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CONGENITAL MINI-FOCUS ISSUE

INTERMEDIATE

IMAGING VIGNETTE: CLINICAL VIGNETTE

Natural History of Double Inlet Left Ventricle and Pulmonary Hypertension in an Adult Patient

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ABSTRACT

Almost 80% of univentricular cardiac malformations with left morphology consist of a double inlet left ventricle (DILV). We report on the natural history of a 28-year-old male patient with DILV and ventriculoarterial discordance, patent ductus arteriosus, pulmonary hypertension and juxtaductal aortic coarctation. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2019;1:532–4) © 2019 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

An 18-year-old male patient was admitted to our institution due to dyspnea of moderate efforts for the last 5 years. He was known to have a heart murmur and cyanosis on lips and nails since the age of 5. On physical examination, he had acrocyanosis and acropachy (**Figure 1A**), thoracic deformity, clubbing, and hyperactive precordium, and a mesosystolic murmur was heard over the left lower sternal border II/IV. He had a single second heart sound that increased in intensity, as well as a third sound. Systemic oxygenation saturation was 80%.

Laboratory blood analysis demonstrated polycythemia (hemoglobin 24.4 mg/l, hematocrit 75.9%), hypochromia, anisocytosis, and hyperuricemia. An electrocardiogram demonstrated sinus rhythm with biventricular enlargement (**Figure 1B**). A transthoracic echocardiogram showed situs solitus, double inlet left ventricle (DILV), anatomically left-sided morphologically rudimentary right ventricle (Video 1), perimembranous ventricular septal defect (9 mm), patent ductus arteriosus (7 mm), juxtaductal aortic coarctation, hypoplasia of aortic transverse portion, levo-transposed great arteries (**Figures 1C to 1E**), and severe pulmonary arterial hypertension with systolic pulmonary artery pressure of 98 mm Hg. These findings were corroborated by computed tomography (**Figure 1F**).

DILV is a form of functionally univentricular heart defined by atrioventricular connection, in which the morphologically left ventricle receives more than 50% of the atrioventricular valves when they are separated, or more than 75% of the common atrioventricular valve (1). The anatomic heterogeneity in the group of DILV hearts is characterized by the position of the ventricles to each other and the relationship of the great arteries to each other as well as to the ventricles (2).

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Informed consent was obtained for this case.

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The survival of our patient until the third decade of life without surgical treatment is extraordinarily rare and exceeds the expectations of overall survival of DILV. The noninvasive imaging studies play an important role in its diagnosis and follow-up.

ABBREVIATIONS AND ACRONYMS

DILV = double Inlet left ventricle

In the largest series of patients with only 1 ventricle without surgical treatment, it was found that in the spectrum of DILV, most patients die in their late teens to early 20s, with 50% mortality 14 years after diagnosis, and 70% of patients with DILV died at 16 years of age. The most common causes of death for patients with this type of congenital heart disease without surgery include congestive heart failure, arrhythmias, or sudden death (3).

Currently, our patient is 28 years of age and has survived due to duct-dependent congenital heart disease and high pulmonary resistance from birth. In the absence of elevated pulmonary vascular resistance, the patient would probably have died in the first year after birth with congestive heart failure.

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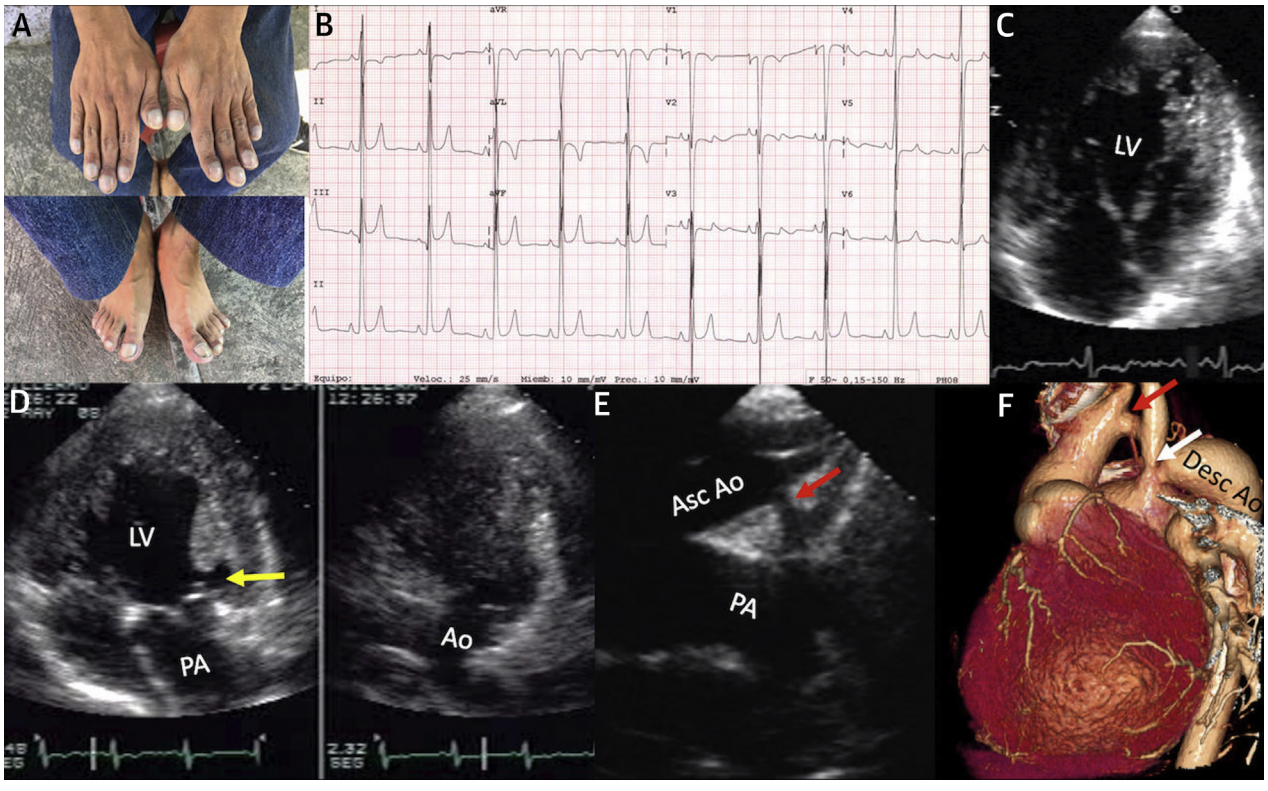
FIGURE 1 Clinical and Imaging Findings



(A) Acrocyanosis and acropachy. **(B)** Electrocardiogram. **(C)** Transthoracic echocardiogram: 4-chamber view demonstrating 2 atrioventricular valves entering the left ventricle (LV). **(D)** Five-chamber view with discordant atrioventricular connection; the **yellow arrow** points the rudimentary ventricle morphologically right. **(E)** Suprasternal view showing the right aortic arch with hypoplasia in the transversal portion **(red arrow)**. **(F)** Three-dimensional reconstruction of the heart showing ventriculoarterial discordance, right aortic arch with hypoplasia of its transverse portion **(red arrow)**, juxtaductal aortic coarctation, and persistence of ductus arteriosus **(white arrow)**. See Video 1. Ao = aorta; Asc = ascending; Desc = descending; PA = pulmonary artery.

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KEY WORDS double inlet left ventricle, computed tomography, echocardiography, natural history, pulmonary artery hypertension

APPENDIX For a supplemental video, please see the online version of this paper.