Cardiology in the Young

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A rare ventriculoarterial connection: double outlet of both ventricles

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Brief Report

Cite this article: Genc HZ, Ozyilmaz I, Baş S, Kose B, Ozturk E, Hatemi AC, and Tanidir IC (2023) A rare ventriculoarterial connection: double outlet of both ventricles. *Cardiology in the Young* **33**: 2390–2392. doi: 10.1017/S1047951123001130

Received: 18 March 2023
Revised: 6 April 2023
Accepted: 19 April 2023
First published online: 19 May 2023

Keywords:
Congenital cardiac malformations; double outlet of both ventricles; echocardiography; CT angiography; 3-dimensional modelling

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Abstract

Ventriculoarterial connection is one of the important points of the segmental approach to congenital cardiac malformations. Double outlet of both ventricles is a rare form where both great arterial roots override the interventricular septum. In this article, we aimed to draw attention to this very rare form of ventriculoarterial connection by presenting an infant case diagnosed using echocardiography, CT angiography, and 3-dimensional modelling.

In the presence of CHDs, the forms of ventriculoarterial connection show diversity. Due to this diversity, there are difficulties in the definitions, classifications, and treatment options in the reported series.

Double outlet of both ventricles is a rare form of ventriculoarterial connection where both great arterial roots override the interventricular septum and are equally associated with both ventricles.[1]

In this article, we aimed to draw attention to this very rare form of ventriculoarterial connection by presenting an infant case diagnosed as double outlet of both ventricles using echocardiography, CT angiography, and 3-dimensional modelling.

Case

A two-month-old male infant, weighing 5.5 kg, applied to five different centres because of rapid breathing and hearing murmur. He was referred to our clinic because he had different diagnoses as double outlet right ventricle, double outlet left ventricle, and ventriculoarterial discordant connections in terms of ventriculoarterial connection. On physical examination, there was a 1/6 systolic murmur. The room air oxygen saturation was 88%. Electrocardiogram showed normal sinus rhythm with right axis deviation. In transthoracic echocardiography, situs was solitus and atrioventricular connection was concordant. In the ventriculoarterial connection, it was observed that both great arteries were arising from the right ventricle at some views, from the left ventricle at some views and were discordant at some views (Fig 1). When the direction of the echocardiography probe was changed, it was observed that the outlets of the aorta and pulmonary artery changed from the right ventricle to the left ventricle. It was observed that the outlet septum was placed perpendicular to the muscular ventricular septum, so that both great arteries became associated with both ventricles. The pulmonary artery was placed to the left of the aorta, as side-by-side (Supplementary Video-1). There was no atresia or stenosis in both atrioventricular valves. A subaortic-subpulmonic 9.4 mm defect with laminar bilateral shunt was present in the perimembranous outlet region. Patent foramen ovale with left-right shunt was present in the interatrial septum. There was no stenosis in the left and right ventricular outflow tract. Arcus and descending aorta diameters were normal. Pulmonary artery z scores were normal. Infundibular branch of the right coronary artery crossing the right ventricular outflow tract was observed.

Additional CT angiography was performed to detail the anatomical condition of the patient. Similar to echocardiography, it was revealed that there was subaortic and subpulmonic wide ventricular septal defect, and both aorta and pulmonary artery were overriding with equal dominance with the interventricular septum (Fig 2). Preoperative 3D modelling showed that the outlet septum was placed perpendicular to the interventricular septum and both ventricles were equally related to the aorta and pulmonary artery (Supplementary Video-2, Fig 3). Biventricular repair was planned with a two-patch routing and a tunnel between the left ventricle and the aorta.

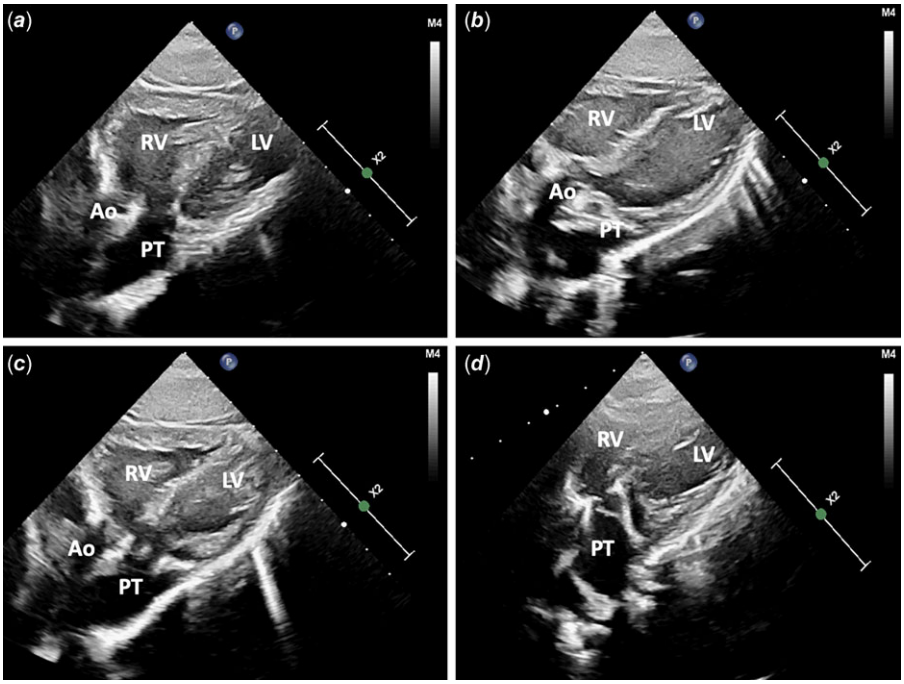

Figure 1. Transthoracic two-dimensional echocardiographic images. (***a***) The angle that suggest double outlet right ventricle in subcostal view. (***b***) The angle that suggest double outlet left ventricle in subcostal view. (***c***) The angle that suggest discordant ventriculoarterial connection in subcostal view. (***d***) The angle that suggest concordant ventriculoarterial connection (right ventricle-pulmonary artery) in subcostal view. Ao=aorta; PA=pulmonary artery; RV=right ventricle; LV=left ventricle.

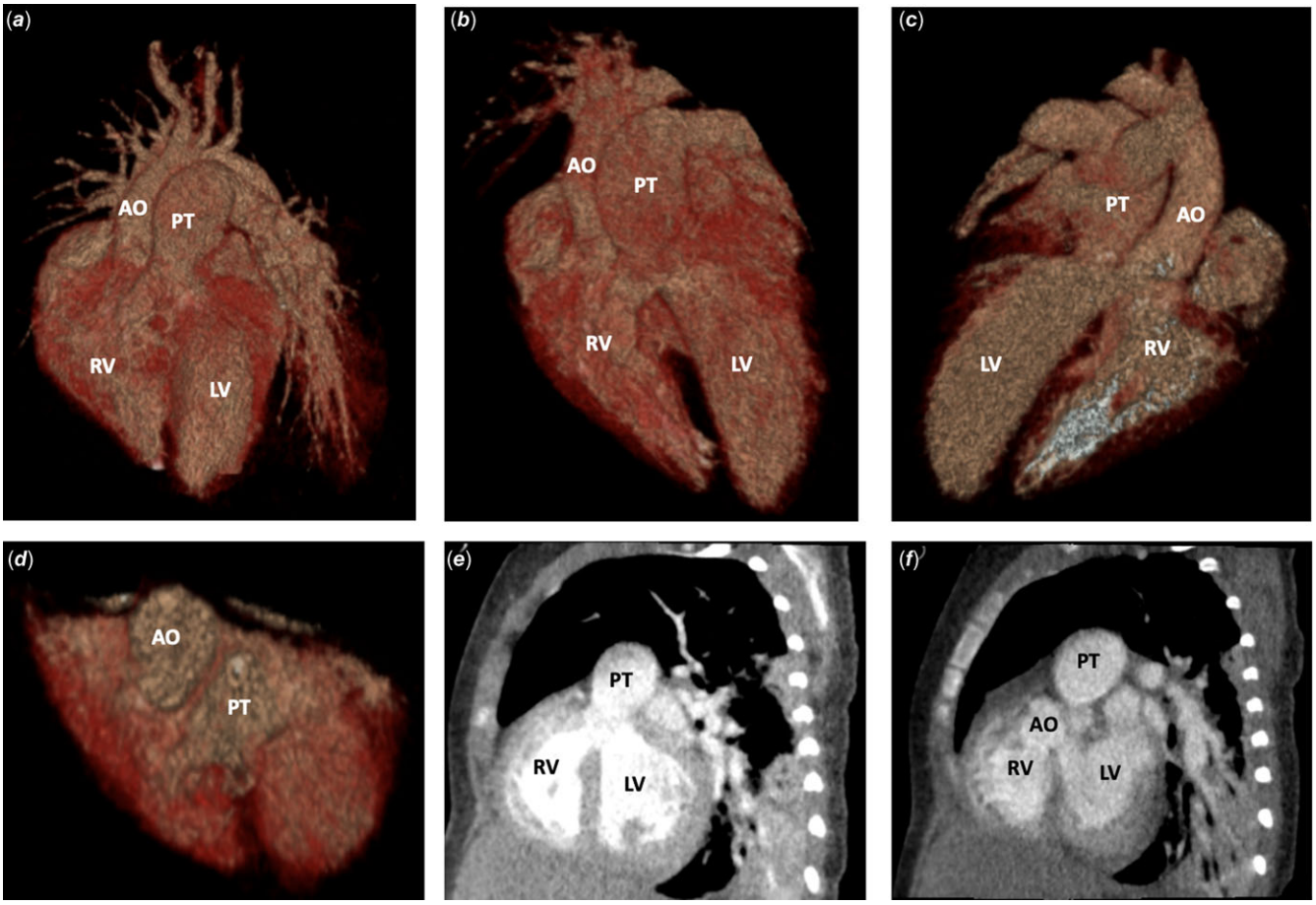

Figure 2. Computerized tomography (CT) angiography images. (***a*** and ***b***) Volume Rendering Technique (VRT) images showing the anterior plane of the heart. (***c***) VRT image showing the posterior plane of the heart. (***d***) VRT image showing the axial plane of the heart. (***e***) Sagittal plane showing the pulmonary artery overrides to both ventricles. (***f***) Sagittal plane showing the aorta overrides to both ventricles. Ao=aorta; PA=pulmonary artery; RV=right ventricle; LV=left ventricle.

https://doi.org/10.1017/S1047951123001130 Published online by Cambridge University Press

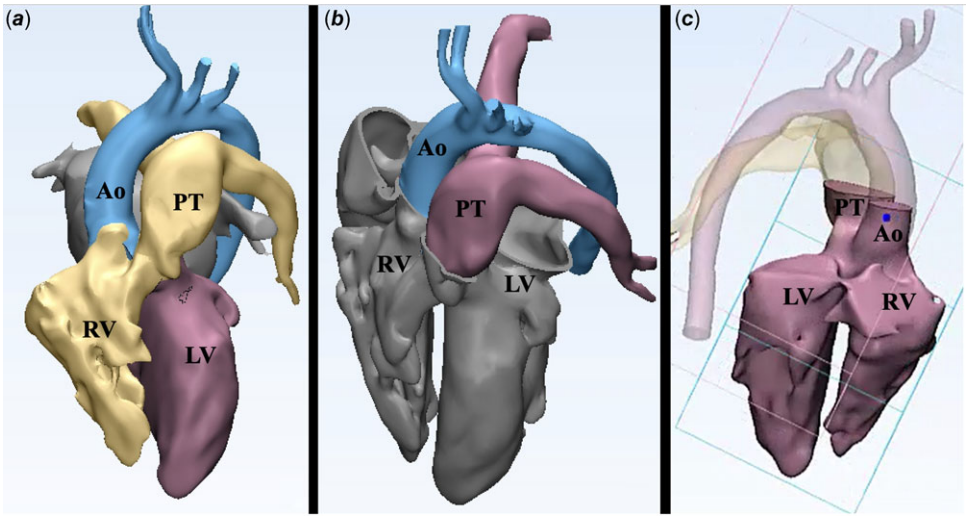

Figure 3. Three-dimensional modeling images. (***a***) Anterior view. (***b***) Craniocaudal view from above. (***c***) Posterior view. Ao=aorta; PA=pulmonary artery; RV=right ventricle; LV=left ventricle.

Discussion

The segmental approach to complex congenital cardiac malformations is widely accepted by many authors. Ventriculoarterial connection is one of the important points of this assessment. Basically, it is grouped as concordant, discordant, double outlet, or single outlet.

Double outlet ventricles are malformations in which both arterial roots arise entirely or predominantly from one of the ventricles. In these cases, interventricular septal defect is usually accompanied. In a rare form, the defect is seen as doubly committed, the outlet septum is fibrous or rudimentary. So, the defect becomes associated with both arterial roots. This pathology was first described by Brandt et al.[1] It was later described by Ueda and Becker[2] and Aiello et al.[3] It is sometimes difficult to define this anatomical condition with echocardiography, and it may lead to different diagnoses of the cases as double outlet right ventricle, double outlet left ventricle, and transposition of the great arteries. As a matter of fact, in our case, it was associated with all three conditions.

Different clinical findings such as growth retardation, cyanosis, feeding difficulties, and recurrent respiratory tract infections can be observed in the cases.

Our case also had rapid breathing. He had a history of hospitalisation once due to lower respiratory tract infection.

Echocardiography can be helpful in diagnosis. In the series of six cases by Iver et al, large interventricular communication and side-by-side great artery relationship were observed in all cases.[4] Our case also had a similar feature. In addition, appropriate surgical planning can be performed by confirming the diagnosis with CT angiography and 3-dimensional modelling.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951123001130.

Financial support. This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Competing interest. None.

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