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CASE REPORT

A Rare Case of Double Outlet Right Ventricle with Pulmonary Atresia and Aortopulmonary Window

ABSTRACT

Double outlet right ventricle (DORV), a congenital heart abnormality, occurs when the aorta and pulmonary artery are predominantly or entirely connected to the morphological right ventricle. Classified based on the relationship between the ventricular septal defect (VSD) and the great arteries, DORV exhibits diverse anatomical and hemodynamic variations resembling VSD, tetralogy of Fallot (TOF), transposition of the great arteries (TGA), and functional single ventricle. In this case, we present a coexistence of aortopulmonary window (APW) in a patient with DORV-pulmonary atresia.

Keywords: Aortopulmonary window, double outlet right ventricle, pulmonary atresia

CASE REPORT

An eight-month-old male patient, previously monitored in the neonatal intensive care unit for cyanotic congenital heart disease after birth, and who had not attended regular follow-up appointments after discharge, was referred to our center. During the physical examination, the patient exhibited cyanosis, and non-invasive oxygen saturation measurement showed a level of 82%. Echocardiography revealed an intact interatrial septum and a 13 mm large perimembranous outlet ventricular septal defect (VSD) in the interventricular septum. The echocardiogram also showed that the aorta was overriding the VSD by 80%, characterized by two cusps and a wide structure. Notably, no pulmonary artery structure was observed emanating from the right ventricle, and a muscular barrier was identified between the right ventricle outflow tract and pulmonary artery. Furthermore, a type 1 aortopulmonary window (APW) was identified at the supravalvular level, positioned to the left of the aorta and situated above the barrier between the right ventricle and pulmonary artery (Fig. 1). To provide a more precise diagnosis and pathology definition, computed tomography angiography (CTA) was conducted, incorporating 3D modeling.

The CTA images revealed that the aorta was predominantly separated from the right ventricle, overriding the VSD, and no main pulmonary artery structure originating from the right ventricle was detected. However, at the supravalvular level, an anatomical anomaly involving a pulmonary artery structure was identified adjacent to the left lateral aspect of the aorta, presenting an approximate 8 mm diameter defect between the aorta and the pulmonary artery (Fig. 2). Additionally, there appeared to be dilation in the ascending aorta, measuring 25 mm in diameter. Consequently, based on these findings, the diagnosis of double outlet right ventricle (DORV)-pulmonary atresia accompanied by an APW was confirmed.

Based on this information, a decision was made to proceed with surgical correction. The defect in the ascending aorta was directly repaired, followed by a vertical incision in the right ventricle, through which the ventricular septal defect was closed with a Dacron patch. After this procedure, continuity between the right ventricle and the bifurcation of the pulmonary artery was established using a 12 mm valved xenograft conduit (Contegra®). At the end of the procedure, cardiopulmonary bypass support was gradually weaned; however, it could not be discontinued due to inadequate cardiac output and oxygenation. Consequently, the patient was transitioned to extracorporeal membrane oxygenation (ECMO) support and transferred to the intensive care unit. In the intensive care unit, in addition to

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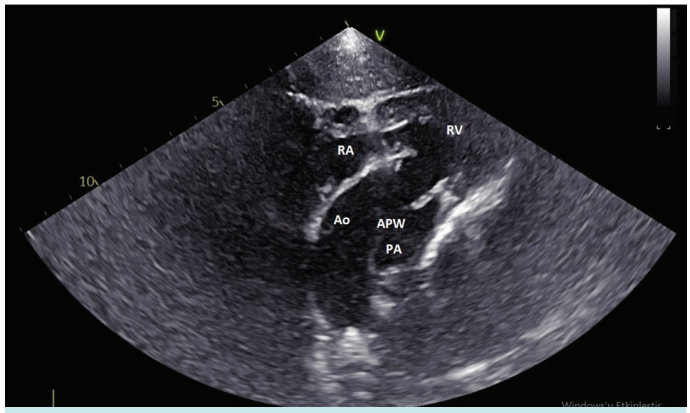

Figure 1. Transthoracic echocardiography showing the aorta, atretic pulmonary valve and pulmonary artery, and aortopulmonary window on subcostal long axis view.
RA: Right atrium; RV: Right ventricle; Ao: Aorta; PA: Pulmonary artery; APW: Aortopulmonary window.

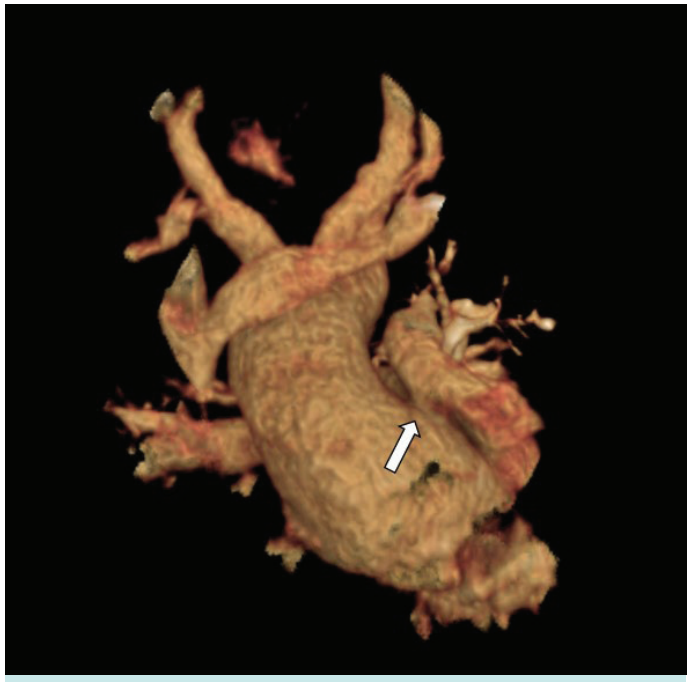

Figure 2. CTA image of the aortopulmonary window.

ECMO and mechanical ventilation support, treatment with dopamine (10 µg/kg/min), adrenaline (0.1 µg/kg/min), and milrinone (0.5 µg/kg/min) was continued. Subsequent echocardiographic evaluation revealed minimal tricuspid insufficiency along with diminished ventricular functions, and confirmed the patent status of the right ventricle to pulmonary artery conduit. During monitoring under ECMO support, the patient, who exhibited elevated acute phase reactants and developed sepsis, succumbed on postoperative day 28.

DISCUSSION

DORV is a rare congenital heart disease where both major arteries are primarily or entirely connected to the right ventricle, constituting about 1% of all congenital heart diseases (1-3). DORV was first described pathologically by Witham et al. (4) in 1957. Goor et al. (5) provided a definition of DORV in 1982, stating that it involves both great arteries arising from the morphologic right ventricle by 50% or more and the presence of an interventricular connection such as VSD or atrioventricular septal defect (AVSD) (6). The classification of DORV by Lev et al. (2), who classified DORV into four main groups as subaortic, subpulmonic, double committed and non-committed VSD types using the relationship of the VSD to the great arteries, remains the most widely used classification of DORV.

The diverse anatomical combinations of DORV can result in either decreased or increased pulmonary blood flow. Variations in pulmonary blood flow can lead to a wide range of clinical presentations, ranging from cyanosis to congestive heart failure and even pulmonary edema. In patients with subaortic or subpulmonary VSD, aggressive diuresis may be required due to pulmonary over-circulation, which can lead to the need for intubation. If pulmonary blood flow is very low, prostaglandin E1 infusion and/or atrial septostomy may be necessary. Some patients may have sufficient pulmonary blood flow and can be discharged for weight gain and growth until surgical intervention is required (6).

Although differential diagnoses for a single cardiac outflow tract include truncus arteriosus, pulmonary atresia with VSD or intact ventricular septum, the association of DORV with pulmonary atresia is also seen rarely. When such rare anatomy is observed, obtaining information about which route supplies pulmonary blood flow, such as aortopulmonary collaterals or patent ductus arteriosus (PDA), is crucial for the management of the disease (7).

Our patient presented with a rare combination of double outlet right ventricle (DORV) and pulmonary atresia. Unlike the typical presentation of DORV with pulmonary atresia, which usually includes a patent ductus arteriosus (PDA) or aortopulmonary collaterals, the pulmonary vascular bed in this case was supplied through the aortopulmonary window (APW). The APW develops when the two opposing conotruncal ridges fail to merge as they should, which typically divides the truncus arteriosus into the aorta and pulmonary artery. It can be observed not only just above the semilunar valves but also in more distal regions of the ascending aorta and in the main pulmonary artery (8). It manifests both independently and in conjunction with other congenital heart anomalies such as patent foramen ovale (PFO), atrial septal defect (ASD), PDA, TOF, aortic arch interruption, aortic arch hypoplasia, and anomalous origin of coronary arteries (9).

Our case holds significance owing to the alteration in pulmonary blood flow compared to the conventional supply routes in pulmonary atresia with DORV, along with its divergence from typical congenital heart diseases linked with the APW. Moreover, there is notable similarity observed in the anatomy and morphology of our case to type 1 truncus arteriosus. The identification of a dysplastic, bicuspid, and wide aortic valve might suggest the possibility of truncus arteriosus; however, the presence of a second outflow tract originating from the right ventricle confirms the diagnosis of DORV with pulmonary atresia accompanied by an APW.

DORV, a rare congenital heart disease with various anatomical features and consequently different treatment methods, has many

subgroups. It should not be considered as a single disease, and morphology should be well defined. Detailed and unequivocal characterization of cardiac anatomy is crucial for appropriate diagnosis and surgical decision-making. Despite our efforts to monitor and provide prolonged supportive treatments in the intensive care unit (ICU) due to the complex pathophysiology and surgical interventions of the disease, we unfortunately lost our patient due to an infection. However, we believe that presenting this rare anatomy in our case report and increasing awareness on this matter will contribute to the survival of other patients.

Informed Consent: Informed written consent for the publication of the case report and accompanying images was obtained from both the patient›s father and mother.

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