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Case report: Congenitally corrected transposition of the great arteries (CCTGA)

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Abstract
Corrected transposition of the great arteries (ccTGA) accounts for <0.5% all congenital heart diseases [4]. It is a physiologically "corrected" transposition of the great arteries [1]. ccTGA characterized by discordant Atrioventricular (AV) and Ventriculoarterial (VA) connections [4]. The main defect of this abnormality comprises a combination of atrioventricular as well as ventriculoarterial discordance [2]. Three other abnormalities, including ventricular septal defect, left ventricular (pulmonary) outflow tract obstruction and tricuspid valve abnormalities are associated with the ccTGA [4]. We introduce the case of a 40-year-old active labor referred to the Cardiology department of Aliabad teaching hospital for evaluation of valvular heart disease for transthoracic echocardiogram.

Keywords: congenital heart disease; corrected transposition of the great arteries; echocardiography

Introductions
ccTGA describes discordant AV and VA connections, the atria connect to the inappropriate ventricles, which then connect to the inappropriate great arteries [4]. Congenitally corrected transposition of great arteries is uncommon congenital heart disease associated with multiple cardiac morphological abnormalities and conduction defects [5]. ccTGA accounts for <0.5% of all congenital heart diseases [4]. Transposition of the great arteries is a congenital heart defect [2]. Due to abnormal development of the fetal heart during the first 8 weeks of pregnancy, the large vessels that carry blood from the heart to the lungs and to the body are improperly connected [1]. As the name suggests, the great arteries are transposed as they relate to the heart [5]. The systemic veins join the Right Atrium (RA), which is connected by a mitral valve to the sub pulmonary Left Ventricle (LV). The Left Atrium (LA) receives pulmonary venous blood from the pulmonary veins and is connected by a tricuspid valve to the sub aortic Right Ventricle (RV) [4]. The VA connections are also discordant wherein the aorta arises from the morphologic Right Ventricle and the pulmonary artery from the morphologic Left Ventricle [4]. Discordant AV and VA connections can occur in isolation; but, associated congenital cardiac anomalies like ventricular septal defect, left ventricular (pulmonary) outflow tract obstruction and tricuspid valve abnormalities are common [4].

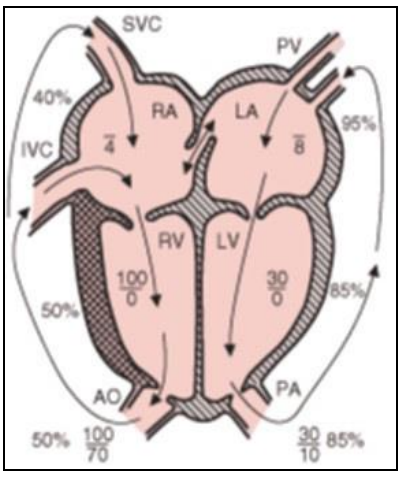

Fig 1: Anatomical confirmation of TGA to ccTGA.

If we compere Patients with ccTGA to patients with normal cardiovascular anatomy, they have reduced exercise tolerance and reduced health-related quality of life [1]. Furthermore, Patients with ccTGA usually diagnosed during investigations like ECG, chest X-ray and Trans Thoracic Echocardiographic done for other reasons [3]. This anomaly is often missed because of failure to recognize the above mentioned anatomical variations [2]. We introduce the case of a 40 year-old active labor referred from Outpatient Department (OPD) of Aliabad Teaching Hospital to the Cardiology department of Aliabad teaching hospital for evaluation of valvular heart disease and Echocardiography. This case also illustrates how Echocardiography can be helpful in diagnosing such uncommon congenital heart diseases.

Ethical consideration: The ethical consideration of this study is reviewed and approved by ethical research committee of Kabul university of medical science.

Case Report

A 40-year-old man presented to the OPD of Aliabad Teaching Hospital with mild exertional dyspnea, mild neck vein distention and harsh systolic murmurs (grade III) at 2nd left intercostal area at mid clavicular line with unknown significant past medical history. For evaluation and accurate diagnose he referred to Cardiology department of Aliabad Teaching Hospital. His 12-lead ECG demonstrated first degree AV block.

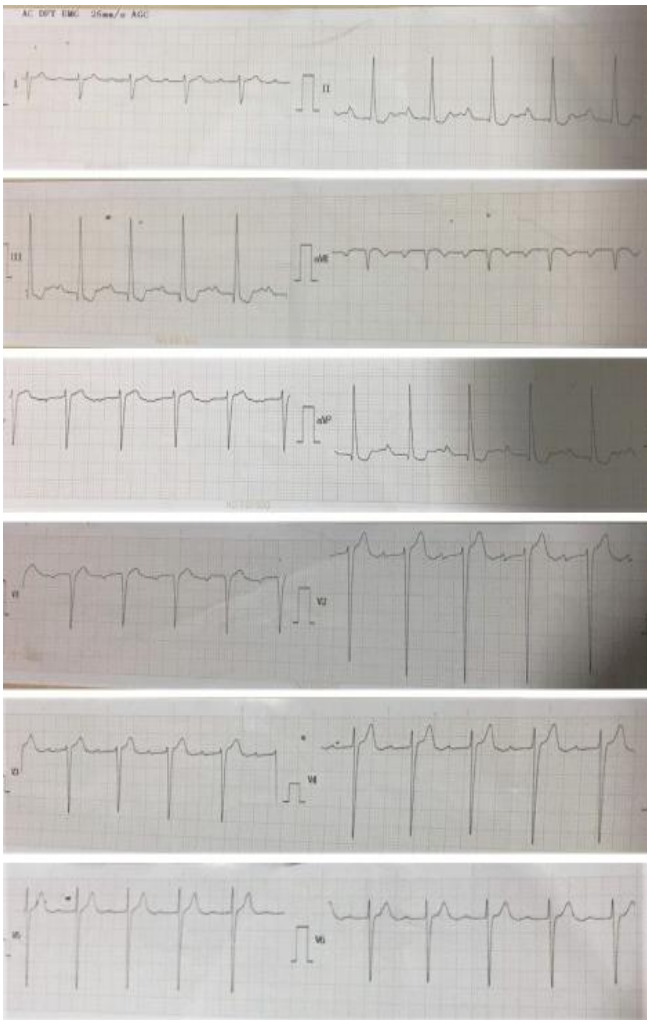

Fig 2: ECG changes seen in ccTGA

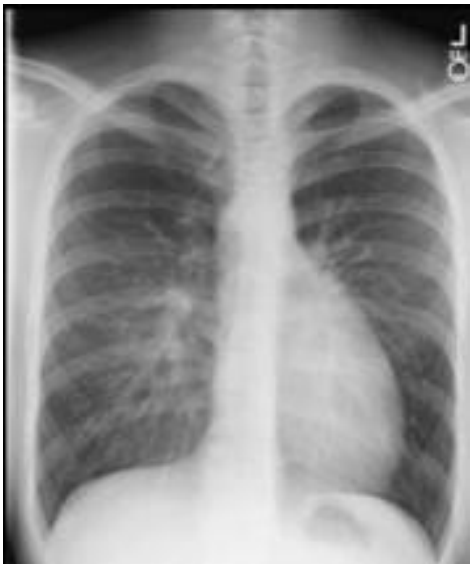

Fig 3: X-ray of the patient

X-ray image which was available in the patient file was reported normal. The patient felt fatigued during physical activity (200–400 meters).

Exercise Stress Test (Treadmill testing) with standard Bruce protocol during 6 minutes and 50 seconds was suddenly stopped due to shortness of breath and fatigue. The patient couldn't achieve the maximum predicted heart rate (65%) and workload of 7 Metabolic equivalents (METS). No ischemic changes were noted on Electrocardiogram during the study.

Subsequently, the patient underwent transthoracic echocardiogram for further evaluation of cardiac structure. Transthoracic echocardiogram completed. His Transthoracic echocardiogram showed severe pulmonary stenosis (confirmed by three-dimensional (3D) echo) and large peri-memembranous VSD (26mm), morphologically right ventricular location is on the left side and left ventricular is on the right side. Patient ventricular function was normal (LVEF=60%), but the tricuspid left AV was mildly regurgi-tated. There were not any obvious significant valvular abnormalities on the study. The above mentioned findings (atrioventricular and ventriculoarterial discordance) were in keeping with congenitally corrected transposition of great arteries.

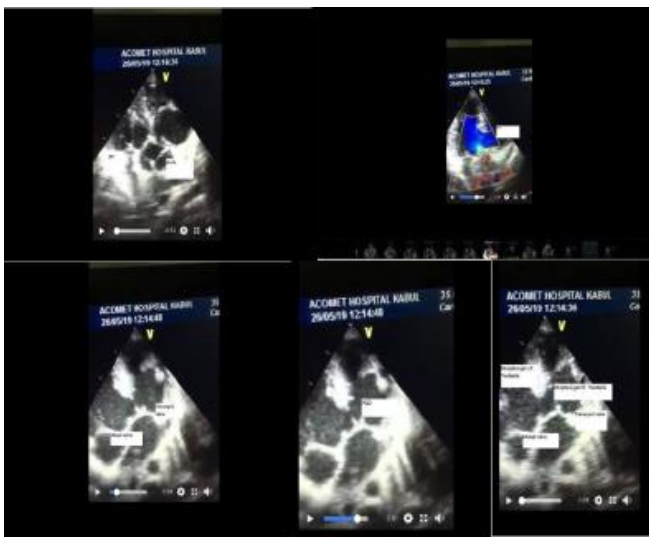

Fig 4: Echocardiography findings in ccTGA.

Discussion

Congenitally corrected transposition of the great arteries (ccTGA) accounts for less than 0.5% of all congenital cardiac anomalies, 4 and often diagnosed in adult age. ccTGA is commonly associated with other cardiac defects and its isolated occurrences are rare [5]. Most patients have at least one additional cardiac abnormality consisting of left ventricular (pulmonary) outflow tract obstruction (50%), ventricular septal defects (60%), and atrial septal defects (19%) as well as abnormality of tricuspid valve (90%) [1, 3]. Aorta is positioned anteriorly and to the left side of the pulmonary artery [1]. 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. Sudden Cardiac Death (SCD) is still the leading cause of death in patients with Congenitally Heart Disease (CHD). ccTGA has the highest mortality rate among all CHD patients [1]. It is not unusual for ccTGA to be found on later adulthood [2]. Some patients are asymptomatic throughout their life and diagnosed incidentally [3]. Often a new murmur, rhythm problems, or heart failure symptoms – that leads to a diagnosis of ccTGA. So it is found accidentally when routine heart tests are done. Echocardiography can be used to diagnose ccTGA in adulthood [4]. But rates of misdiagnosis of ccTGA using echocardiography at Adult cardiac centers are high [3]. The prognosis depends on correct diagnosis and associated heart defects. The 20-year survival rate for congenitally corrected transposition of great arteries is 75%.1 Since ccTGA is a complex defect, specialized care from cardiologist is mandatory in every case.

Conclusion

Congenitally corrected transposition of the great arteries is not a prevalent cardiac anomaly in which there won't be symptoms for many years, unless heart failure symptoms appear, that's why such patients are diagnosed by chance with in a long time. By comparing these patients to controlled group it turns out that they have decreased exercise capacity in their daily activities. Complications such as heart failure or some possible symptoms could be prevented by Tran Thoracic Echocardiography which is a nice modality for early diagnosis.

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