfunction of the systemic right ventricle (ejectionfraction >45%).[10] When the tricuspid insufficiency is combined with dysfunction of the systemic right ventricle, the double switch operation (arterial and atrial) is considered appropriate.[11,12] A total cavopulmonary connection may be definitive surgery.[13] and Heart transplantation remains the final option[2] The clinical course of corrected transposition of the great vessels depends on the presence and severity of the associated defects. Even in the absence of such anomalies, or after their surgical repair, the question remains whether the anatomical right ventricle is capable of maintaining an adequate cardiac output over a long period.[14] Regular schedule follow up

examinations are recommended to detect progressive AV conduction disorders and the progression or late appearance of left AV valve incompetence. Antibiotic coverage as protection against infective endocarditic is recommended.[15] The international literature contains very few cases of adult patients with congenitally corrected transposition of the great arteries without associated defects or surgical intervention [16] where with rastelli procedure RV to PA condui has been successfully performed[2]It is estimated that approximately 1% of individual with corrected transposition have an otherwise normal heart. [15]

Our patient adds one more case to this short list. CCTGA without associated defect.

Conclusion

Congenitally corrected transposition of the great arteries is a rare cardiac anomaly where many patients will remain asymptomatic for much of their lives. CCTGA patients have a reduced tolerance for exercise and have reported reduced health-related quality of life compared to a control population.

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