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CASE REPORTS

A Rare Case of Congenitally Corrected Transposition of Great Arteries with Complete Heart Block

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Introduction

Congenitally corrected transposition of great vessels (CCTGA) is a rare congenital heart defect (CHD) and constitutes less than 1% of all congenital heart disease. The two fundamental anatomic abnormalities in CCTGA consist of transposition of the ascending aorta and pulmonary trunk, as well as inversion of 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. Thus the circulation corrected functionally. The clinical presentation, course and prognosis of patients with CCTGA vary depending on the nature & severity of any complicating intracardiac anomalies, as well as development of dysfunction of the systemic subaortic right ventricle. Cardiac deaths have been attributed to the development of complete atrioventricular (AV) block, systemic ventricular dysfunction or systemic AV valve incompetence.[1-4] Very rarely in patients of CCTGA, ventricular tachycardia (VT) has been attributed to the cause of sudden cardiac death (SCD).[5] Furthermore , fatal arrhythmia is very rare in the absence of systemic ventricular dysfunction.

Case Report

A boy of 13 year presented with recurrent episodes of unconsciousness and exertional palpitation for six months. While playing in the field, he first experienced black out followed by unconsciousness of 10-15 minutes duration with uneventful recovery, six months back. He experienced same type of episodes repeatedly. He had exertional palpitation, subsided spontaneously after taking rest along with it. In early childhood, he had history suggestive of recurrent respiratory tract infection. He had no history of chest pain, breathlessness, nausea, vomiting, headache, convulsion, tongue bite, involuntary micturition & defecation or any change of posture.

He was mildly anemic, Pulse was 50/min, regular, high volume; BP was normal & Cannon wave was present in JVP. His apex beat was normal, palpable P2 and Left parasternal heave present with variable S1, loud P2 in pulmonary area and 3/6 Pansystolic murmur all over the

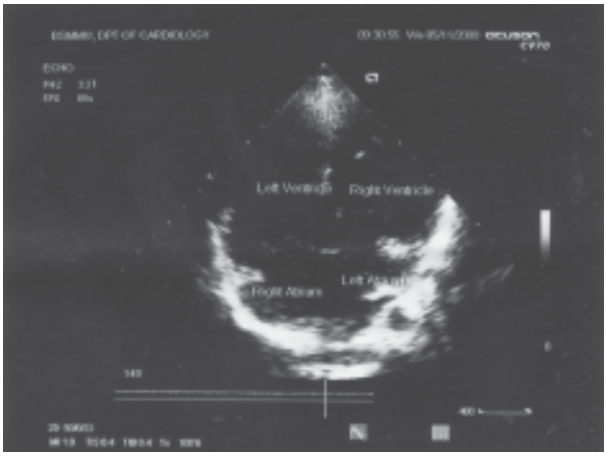

Fig.-1

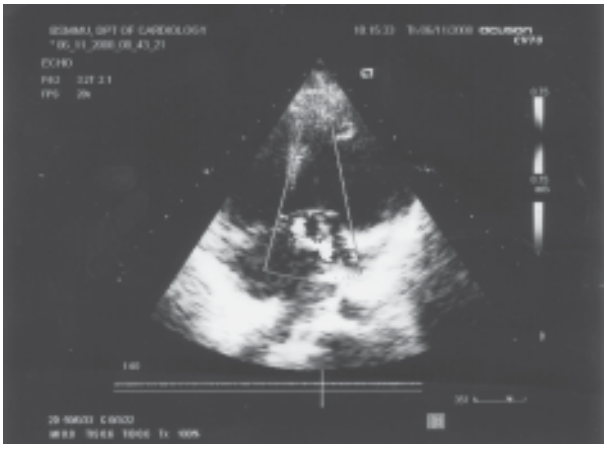

Fig.-2

precordium, especially in left lower parasternal area. Electrocardiography (ECG) showed complete heart block with heart rate around 50.

Transthoracic echocardiography showed an L-TGA (congenitally corrected) pattern, moderate Tricuspid valve insufficiency (systemic AV valve), normal RV systolic function, no residual VSD. He was managed with ACE inhibitor & diuretics and discharged in clinically stable condition. He was also suggested for Left sided Tricuspid valve replacement along with permanent pace maker (PPM) implantation in DDD mode with active fixation of electrodes.

Discussion

CCTGA has the highest mortality among all CHD patients. SCD is the most common cause of death in patients with CHD.[7] SCD has been attributed to the presence of complete AV block or a rapid progression from first degree AV block to complete AV block.[8-10] Very rarely in a patient with CCTGA, VT has been attributed to the cause of SCD.[5] In our patient like the majority of patients with CCTGA; complete AV conduction block was present. Patients with an anatomical systemic RV are at significant risk (22%) for heart failure,[11] as the ventricle was not meant to take the systemic load developmentally. Graham et al [12] concluded that after childhood, systemic ventricular dysfunction is more common and may reflect the inability of the anatomic RV to function as a systemic pumping chamber over a normal lifespan. The systemic ventricular dysfunction makes the heart more prone to arrhythmias. The best predictors of mortality in patients with systemic RV dysfunction are NYHA class and systemic ventricular ejection fraction.[11] Our review of literature focusing on the coronary arterial anatomy in CCTGA has demonstrated that some patients had significant coronary abnormalities,[13-15] but interestingly this was less frequently than what has been observed in other common congenital cardiac lesions.[16-19] It is important to know of the presence of a coronary anomaly (in origin, course and distribution) in order to determine the optimal surgical approach and outcome.[11] Previous studies have suggested a consistently inverted coronary arterial pattern in patient with CCTGA and, rarely, a coronary ostium or artery.[6]An anatomical systemic ventricle ,which makes a venous ventricle in a patient with CCTGA , presents challenges that are unique during PPM or ICD lead placement. Unlike a true RV, which is full of trabeculi where the PPM or ICD lead can be positioned, the anatomical systemic ventricle is devoid of these structures, thus making it harder to obtain a stable lead position and possibly increasing the risk of lead displacement. This can be overcome by selecting an active-fixation PPM or ICD lead from the beginning and actively fixing it to interventricular septum. Secondly, the anatomical and fluoroscopic landmarks of the ventricle are completely altered, as chambers are backwards due to transposition of the ventricles. This requires the operator to use oblique views more frequently and to constantly remain aware of the anatomy during the implantation procedure.

All patients should have at least annual Cardiology follow-up. Regular assessment of systemic (tricuspid) AV valve regurgitation by serial echocardiographic studies and systemic ventricular function by MRI or radionuclide angiography should be done. Holter recording can be useful if paroxysmal atrial tachycardia or transient complete AV block is suspected.

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