

# Double-Outlet Left Ventricle: The Importance of Echocardiographic and Computed Tomographic Assessment



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## INTRODUCTION

Double-outlet left ventricle (DOLV) is a very rare congenital cardiac anomaly in which both the aorta (Ao) and pulmonary trunk arise entirely or predominantly from the left ventricle (LV).<sup>1</sup> It represents a heterogeneous group of patients with various clinical presentations, associated cardiac abnormalities, and long-term outcomes.<sup>2</sup> We report the case of a newborn with DOLV, who presented with cyanosis within the first day of life.

## CASE PRESENTATION

This male infant was born at 41 weeks of gestation via a normal, spontaneous vaginal delivery after an uncomplicated pregnancy. Birth weight was normal, and he did not require any resuscitation maneuvers (Apgar 8/8). However, he became hypoxic within the first 24 hours of life. On physical examination, he was cyanotic (O<sub>2</sub> saturation 70%) but well perfused. He had a normal heart rate and a regular rhythm. There was a III/VI systolic ejection murmur audible at the left lower sternal border. Chest radiograph showed a normal cardiac silhouette, cardiothoracic ratio, and pulmonary markings. An electrocardiogram exhibited normal sinus rhythm and nonspecific T wave changes.

A transthoracic echocardiogram was performed on the second day of life, which revealed situs solitus of the atria and abdominal viscera, levocardia, and atrioventricular (AV) concordance. The great arteries were in a side-by-side orientation, both arising predominantly from the LV (Figure 1, Video 1), with the Ao slightly anterior and rightward, overriding a large membranous ventricular septal defect (VSD; Figure 2; Video 2). The pulmonary artery (PA) arose entirely from the LV (Figure 3, Video 3), the pulmonary valve was bicuspid, and the branch pulmonary arteries were confluent.

Both right ventricular (RV) and LV sizes and systolic function were normal, with a calculated LV ejection fraction of 63% by 2D echocardiography. Both AV valves were normal. A single coronary artery was noted originating from the left anterior sinus, with the right coronary artery coursing anterior to the Ao.

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The patient underwent cardiac computed tomography (CT) with 3D reconstructions to better delineate the relationship of the great arteries to the LV and the course of the coronary arteries for appropriate surgical planning. The great arteries were visualized side by side, originating from the LV. It was noted that >50% of the Ao arose from the LV (Figure 4), overriding the VSD (Figure 5). The PA was confirmed to arise entirely from the LV, with no pulmonary-mitral fibrous continuity (Figure 6). This verified the diagnosis of DOLV.

The relationship of both great arteries to the LV and the mitral valve annulus was better appreciated in the animation from the 3D reconstruction (Video 4). In this animation, the Ao and PA were clearly shown arising from the LV. The two major crescents inside the LV chamber are the papillary muscles. The protrusion perpendicular to the long axis of the LV represents the mitral annulus, and the hollow part behind it is the left atrial chamber.

At 7 days of age, the patient underwent a Rastelli-type repair. The LV was baffled to the aorta through the VSD with patch exclusion of the RV, the main PA was divided, and the pulmonary valve was oversewn. An RV to PA conduit with a 9-mm valved homograft was placed.

## DISCUSSION

Double-outlet ventricles with concordant AV connection account for 1% of all cases of congenital heart disease, and DOLV accounts for <5% of those cases.<sup>1,3</sup> The exact incidence is not known, but <1/200,000 live births have been reported.<sup>1</sup>

DOLV was considered to be an embryologic impossibility until 1967, when Sakakibara *et al.*<sup>4</sup> reported the first successful surgical repair of DOLV, suggesting the existence of this type of ventriculoarterial connection. To date, no single hypothesis has explained the embryology of all known DOLV cases.<sup>1,3</sup> Anomalous differential conal growth, first explained by Paul *et al.* 1970,<sup>5</sup> emphasized that infundibular growth beneath the semilunar valves represents one of the most important factors in the morphogenesis of normal and abnormal relations between the great arteries. This hypothesis implies that bilateral absence of conus is a prerequisite for DOLV.<sup>3</sup> An alternative hypothesis, proposed by Anderson *et al.*<sup>6</sup> in 1974, suggested that the development of DOLV could be explained by anomalous absorption or malorientation of the subarterial portion of the ventricular septum, separating the RV infundibulum from both great arteries.<sup>6,7</sup>

DOLV is defined as a type of ventriculoarterial connection in which both great arteries arise entirely or predominantly from the morphologic LV.<sup>1-3,8</sup> Although it has been reported with either situs solitus or inversus of the atria, concordant or discordant AV connections, with or without intact ventricular septum, and with functionally single-ventricle hearts,<sup>3</sup> it most commonly occurs in the form of atrial situs solitus with AV concordance.<sup>1,3</sup> Similar to double-outlet right ventricle, DOLV is currently classified in terms of the location of the VSD relative to the great arteries. Further variations include the

## VIDEO HIGHLIGHTS

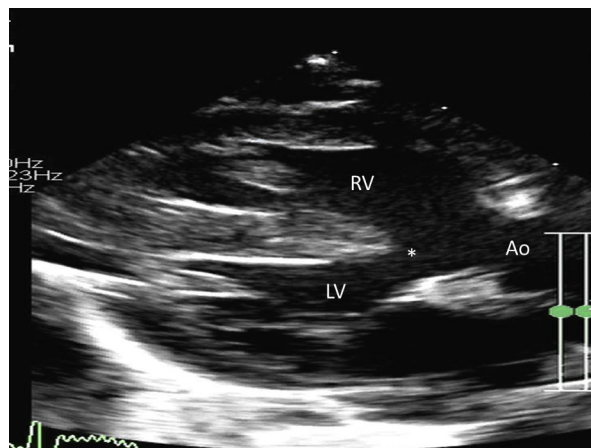
**Video 1:** Transthoracic 2D echocardiography with color Doppler comparison, obtained in the subcostal view, showing laminar flow across the great arteries, both arising from the LV.

**Video 2:** Transthoracic 2D echocardiographic with color Doppler comparison, obtained from parasternal long-axis view. The Ao overrides a large membranous VSD, with left-to-right shunt.

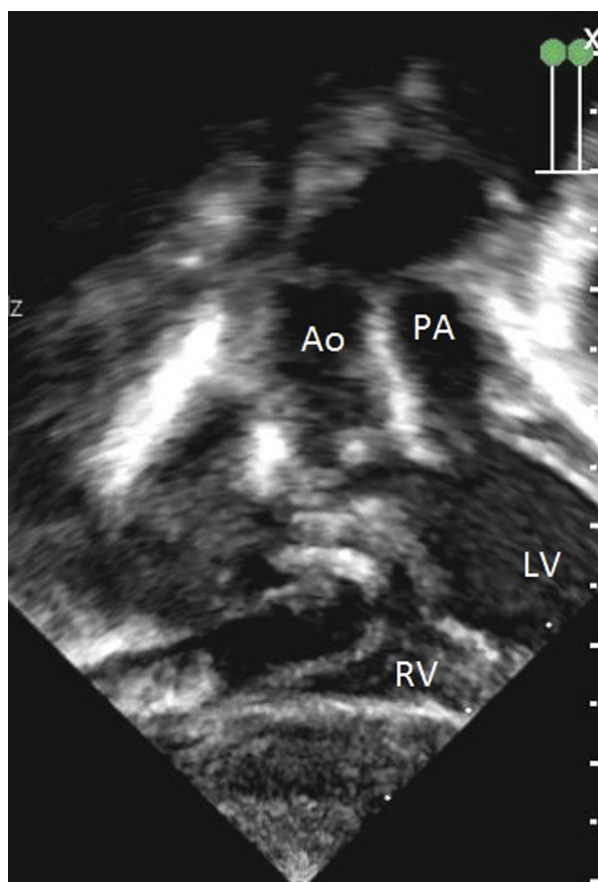
**Video 3:** Transthoracic 2D echocardiography with color Doppler comparison, obtained in a more posterior parasternal long-axis view. The PA is seen arising from the LV.

**Video 4:** Multiplanar 3D reconstruction of CT angiography. The hollow parts are the LV chamber, with the Ao on the right and PA on the left.

View the video content online at [www.cvcasejournal.com](http://www.cvcasejournal.com).



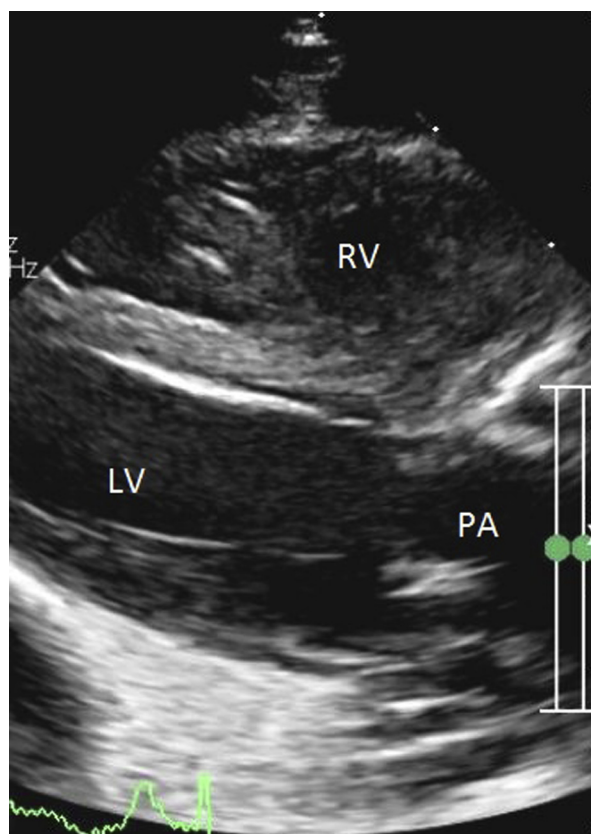
**Figure 2** Transthoracic 2D echocardiographic image in parasternal long-axis view. The Ao overrides a large VSD (\*).



**Figure 1** Transthoracic 2D echocardiographic image obtained in the subcostal view. Both great arteries are seen arising from the LV.

presence of pulmonary and subpulmonary stenosis, the location of subaortic conus, and whether aortic valve stenosis is present.<sup>3,8</sup>

Van Praagh *et al.*<sup>9</sup> provided the most comprehensive review of 109 cases with DOLV. In their series, the most common form of DOLV



**Figure 3** Transthoracic 2D echocardiographic image obtained in a more posterior parasternal long-axis view. The PA is seen arising entirely from the LV.

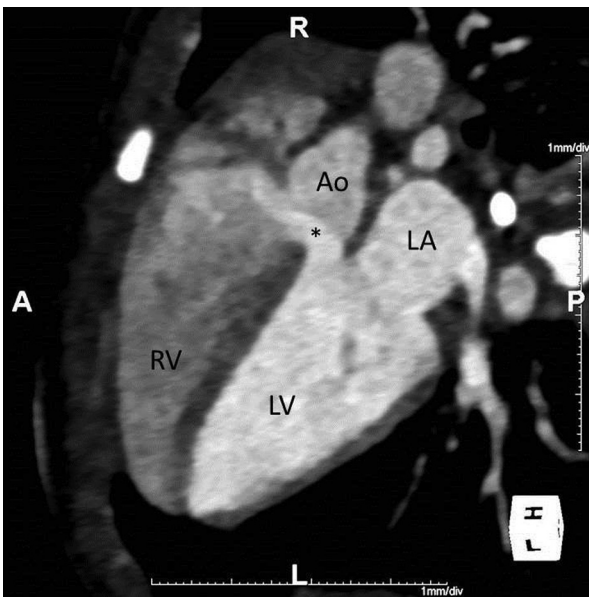
had a subaortic VSD, comprising 48% of the 109 cases, or 73% of the cases with situs solitus and AV concordance. Most frequently, the Ao was rightward and anterior or right and lateral. Furthermore, the most common type also had subvalvar or valvar pulmonary stenosis (Figure 7). This explains why these patients manifest clinical and angiographic findings of tetralogy of Fallot.



**Figure 4** CT angiography. An oblique coronal multiplanar reformatted image shows the Ao and PA in side-by-side orientation, originating from the LV. More than 50% of the Ao is seen arising from the LV.



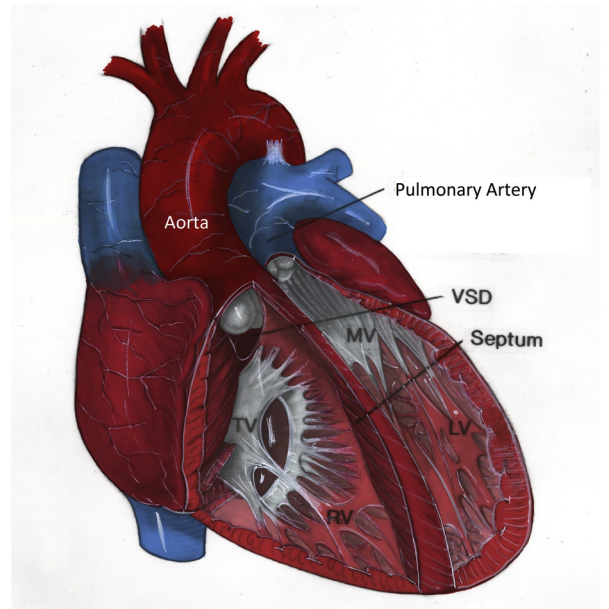
**Figure 6** In the long-axis view, the PA arises entirely from the LV, with no pulmonary-mitral fibrous continuity due to the presence of a part of the subarterial conus (arrow). Ao, Aortic arch; LA, left atrium.



**Figure 5** Multiplanar reformatted image of CT angiography in a long-axis orientation of the LV. The Ao overrides the VSD (\*), with left to right shunt. LA, Left atrium.

When DOLV is associated with a subaortic VSD, a right and anterior Ao, and no significant pulmonary outflow tract obstruction, patients present clinically with transposition physiology. This was the case for our patient. When DOLV presents with a subpulmonary VSD (15% of cases), it has clinical features of a large VSD with pulmonary overcirculation. But more commonly, some degree of outflow obstruction is present in 80% of these cases, including coarctation of the Ao or interrupted aortic arch.<sup>3,8</sup>

After the first surgical correction by Sakakibara *et al.* in 1967,<sup>3,4</sup> other procedures were attempted, such as pulmonary root translocation, RV outflow tract patch reconstruction,<sup>7</sup> and a modified



**Figure 7** Diagram of the most common form of DOLV showing situs solitus and AV concordance, rightward/anterior or right/lateral Ao with subvalvar or valvar pulmonary stenosis. MV, Mitral valve; TV, tricuspid valve.

Lecompte procedure,<sup>10</sup> with variable results. However, later reports emphasized surgical correction of DOLV with a two-ventricle repair by VSD closure and placement of an RV to PA conduit (extracardiac or “Rastelli-type” repair).<sup>3,7,8</sup> If a two-ventricle repair could not be

performed, single-ventricle palliation was preferred. In our case, the preoperative comprehensive echocardiographic and tomographic studies allowed a correct assessment of the great arteries and their relationship to the VSD, enabling a Rastelli-type approach.

## CONCLUSION

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DOLV encompasses a broad spectrum of clinical and pathologic features of various physiologic entities, ranging from simple VSD to tetralogy of Fallot and complete transposition of the great arteries. We presented the case of a newborn who developed cyanosis early in life and had extensive echocardiographic and tomographic assessment permitting a surgical correction during the first week of life.

## SUPPLEMENTARY DATA

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Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2019.04.004>.

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