

A case of double-outlet left ventricle with atrioventricular discordance

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The clinical, echocardiographic and angiographic aspects of a four-month-old boy with double-outlet left ventricle, atrioventricular discordance, L-malposition of the great arteries, ventricular septal defect and pulmonary stenosis are described. Additionally, in this patient, the right arcus aorta and the ligament of ductus arteriosus caused anterior compression of the trachea. The anterior position of the morphological left ventricle is the most interesting feature.

Key words: double-outlet left ventricle, atrioventricular discordance, vascular ring.

Double-outlet left ventricle (DOLV) was not considered compatible with the prevailing theories of embryology¹ until Sakakibara et al.² reported several cases in 1967. Numerous studies, most of which are summarized in the reviews by Van Praagh et al.³ and Otero-Coto et al.^{4,5}, have described the anatomic, angiographic and surgical aspects of this anomaly over the past 30 years⁴. We present a distinct case of DOLV with atrioventricular discordance whose left ventricle was situated anteriorly.

Case Report

A four-month-old boy was referred to the pediatric Cardiology Unit because of cyanosis and systolic murmur. He also had dyspnea, wheezing and stridor. Cardiovascular system examination revealed a grade 3/6 systolic murmur maximal at upper left sternal border. The second heart sound was single. The electrocardiogram revealed QRS axis of 100°, and right ventricle hypertrophy. The chest X-ray showed normal-sized cardiac silhouette with a broad cardiac base and left-sided apex. The pulmonary vasculature was normal.

The two-dimensional echocardiography revealed levocardia, viscerocardiac concordance, normal systemic and pulmonary venous return, and atrioventricular discordance. The anatomic right ventricle was situated to the left of the anatomic left ventricle (L-loop). A prominent subpulmonic conus was causing severe

pulmonary stenosis having systolic gradient of 77 mmHg. The aorta was situated anterior and to the left of the pulmonary artery (L-loop). The infundibular morphology resembled that of congenitally corrected transposition of great arteries, but both arteries arose from the left ventricle. The patient had right-sided arcus aorta (Fig. 1). We also detected the enlarged right atrium, bulging atrial septum towards the left atrium, right-to-left shunt through a small patent foramen ovale, and a large subaortic ventricular septal defect. The echocardiographic diagnosis was “{S, L, L} DOLV, atrioventricular discordance, large ventricular septal defect and pulmonary stenosis with subpulmonic conus”. Figure 2 summarizes the anatomic features of our case.

An aorto-pulmonary shunt procedure was scheduled for the patient; however, he had several attacks of lower respiratory tract infections and an irritating cough suggesting bronchial compression. We performed cardiac catheterization and angiography in order to reveal the anatomic details and demonstrate a possible abnormal vascular structure compressing the airways. The systolic, diastolic and mean pressures in the right-sided ventricle were 82, 4, and 38 mmHg, respectively; those of the aorta were 75, 46, and 62 mmHg. We could not advance the catheter through the posteriorly situated, stenotic pulmonary valve. The oxygen saturation in the aorta was 89%. The radio-opaque solution injected

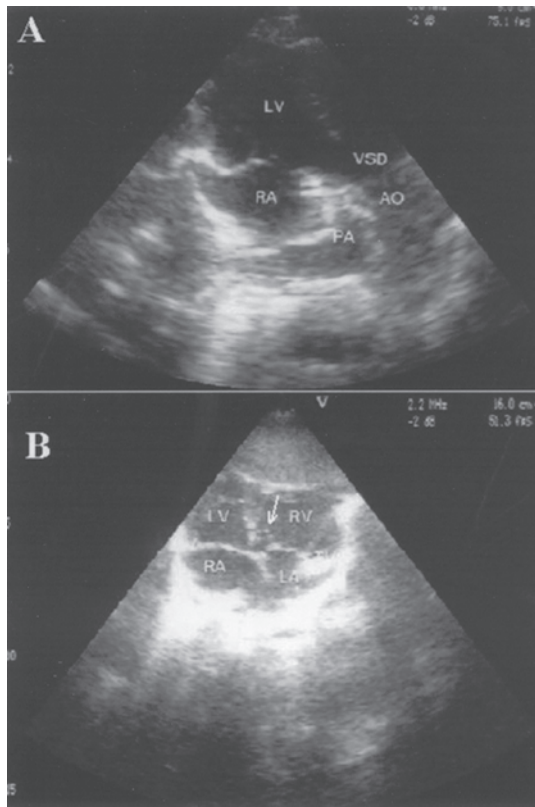


Fig. 1. The echocardiographic images of the patient.
 A) The subcostal long-axis view. The right atrium is connected to the right-sided anatomical left ventricle. Pulmonary artery and the aorta arise from this ventricle.
 LV : left ventricle. RA: right atrium.
 VSD: ventricular septal defect. PA: pulmonary artery, Ao : aorta.
 B) The parasternal four-chamber view. The pulmonary veins drain into the left atrium, which is connected to the left-sided anatomical right ventricle. The arrow depicts the chordal attachment of the septal leaflet of the tricuspid valve on the interventricular septum.
 LV: left ventricle. RV: right ventricle.
 RA: right atrium. LA: left atrium.

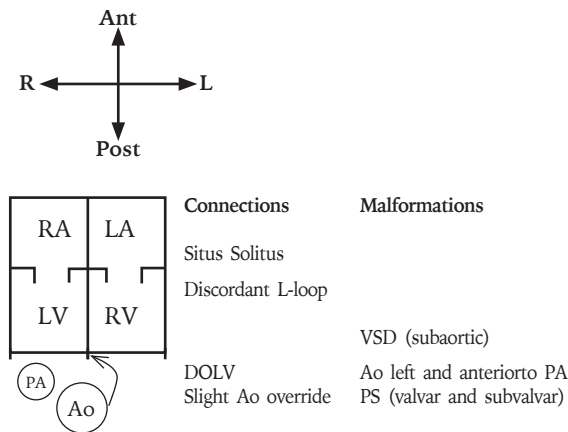


Fig. 2. The anatomic features of the case.

into the anteriorly located smooth ventricle visualized both the aorta and the pulmonary artery arising from this ventricle (Fig. 3). The subpulmonic region and the origin of the main pulmonary artery were very stenotic, whereas the branches were of adequate size. The diameters of the right and left pulmonary arteries were 7.3 mm and 6.3 mm, respectively. The aorta arising from the same ventricle became visible following the demonstration of the pulmonary arteries. Approximately 1 cm distal to the bifurcation, the left pulmonary artery was drawn toward the right-sided arcus aorta by the ligament of the closed ductus arteriosus (Fig. 3). Thus, the ligament of the ductus arteriosus and the right-sided arcus aorta surrounded the left main bronchus,

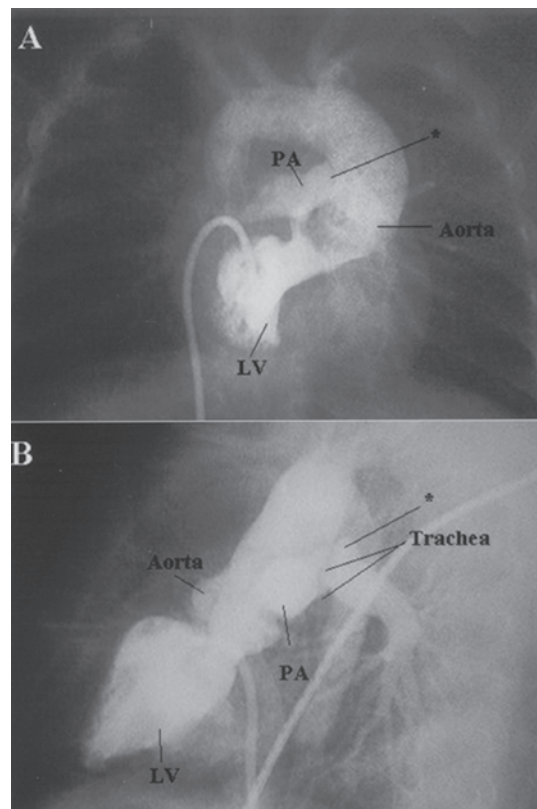


Fig. 3. The angiographic images of the case.
 A) Injection of contrast material into the right-sided (left) ventricle. Both the aorta and the main pulmonary artery arise from this non-trabeculated ventricle.
 B) Injection of contrast material into the anteriorly situated, non-trabeculated ventricle. The aorta is slightly in front of the pulmonary artery.
 LV: left ventricle, PA: pulmonary artery.
 * The ligament of the closed ductus arteriosus pulling the pulmonary artery toward the right-sided arcus aorta.

embracing the air within it. Furthermore, the arcus aorta compressed the trachea by displacing it posteriorly (Fig. 3).

Discussion

DOLV is a very rare congenital heart defect occurring with multiple other congenital heart defects. It has been mostly reported in situs solitus with atrioventricular concordance, but rarely seen in situs inversus totalis with atrioventricular concordance or discordance⁴. There are very few cases of DOLV with atrioventricular discordance^{6,7}. The conal anatomy is also variable. The most common anatomy is near normal deficiency of subpulmonic conal muscle and short, usually stenotic subpulmonary conus. In the majority of cases there is a subaortic ventricular septal defect. A subpulmonary defect or additional muscular defects have been detected^{8,9}. There are also three known cases with intact ventricular septum⁸. Pulmonary or aortic outflow tract obstructions and D-malposition of the great arteries are common with DOLV, whereas L-malposition, with aorta on the left, is a rarer occurrence⁸.

Most of the common features described above were also present in our case. However, our case is an S, L, L type (normal viscerotransverse situs, L-loop of the right ventricle, L-malposition of the great arteries) with atrioventricular discordance. The feature that makes our case unique is the anteriorly placed left ventricle. It is well known that the left ventricles, even when hypoplastic, are always found postero-inferiorly within the ventricular mass¹⁰. Among the previously reported cases of DOLV with atrioventricular discordance^{6,7}, the one described by Subirana et al.⁷ seems to have similar features to our case in terms of the position of the morphologic left ventricle. In that particular case, the morphologic left ventricle also lay unusually anteriorly and bulged into the morphologic right ventricle, as in our case⁷.

Moreover, our case had associated features of an incomplete vascular ring. The compression of the trachea and the embracement of the left main bronchus were caused by a ring formed by the right arcus aorta and ligament of ductus arteriosus. Although this anatomy does not comply with any of the previously described

vascular rings, clinically or by definition, we believe it may be accepted as an incomplete vascular ring. Associated anomalies are common with DOLV; however, no case of DOLV with this type of association was described.

The case of Subirana et al.⁷ was treated surgically with Mustard procedure, transaortic closure of the small outlet muscular ventricular septal defect and connection of the left-sided right ventricle to the pulmonary trunk with an extracardiac valved conduit. In our case, a palliative operation consisting of creation of a Blalock-Taussig shunt and correction of the vascular ring was done. A similar procedure consisting of an atrial switch operation with closure of the ventricular septal defect and connection of the pulmonary artery to the anatomic right ventricle via a conduit was planned as the final operation.

The two-dimensional echocardiography was an important diagnostic tool in complete and accurate diagnosis of the complex anatomy in this case. The segmental approach is of great value especially in clarifying the atrioventricular connections and spatial relations of the great arteries and outflow tracts. The existing case reports of DOLV clearly demonstrate that a wide variation in anatomy is possible. Our case is a distinctive one of the S, L, L type with atrioventricular discordance, anteriorly placed left ventricle, subpulmonic conus, and pulmonary stenosis ventricular septal defect, which also had an associated incomplete vascular ring anomaly. The presence of such a unique case may provide a new insight into this complex anatomy.

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