

An Asymptomatic Double Outlet Right Ventricle (DORV).

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ABSTRACT

We report a case of DORV in an 18-year-old female patient presented to NIMS Medical College and Hospital with unrelated symptoms of urinary tract infection. Diagnosis of this case is made by 2D-echocardiography with cardiologist consultation.

Keywords: DORV, Large VSD, TGA, cyanosis, single ventricle.

INTRODUCTION

At normal heart, the pulmonary artery (PA) arises from the right ventricle carrying deoxygenated blood to the lungs for oxygenation.

This oxygenated blood travels back to the left side of the heart, which is carried away to body organs by the aorta arising from left ventricle.^[1] Double outlet right ventricle (DORV) is characterized by a malposition of the great arteries in which both aorta and pulmonary artery arises from right ventricle, the only way of blood outflow from left ventricle is through ventricular septal defect (VSD).^[2]

The word 'double outlet' was used to describe a ventriculoarterial connection in which >50% of both arterial valves are connected to same ventricle. Depending on the location of the VSD, the degree of malposition of the great vessels and the presence of right ventricular outflow tract obstruction, DORV can mimic clinically an unrestrictive VSD, a tetralogy of fallot (TOF) or a transposition of great arteries(TGA),

with which its classification and treatment varies.^[3,4] Generally patients with this heart disease presents with variety of clinical manifestations like shortness of breath, cyanosis, failure to thrive etc., but our patient has no bothersome symptoms related to heart disease. Hence, this has become a point of interest in our case.

Rarity: DORV affects between 1% & 3% of people born with congenital heart defects.^[5]

On routine auscultation, we found holosystolic murmur on left lower border of sternum and mitral area radiating to whole pericardium increasing with inspiration. On further asking the patient's attendants, they gave a history of recurrent chest infections, breathlessness & cyanotic spells and frequent hospitalizations during childhood. However, after the age of 5 years patient remained asymptomatic till date. On examination her pulse rate was 96/minute regular, normal volume and character; blood pressure was 110/60 mm of Hg in right upper limb supine position. Her SpO₂ was 88%, 89%, 93%, 94% in right upper limb, left upper limb, right lower limb and left lower limb respectively. Her height was 157 cm, arm span 155 cm, no marfanoid features and her weight was 40 kg. There are no signs of pallor, icterus, clubbing, lymphadenopathy & edema on systemic examination- apex beat is in 5th intercostal space 2 inch lateral to mid clavicular line, pansystolic murmur heard all over the pericardium, best heard at left lower sternal border during inspiration. Chest radiogram revealed cardiomegaly. Henceforth, 2D-echocardiogram was advised with cardiology consultation, which revealed double outlet right ventricle with transposition of great vessels with large VSD [Figure 1]. She was advised open-heart surgery with high risk, but asymptomatic nature of the disease preferred the attendants not to go under knife at present. Hence, she was discharged on request [Figure 2].

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CASE REPORT

An 18 year old female patient came to medicine OPD with complaints of fever, pain in lower abdomen, burning micturition for 3 days, which was diagnosed as urinary tract infection (UTI) and treated.



Figure 1: 2D echo suggestive of DORV



Figure 2: Patient

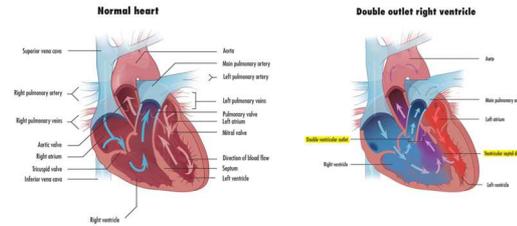


Figure 3: Blood flow in normal heart and DORV

DISCUSSION

Society of Thoracic Surgeons(STS) and European Association of Cardiothoracic surgery (EACTS), Association of European Pediatric Cardiology (AEPC), defined DORV into 4 anatomic subtypes based on relationship of VSD with great vessels and the presence of Right ventricle outflow tract obstruction (RVOTO) [Table 1].^[6-9]

- 1) VSD- type
- 2) Fallot- type
- 3) TGA- type
- 4) Non- committed VSD- type.

Table 1: Classification of DORV types

DORV TYPE	PERCENTAGE OF TYPE	TREATMENT ¹⁰	SURGICAL MORTALITY ¹⁰
VSD-type (DORV with sub aortic VSD)	50-70%	biventricular repair	<5%
Fallot type (DORV with sub aortic or double committed VSD and pulmonary outflow stenosis)	<5%	Complete repair with an arterial switch operation and a VSD-PA baffle	6%
TGA-type (taussig type- DORV with a sub pulmonary VSD)	10-30%	TAPVR repair with an accompanying BT shunt.	11%
Non committed VSD type (DORV with a remote VSD)	10%	Fontan procedure	9%

Cardiologists diagnosed this case as TGA-type based on 2D- echocardiogram findings [Figure 3]. The reason for the asymptomatic nature of the disease is due to development of PAH, once PAH is established it is expected to come out with full-blown symptoms of cyanotic heart disease, which could be fatal.^[9,10]

CONCLUSION

DORV is a rare congenital heart disease with dismal prognosis, surgical intervention is the only treatment of choice, ideally such patients should be operated in neonatal period itself to prevent long term consequences. It is very much astonishing to see a patient with DORV without any symptoms. One thing, which I learnt from this patient, is not to miss auscultation in any patient in medical outdoor.

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