

Double Outlet Right Ventricle with Pulmonary Valve Stenosis in a Young Girl: A Letter to the editor

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Dear Editor,

Double outlet right ventricle (DORV) includes abnormal junctions between the ventricle and arteries, which both the aorta and pulmonary artery originates from the right ventricle structurally.¹ About 1 to 1.5 percent of patients with congenital heart diseases may have this rare condition. It may be associated with some other congenital anomalies. Some types which are more frequent variants include DORV are: Pallot type, Taussig-Bing malformation and Eisenmenger anomaly, and less frequent are associated with: Non-committed Ventricular septal defect (VSD) and ect.² It requires to be stated that the written consent form was taken from the patient before decision to write this article. Also, no conflict of interest exists.

A 19-year-old girl referred to the cardiology clinic of Fatemeh-Zahra hospital Sari-Iran with chief complaint of palpitation. At first a complete history was taken. She had not any past history of dyspnea, weakness followed by activity, syncope and chest pain. On cardiovascular examination, distal pulses were fully palpable. The first and second heart sounds, S1 and S2 were normal, but a systolic sufl in left intercostal space and left boarder of the sternum was sound. There were no evidence of clubbing and acrosianosis in physical examinations. Lung auscultation was normal too. Echocardiography was performed for evaluating of the cardiac chambers, which revealed extreme hypertrophy of right ventricle (RV), double chamber RV due to muscular band, severe stenosis of RVOT, sub pulmonic PS (PG pick-120 mmHg), no Atrial septal defect (ASD), Patent Foramen Ovale (PFO) and Partially Anomalous Pulmonary Venous Connection (PAPVC). Also a dumb shape pulmonary valve, severe interventricular septum (IVS) hypertrophy with paradoxical and abnormal movements due to

extra load of right ventricle pressure was seen (**Fig.1**). A double chamber right ventricle that is divided by a fibromuscular layer and also pulmonary valve stenosis were seen in ventriculography (**Fig.2**). Therefore, based on these results and for the confirming of ventricular problem and pulmonary valve stenosis in this patent, we decided to perform a right anterior oblique (ROA) angiography, which revealed two proximal and distal chambers in the right ventricle (**Fig.3**).

The patient was successfully treated through the staged surgical interventions. Various cardiac anomalies accompanied by DORV. It can be said the most frequent associated heart anomaly is VSD, that observed in 63-77% of the patients with DORV.³ Also other associated anomalies include valvar pulmonary stenosis, atrial septal defect, tricuspid valve regurgitation, aortic valve regurgitation, ruptured sinus of valsalva aneurysm, tetralogy of Fallot, transposition of the great vessels, and Ebstein anomaly.⁴ In our case VSD was not seen and out of these associated anomalies just pulmonary stenosis and right ventricular hypertrophy was seen. The nature of DORV contains an inclination towards obstruction progression.⁵ Patients can develop severe and different symptoms like easy exhaustion, shortening of breath, recurrent chest infection, while in our case no symptom existed except palpitation.⁶ Calcification of valves, especially tricuspid valve was not observed in our case, although this finding has been reported in some cases. Reports of calcification in the tricuspid valve are very rare.⁷ The mechanism of calcification is thought to be because of the high pressure or excessive volume overload of the right ventricle associated with congenital disorder.⁸ Most cases are associated with pulmonary stenosis that this characteristic also was observed in our study.^{5, 6}

DORV is a rare cardiac malformation associated with high mortality and guarded outcomes from residual lesions. Diagnosis of this abnormality is difficult, especially based on the physical examination. So performing more paraclinic evaluations in patients with cardiovascular problems and having in mind of

rare cases can help physicians in more rapid and exact diagnosis. Based upon the review of literature, this is the first such reported case in Iran.

Figure1.



Figure 2.

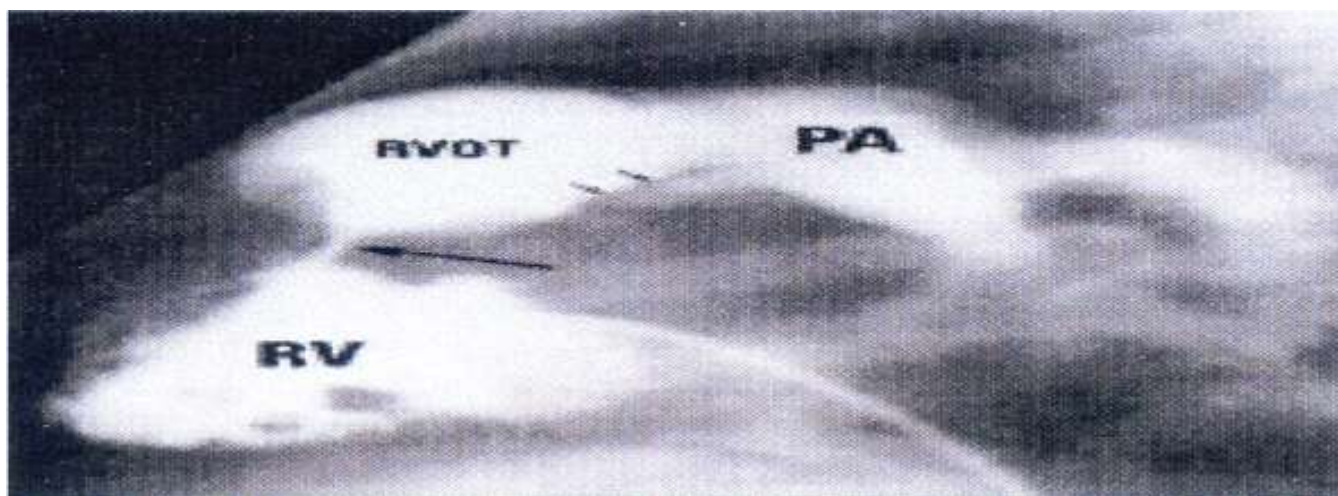
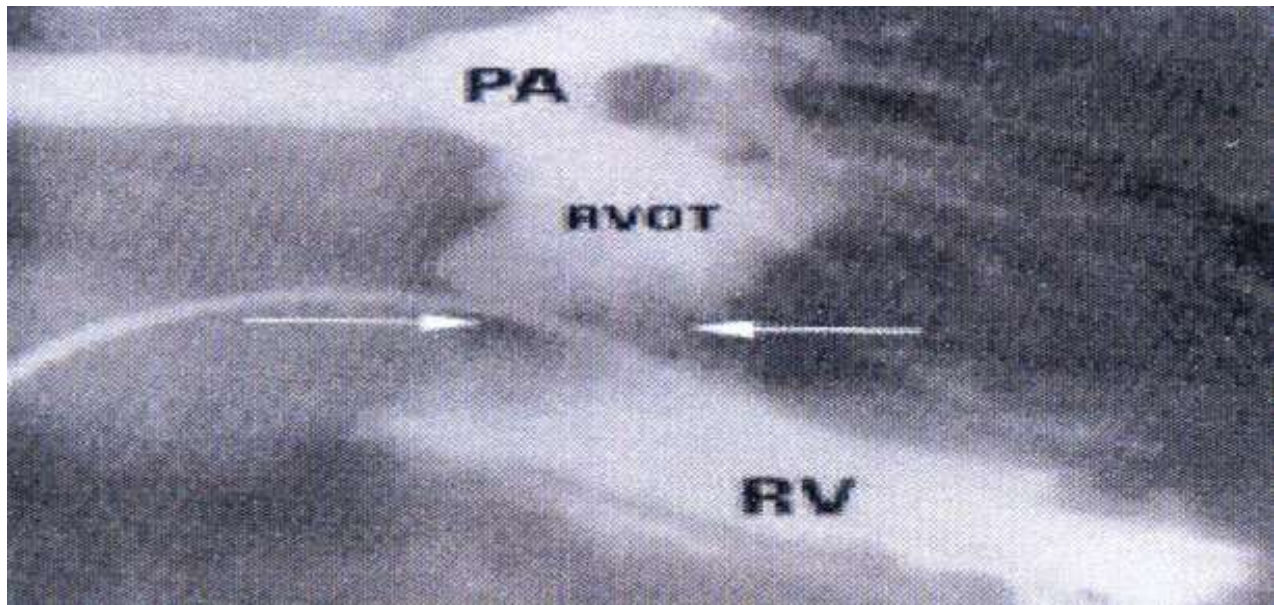


Figure3.



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