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RESEARCH ARTICLE

CCTGA WITH DORV WITH EISENMENGERIZATION: A RARE CYANOYIC CONGENITAL HEART DISEASE

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ABSTRACT

Congenitally Corrected Transposition of the Great Arteries CCTGA and Double Outlet Right Ventricle DORV are two complex congenital heart defects affecting the structure and function of the heart. CCTGA is characterized by atrioventricular and ventriculoarterial discordance allowing for relatively normal circulation despite the anatomical abnormality[i] In contrast DORV occurs when both the aorta and pulmonary artery arise predominantly from the right ventricle often accompanied by a ventricular septal defect VSD leading to variable degrees of oxygenation impairment and cyanosis [ii]. We are reporting a rare case of CCTGA with DORV in a young 32 year male was admitted with difficulty breathing since childhood on exertion . 2D ECHO and cardiac MRI suggestive of CCTGA WITH DORV with late complication eisenmengerization.

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INTRODUCTION

CCTGA is a rare congenital heart defect where the heart's ventricles and the great arteries are swapped in a way that compensates for the defect. The blood flow relatively normal despite the anatomical differences.

- The right atrium connect to left ventricle, which pumps blood into the pulmonary artery
- The left atrium connects to the right ventricle, which pumps blood into the aorta.

This double switch (atrioventricular and ventriculoarterial discordance) allows oxygenated blood to flow to the body and deoxygenated blood to flow to the lungs. DORV (Double Outlet Right Ventricle) both the aorta and the pulmonary artery arise predominantly from the right ventricle, which disrupts normal blood flow.

- There is often a ventricular septal defect (VSD), which serves as the only outlet for blood to move between the ventricles.
- The position of the VSD and the relative alignment of the great arteries determine the blood flow pattern.
- Oxygen-rich and oxygen-poor blood can mix, leading to cyanosis (low oxygen levels in the blood)

Eisenmengerization is a late stage complication of CHDs with significant left to right shunting, such as VSD, ASD, PDA. Eisenmenger syndrome significantly reduces life expectancy and quality of life [iii].

CASE REPORT

A 32 year old married male patient was admitted to Male Medicine Ward of RNT Medical College, Udaipur, Rajasthan with complaints of shortness of breath during exertion since childhood which worsened in the last two months.Patient also have complaint of easy fatigability for last few days. There was no history of orthopnoea and PND. The patient was never admitted anywhere before. At the time of admission patient was conscious and oriented to time place and person. His vital signs were pulse 84/min regular, respiratory rate 20/min , oxygen saturation (spo2) was 90% at room air and temprature was normal. His general physical examination revealed cyanosis on lips, tongue, ear lobes,tip of nose, mucous membranes and clubbing grade 2, no pallor, icterus, edema, lymphadenopathy, patachiae/purpura were seen.on

examination chest is bilateral symmetrical and normal bronchovesicular breath present on auscultation and no added breath sound was present. On cardiac auscultation harsh pansystolic murmur was present at the lower left sternal border. Comprehensive laboratory investigations were performed. The complete blood count (CBC) showed hemoglobin 19.5 g/dl, TLC -9970/cu mm, platelet count 1.01 lakh /cu mm. The liver and kidney functions were normal.ECG was showing P pulmonale and T wave inversion in inferior leads and V4 to V6 . Chest Xray showing cardiomegaly (figure 1).

2D ECHO suggesting situs soleus, levocardia, intact IAS, atrio ventricular discordance is present, morphological LV connected to RA & RV connected to LA, pulmonary artery and aorta is arising from morphological RV, large perimembranous VSD size ~ 34-35 mm with bidirectional shunt predominantly right to left shunt, pulmonary artery is posterior to aorta, normal LV systolic function but severe morphological RV systolic dysfunction, moderate PR ,left sided aortic arch, no CoA/PDA/AS, no clot/vegetations and pericardial effusion impression is CCTGA with DORV with Eisenmengerisation.

Aspect	CCTPGA	DORV
Primary problem	Ventricular and arterial connections are reversed	Both great arteries arise from the right ventricle
Blood Flow	Corrected by 'double switch' anatomy	Often mixed oxygenated and deoxygenated blood
Symptoms	May be symptomatic initially , later heart failure	Cyanosis , shortness of breath, heart failure
Associated defects	VSD, pulmonary stenosis, arrhythmias	VSD,pulmonary stenosis , transposition of arteries

Cardiac MRI (figure 2) also revealing same finding with following chamber morphology

Chamber	Dimensions(4ch view) midsection transverse(mm)
Right atrium	55.5 (dilated)
Anatomical right ventricle	61.1(dilated)
Left atrium	52.4 (dilated)
Anatomical left ventricle	35

To rule out any impairment in brain growth and development in CHD patient, MRI brain (Figure 3) also done which was normal. Due to disease has progressed and systemic ventricle developed, surgical intervention not feasible. Only conservative management can be continued.

DISCUSSION

CCTGA is a rare CHD in which both the atrioventricular (AV) and ventriculoarterial (VA) connections are transposed, resulting in a physiologically corrected circulation despite abnormal anatomy. The right atrium connects to the morphologic left ventricle, which pumps blood into the pulmonary artery, while the left atrium connects to the morphologic right ventricle, which pumps blood into the aorta. This "double switch" maintains normal oxygenation, but over time, the systemic right ventricle may fail because it is not designed to handle high systemic pressures [iv].

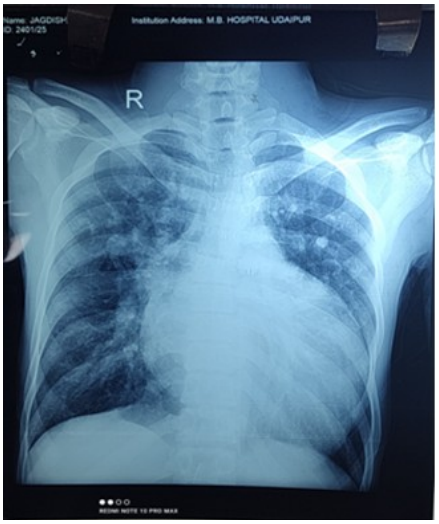

Figure 1. XRAY Chest showing cardiomegaly

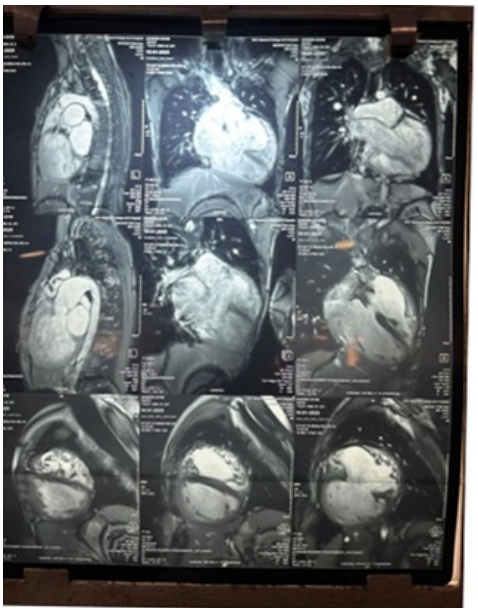

Figure 2a

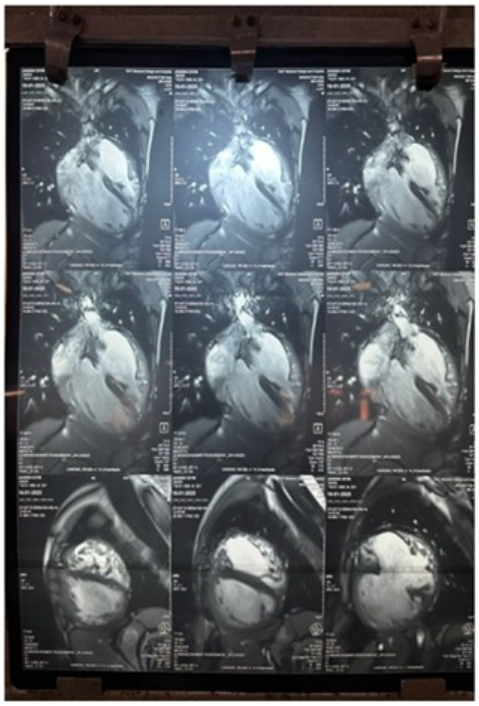

Figure 2b. Cardiac MRI showing CCTGA Cardiac MRI

DORV is characterized by both the aorta and pulmonary artery arising predominantly from the right ventricle, leading to abnormal circulation. The presence and location of a VSD determine the direction of blood flow. Unlike CCTGA, which can maintain physiologic circulation, DORV often results in mixing of oxygenated and deoxygenated blood, leading to cyanosis and heart failure. CCTGA may remain undiagnosed until adulthood due to its physiologically corrected circulation [v]. Symptoms often arise due to right ventricular dysfunction, tricuspid regurgitation, or arrhythmias. Patients may develop heart failure as the right ventricle struggles to sustain systemic circulation DORV usually presents in infancy or early childhood with cyanosis, respiratory distress, and failure to thrive, depending on the degree of mixing and pulmonary blood flow[vi]. Both conditions require detailed imaging for diagnosis and surgical planning.Echocardiography Evaluates ventricular function, great artery alignment, and associated defects and Cardiac MRI/CT provides detailed anatomical assessment.

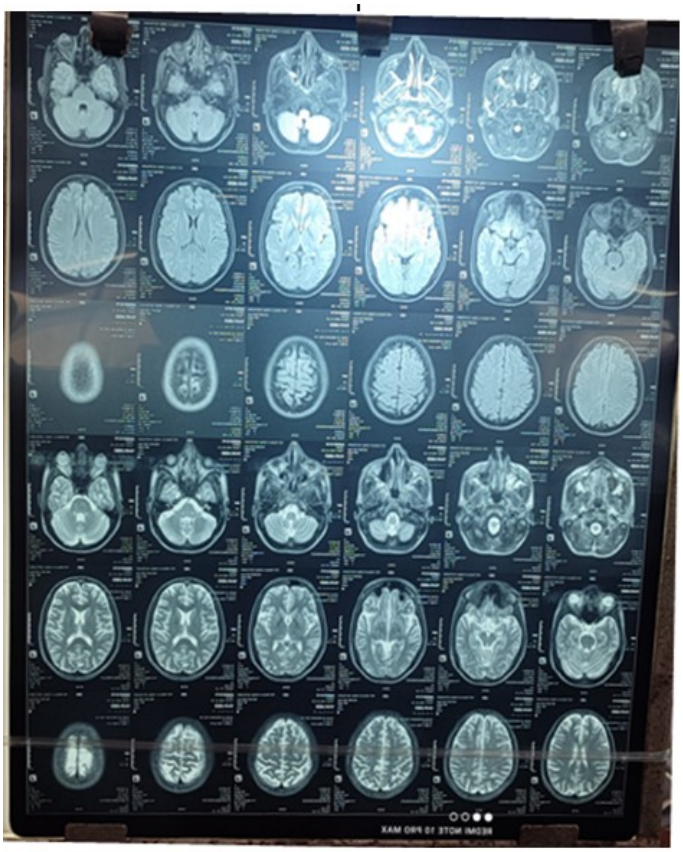

Figure 3. MRI Brain

Management Strategies: CCTGA Patients with mild defects may only need monitoring. Those with systemic right ventricular failure require heart failure medications such as beta-blockers, ACE inhibitors, and diuretics. Many patients develop conduction Issues, requiring pacemaker implantation. Tricuspid valve repair/replacement if severe regurgitation develops. Double-switch procedure (Senning/Mustard atrial switch and arterial switch) in select cases to redirect the right ventricle to the pulmonary circulation and the left ventricle to the systemic circulation[vii].

In DORV patients Intraventricular tunnel repair (to direct the VSD flow to the aorta or pulmonary artery) and Arterial switch operation (for Taussig-Bing anomaly) and Rastelli procedure (using a conduit to connect the left ventricle to the aorta) are avilable surgical interventions [viii]. Palliative procedures, such as pulmonary artery banding, in cases of excessive pulmonary blood flow before definitive repair.

CONCLUSION

Both CCTGA and DORV are complex congenital heart defects with distinct anatomical and physiological implications. While CCTGA allows for near-normal circulation but predisposes patients to late complications, DORV frequently leads to early cyanosis and requires surgical correction[ix]. Advances in imaging, surgical techniques, and long-term management have significantly improved the survival and quality of life for patients with these conditions. Ongoing research into right ventricular function preservation and optimal surgical strategies continues to enhance outcomes for affected individuals.

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