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A Rare Cause of Heart Failure - Congenitally Corrected Transposition of the Great Arteries

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Abstract

Transposition of great arteries is a rare congenital heart disease where the two main blood vessels leaving the heart are in abnormal position where the pulmonary artery arises from the left ventricle and aorta arises from the right ventricle. CCTGA is a rare cardiac condition also called L-TGA, double discordance or Atrioventricular and ventricular arterial discordance where the right atrium is connected to left ventricle supplying blood to the pulmonary artery whereas the left atrium is connected to the right ventricle supplying blood to the aorta despite of this oxygenated and deoxygenated blood flow is in the correct physiologic direction resulting in normal oxygenation and perfusion. In these patients the right ventricle is subjected to systemic pressure and fails prematurely. The most common associations with CCTGA are Ventricular septal defect, pulmonary stenosis, left Atrioventricular valve (morphological tricuspid valve) regurgitation

Keywords: CCTGA, L-TGA, VSD.

Case report

A rare case of heart failure like CCTGA, like in this case of 17yrs old male who came with chest pain and breathlessness on exertion.

2D ECHO showed congenitally corrected transposition of great arteries with VSD and PS .There is a large inlet VSD with sub pulmonic extension with bidirectional shunt. Doming pulmonary valve. There is severe pulmonary stenosis with peak gradient of 80mmHg .Mild aortic regurgitation .No AV valve regurgitation. Good biventricular function. Left aortic arch.

CT chest plain and contrast done preoperatively – Morphological right ventricle is located on the left side and morphological left ventricle is located on the right side. Large inlet VSD seen. Moderate to severe infundibular stenosis seen. Doming of the pulmonary valve noted. Main pulmonary artery is arising from the right ventricle that is morphological left ventricle and aorta is arising from the left ventricle that is the morphological right ventricle.

Diagnostic catch done which showed, Left circumflex and RCA arising from the left cusp and left main arising from the right cusp.

Diagnosis of Congenitally corrected transposition of greater arteries with VSD and PS in a 17yrs old male was made.

Nikaidoh procedure was done; it is a method of anatomic correction in which the left ventricle is connected to aorta and the right ventricle to pulmonary artery via the arrangement of pulmonary trunk behind the ascending aorta

Discussion

CCTGA is a rare disease that accounts for less than 1% of congenital heart diseases. CCTGA without other frequently associated cardiac anomalies sometimes remains undiagnosed for decades because symptoms are often minimal and nonspecific, and many asymptomatic patients are first given a diagnosis of CCTGA

Theoretically, no functional abnormalities exist, but unfortunately, most cases are complicated by associated intracardiac defects, AV conduction disturbances, and arrhythmias.

A. VSD occurs in 80% of all cases
B. PS, both valvular and subvalvular, occurs in 50% of patients and is usually associated with VSD.
C. Systemic AV valve (tricuspid valve) regurgitation occurs in 30% of patients
D. Occasionally, complex associated defects are present with hypoplastic ventricle, AV valve abnormalities, or multiple VSDs
E. Both varying and progressive degrees of AV block and paroxysmal supraventricular tachycardia (SVT) frequently occur.

The cardiac apex is in the right chest (dextrocardia) in about 50% of cases. The coronary arteries show a mirror-image distribution, the right coronary artery supplies the anterior descending branch and gives rise to a circumflex; the left coronary artery resembles a right coronary artery.

Conclusion

Patient with CCTGA can live for years without diagnosis .The prognosis depends largely on the right ventricle to adapt to the systemic circulation .The presence of pulmonary stenosis provides a physiological protection. Surgery helps in correction of the heart failure

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