Case Report

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A Rare Case of Double Chambered Right Ventricle with Anomalous Left Anterior Descending Coronary Artery from Pulmonary Artery (ALADAPA)

Anupama Rao[1], Vijay L[2], Sathya Patnaik[3], P K Dash[4]

Author's Affiliation: [1]Assistant Professor, Department of Pediatric Cardiac Surgery, [3]Associate Professor and HOD, Department of Cardiac Anesthesia, Hi-Tech Medical College, Pandara, Bhubaneshwar, Odisha 751025, India, [2]Senior Consultant, Department of Cardiothoracic Surgeon, Star Hospital, Visakhapatnam, Andhra Pradesh 530040, India, [4]Director of Cardiac Sciences, Department of Cardiac Sciences, Capital Hospital, Bhubaneshwar, Odisha 751020, India.

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Abstract

Double-chambered right ventricle (DCRV) is a rare congenital heart disease in which the right ventricle (RV) is hypertrophied due to anomalous muscle bundles. Anomalous left coronary artery from pulmonary artery (ALCAPA) is also a rare lesion constituting 1 in 3,00,000 live births. Isolated LAD from pulmonary artery (ALADAPA) is further rare. We report a case of DCRV associated with Anomalous left anterior descending Artery from Pulmonary artery (ALADAPA). The patient was initially diagnosed to have DCRV and on further evaluation associated anomalous left anterior descending artery from pulmonary artery was detected. The patient underwent DCRV repair and Takauchi repair and had an uneventful post operative recovery.

Keywords: Congenital Heart Disease; Double Chamber Right Ventricle; DCRV; ALCAPA; ALADAPA.

Introduction

Double-chambered right ventricle (DCRV) is a rare form of congenital heart disease occurring in 0.5 – 2% of all patients with congenital heart disease.[1] In DCRV, the right ventricle (RV) is divided into into proximal high pressure and distal low pressure chamber.[2] It usually presents in childhood with few cases reported in adults.[3] Anomalous left coronary artery from pulmonary artery (ALCAPA) is also a rare lesion constituting 1 in 3,00,000 live births.[4] Isolated LAD from pulmonary artery (ALADAPA) is further rare. We report a rare case of DCRV associated with Anomalous left Coronary Artery from Pulmonary artery (ALADAPA). (Fig. 1)

Corresponding Author: Anupama Rao, Assistant Professor, Department of Pediatric Cardiac Surgery, Department of Cardiac Anesthesia, Hi-Tech Medical College, Pandara, Bhubaneshwar, Odisha 751025, India.

E-mail: : anupama.rao16@gmail.com

Case report

A 4 year old male child was brought with complaints of breathlessness on exertion. At presentation he was afebrile, RR-20/mm, BP-90/60 mmhg. General physical examination was normal. Respiratory system showed few crepitations bilaterally. Cardiovascular examination showed normal heart sounds with systolic murmur grade 3/6 in left Intercostal space, best heard in 3rd ICS without any radiation. Other systemic examination was normal and routin hematological investigations were also normal. Chest X ray was normal. ECG showed evidence of right ventricular hypertrophy. On 2D echo evaluation, he was found to have Double chambered right ventricle with valvular pulmonary stenosis. Cardiac catheterization was done as per institutional protocol, which revealed Anomalous LAD coronary artery from pulmonary artery. Coronary angiography showed a left main coronary artery that originated from the left sinus of Valsalva of the aortic root, and continued with a large left circumflex artery. The right coronary

artery (RCA) was found to originate from the right sinus of Valsalva of the aortic root, and was normal. Both RCA and LCX gave collateral vessels that were retrogradely filling LAD draining into PA. The patient underwent DCRV repair , pulmonary valvotomy and ALCAPA repair (Takeuchi repair). His post operative recovery was uneventful and discharged under stable condition. (Fig. 2 and 3)

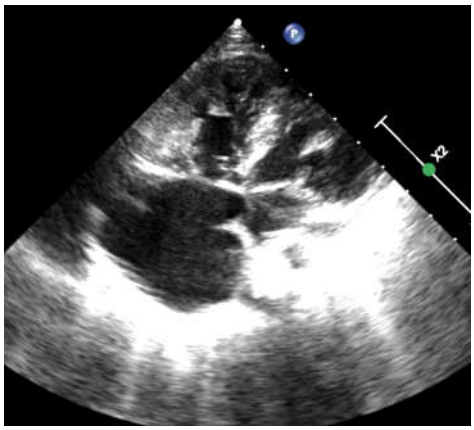

Fig. 1: 2D echo showing double chambered right ventricle.

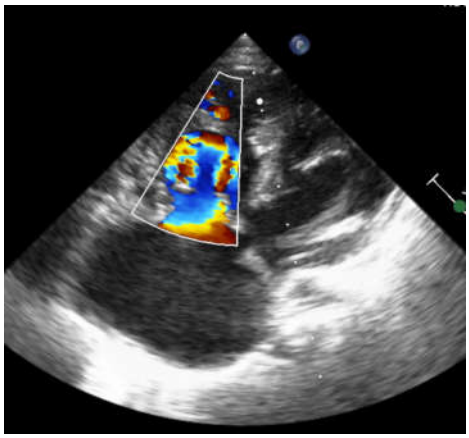

Fig. 2: Colour Doppler showing RV obstruction.

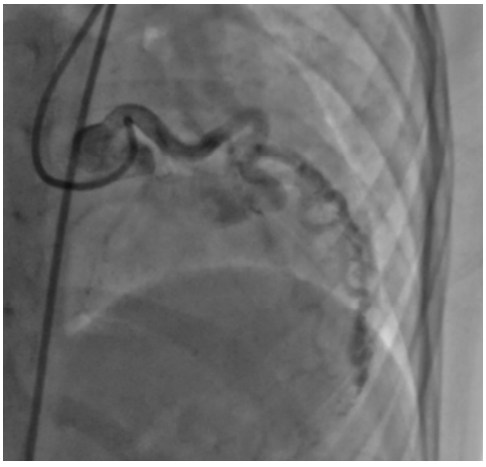

Fig. 3: PA retrogradely filling by collaterals from LCx , which has aortic root origin.

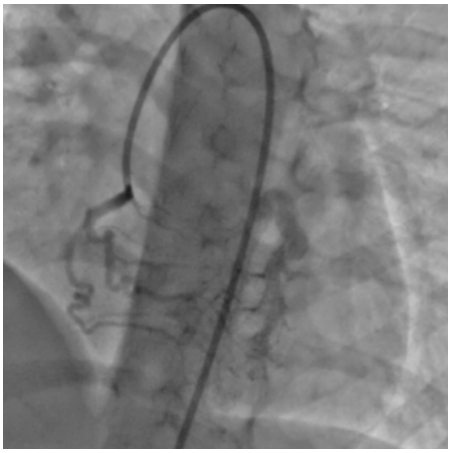

Fig. 4: RCA giving collaterals to LAD.

Discussion

DCRV is a rare congenital cardiac anomaly. The level of obstruction could be in proximity to apex or close to infundibulum. The stimulus for hypertrophy of anomalous muscle bundle in RV is attributed to increased blood flow and pressure within the right ventricular outflow in patients with VSD with increased pulmonary blood flow.[1] The right ventricle is divided into a proximal high-pressure chamber and a distal low- pressure chamber by abnormal muscle bundle.[5] The most common association is VSD,[6] other lesions being atrial septal defect, Sub aortic membrane, coarctation of aorta, sinus of valsalva aneurysm, pulmonary stenosis etc. There is not much data available on association of DCRV with anomalous LAD from pulmonary artery. ALCAPA is a rare congenital anomaly. Without surgery, around 85% of patients with ALCAPA die before reaching adulthood.[7] Patients usually present in the first year of life with signs and symptoms of heart failure or sudden cardiac death.[8] ALADAPA is rarer. There is risk of ischaemia of LAD territory, however, risk is lower than ALCAPA due to collateralization from LCx and RCA. (Fig. 4) Inspite of benign clinical course, ALADAPA still has risk of sudden death. The indications for surgical intervention include right ventricular pressure gradient >20 mmHg by echo-cardiography or by cardiac catheterisation; an angio-graphic evidence of intra-right ventricular obstruction and surgical confirmation of DCRV during the operation.[9]

Conclusion

DCRV is a rare congenital cardiac defect. Though

DCRV is most commonly associated with a VSD, it's rare associations have to be considered during evaluation. Cardiac catheterization may help to rule out less common associations including coronary anomalies which could be missed on 2D echocardiography. This may help to prevent the complications or surprises during surgery. Planned surgical correction of both the lesions improve the outcome.

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