

Double-Outlet Left Ventricle. Echocardiographic Diagnosis

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This is a case report of a double-outlet left ventricle associated with tricuspid atresia and hypoplasia of the right ventricle, diagnosed during echocardiography with color-flow imaging, in a three-month-old child who presented with fatigue and cyanosis. The child underwent palliative pulmonary arterial banding without an invasive procedure, and showed sustained improvement during follow-up.

Double-outlet left ventricle is a rare anomaly in which the aorta and pulmonary arteries arise completely or mostly from the morphologically left ventricle. In 1819,¹ Marchal provided the first description in the literature of this anomaly. It was not until 1967 that Sakakibara et al² described it in detail however. Recent studies report that the incidence is lower than 1 per 200,000 live births³.

Case Report

A three-month-old female child, weighing 2.8kg, was admitted to the hospital with fatigue on light effort. On examination, she was found to be in regular clinical condition, cyanotic, tachypneic, had a respiratory rate of 45 ipm, and a heart rate of 130bpm. Pulmonary auscultatory findings were characterized by diffuse rales, and cardiac auscultation was characterized by a systolic murmur at the medium left sternal border. The second heart sound was split and fixed. Liver enlargement was identified at 2.5cm below the right costal margin, and the pulses were symmetric and palpable. The chest X-ray showed cardiomegaly, and a diffuse pulmonary infiltrate in the right mid-lung zone was revealed on bronchogram and was treated as bronchopneumonia. Doppler echocardiography with color flow imaging showed situs solitus, levocardia, interatrial communication of the ostium secundum type, tricuspid atresia, and a

severely hypoplastic right ventricle (fig. 1). Both arteries arose exclusively from the left ventricle. The pulmonary artery was posterior and left positioned, the aorta was anterior and right positioned, as in D-transposition of great arteries. Subaortic ventricular communication was identified together with mitral-pulmonary Fibrous continuity (fig. 2). A continuous-wave Doppler examination showed increased pulmonary flow, with a 2.12 m/s velocity. Color-flow imaging showed mild mitral and pulmonary regurgitation.

Pulmonary artery banding was indicated, and the postoperative period was uneventful. The patient was sent back to the rejer center for follow-up examination and a fontan operation was planned for the future.

Discussion

Double-outlet left ventricle is a rare anomaly in which the aorta and pulmonary arteries arise completely or mostly from the morphologically left ventricle. According to this definition, double-outlet left ventricle is a very heterogeneous entity, presenting wide variations in morphology. Thus, Van Praagh et al¹ described 14 different types, and Bharati et al⁴ report that it is not a precise entity, due to the heterogeneity of the morphological findings. Otero Coto et al⁵ stressed the need for a precise description of intracardiac morphology for treatment guidance.

Gouton et al⁶, in a review of 126 cases, reported a greater incidence of levocardia (96%), atrial situs solitus (92.8%), subaortic interventricular communication (70%), and D-transposition (65%). Atrioventricular valve anomalies were observed in 30% of cases, mostly in the tricuspid valve (atresia, 16 cases; hypoplasia, 3 cases; stenosis, 7 cases; dysplasia, 8 cases; Ebstein, 3 cases). In this eventuality, as in this study, right ventricle hypoplasia is commonly associated.

Khanolkar et al⁷ reported for the first time the association of double-outlet left ventricle, tricuspid atresia, and *cor triatriatum dexter*, which is very rare due to persistence of the embryonic right valve of sinus venosus.

Due to the great variability of associated malformations, each case has a specific surgical approach, through

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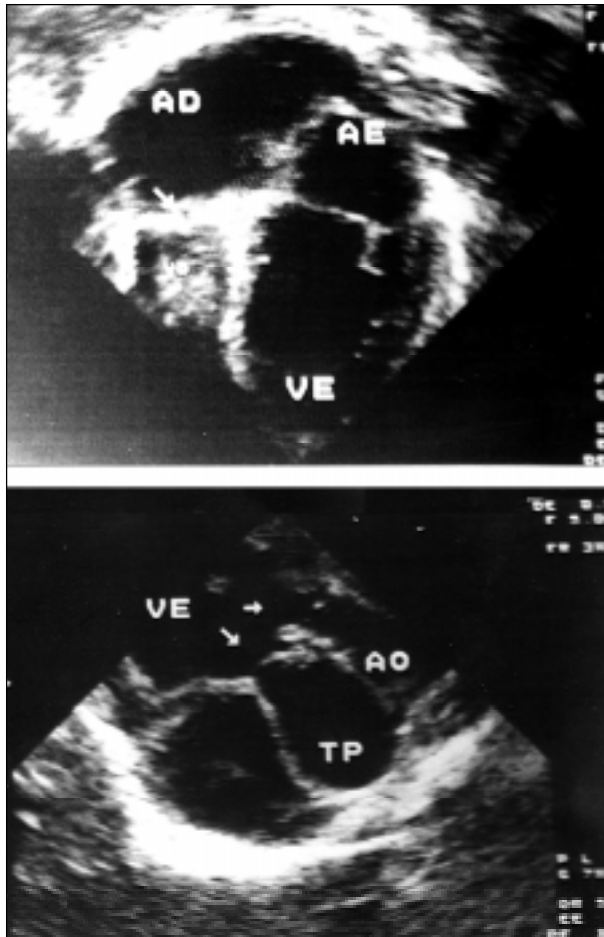


Fig. 1 - Upper panel: Four chamber view depicting. Tricuspid atresia (arrow) and the hypoplastic right ventricle (VD). Lower panel: Aorta (Ao) and pulmonary artery (TP) longitudinally related to the left ventricle. AD - Right atrium; AE - Left Atrium; VE Left ventricle.

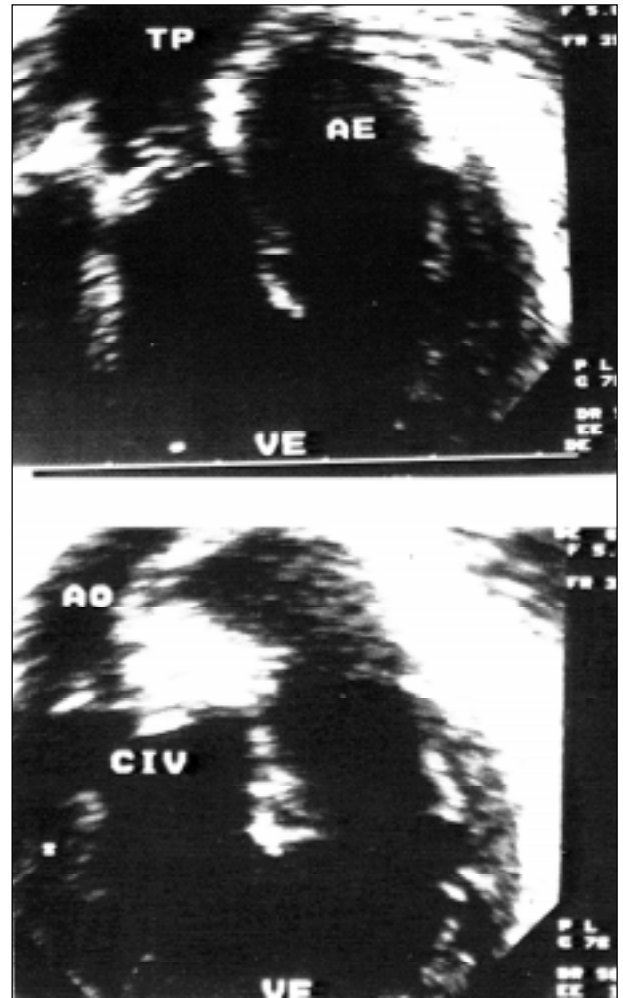


Fig. 2 - Upper panel: Pulmonary artery (TP) totally related to the left ventricle (VE) and mitral-pulmonary continuity, in anterior position. Lower panel: The aorta (Ao) is related to the left ventricle (VE) overlying a ventricular septal (CIV). AE: Left atrium.

the use of several techniques that are classified according to the presence or absence of hypoplasia of the right ventricle, pulmonary stenosis, and a relationship of interventricular communication to the great arteries^{8,9}. In the presence of a normal right ventricle, the best approach is biventricular repair, and in the presence of hypoplasia of the right ventricle, cavo pulmonary derivation of the Fontan type would be the procedure of choice. Tubes are commonly used to translocate the pulmonary artery to the right ventricle in biventricular repair, because pulmonary valve stenosis is a common association.

Before the advent of 2-D echocardiography, the diagnosis of double-outlet left ventricle was only possible through cardiac catheterization, postmortem examination, or even as surgical findings. Currently, 2-D echocardiography together with color-flow imaging enables fast diagnosis, showing both arteries arising from the morphologic left ventricle. No further difficulties occur in identifying the rela-

tionship between sigmoid valves and atrioventricular valves; presence or absence of infundibulum; common associated anomalies, such as aortic coarctation, aortic stenosis, pulmonary stenosis, tricuspid atresia, right ventricle hypoplasia, and interventricular communication.

It is important to highlight the rarity of the case described here because it did not involve pulmonary stenosis (17% of cases). Eighty-three percent of cases described by Donald and William¹⁰ of double-outlet left ventricle associated with subaortic interventricular communication and with anterior and right aorta were also associated with pulmonary stenosis. This case is in agreement with data found in the literature¹¹ where an improvement in echocardiography allowed early diagnosis of double-outlet left ventricle and treatment. It is also responsible for demonstrating aspects of a disease that was considered embryologically impossible.

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