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MINI-FOCUS ISSUE: CONGENITAL HEART DISEASE

ADVANCED

CASE REPORT: DA VINCI CORNER

Complete Transposition of Great Arteries With Dominant Left Ventricle

Long-Term Survival in Natural History

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ABSTRACT

We report the case of an adult patient, affected by complete transposition of great arteries with ventricular septal defect, who survived until 68 years of age without surgery, thanks to the presence of a common atrium and pulmonary stenosis. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2020;2:2107–10) © 2020 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/).

HISTORY OF PRESENTATION

In 1989, at the age of 40 years, the patient had been diagnosed with complex cyanotic congenital heart disease (CHD). The echocardiogram showed a situs solitus with juxtaposition of atrial appendages, hypertrophic left and hypoplastic right ventricles, transposition of great arteries (TGA), and pulmonary stenosis with 50 mm Hg gradient along the pulmonary outflow tract. The case was considered as univentricular heart with TGA and pulmonary stenosis. Fontan intervention was excluded for high pulmonary pressure at catheterization.

LEARNING OBJECTIVE

- This case represents a beautiful example of how nature resolves hemodynamic problems otherwise resolved with multiple surgical interventions.

PAST MEDICAL HISTORY AND MANAGEMENT

The patient presented episodes of atrial fibrillation and flutter that required electric cardioversion and antiarrhythmic and anticoagulant therapies. Secondary erythrocytosis occurred that required periodic therapeutic bloodletting.

The last echocardiogram showed hypertrophic left ventricle (LV) and hypoplastic right ventricle (RV); the peak systolic gradient of pulmonary valve was 50 mm Hg, telediastolic volume of LV was 119 ml/m^2, and ejection fraction was 49% of left ventricle. Arterial pressure was 118/65 mm Hg, heart rate was 60 beats/min, and saturated O_2 was 73% (ambient air). The most clinically significant blood biomarkers were as follows: troponin I as 0.058 μg/l; microcytic hypocromic erythrocytes; secondary erythrocytosis red blood cells: 7.22 × 10^{12}/l, white blood cells: 6.45 × 10^9/l; hemoglobin: 14.4 g/dl; hematocrit: 50.6%; and mean corpuscular volume 70.1 fl. Kidney

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ABBREVIATIONS AND ACRONYMS

Ao = aorta
CHD = congenital heart disease
CT = cresta terminalis
LAA = left atrial appendage
LV = left ventricle
M = mitral valve
PH = pulmonary hypertension
PV = pulmonary valve
RAA = right atrial appendage
RV = right ventricle
T = tricuspid valve
TGA = transposition of great arteries
VSD = ventricular septal defect

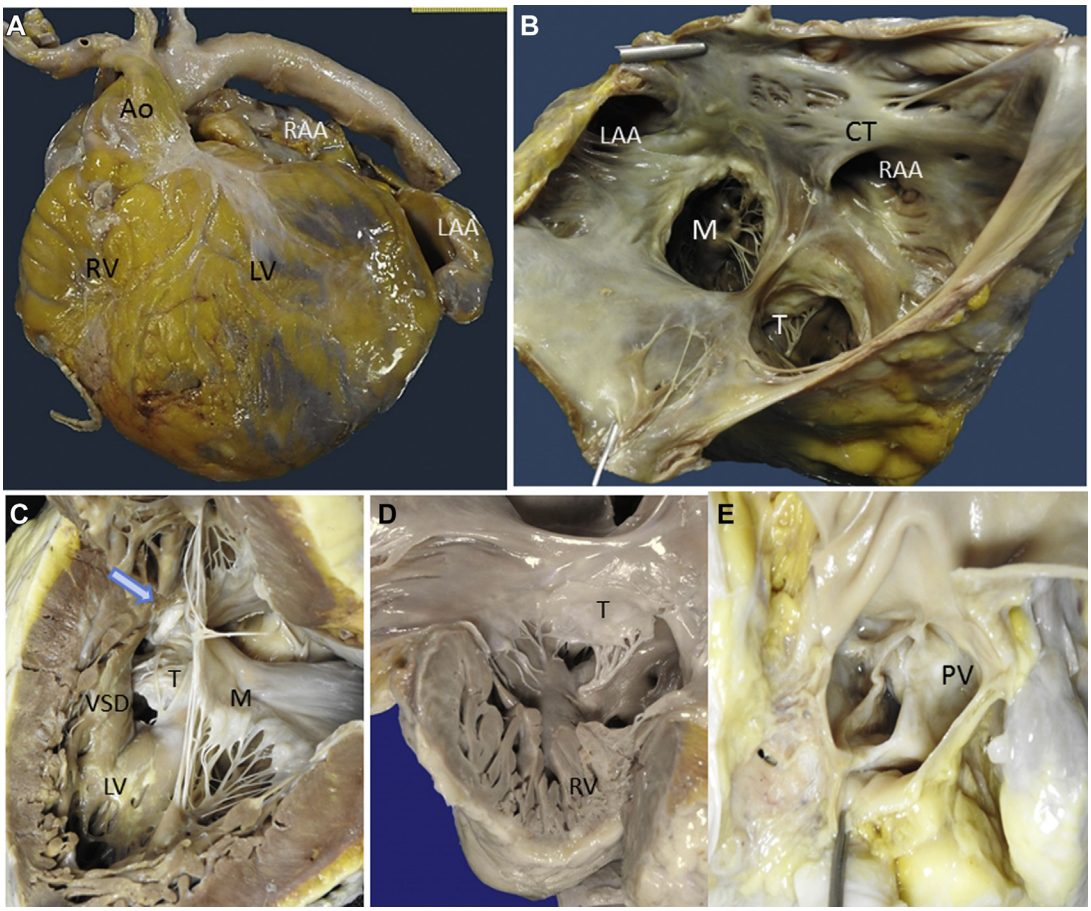
and liver function was normal and creatinine was 81 μmol/l.

The patient died at home due to sudden cardiac arrest.

AUTOPSY INVESTIGATION

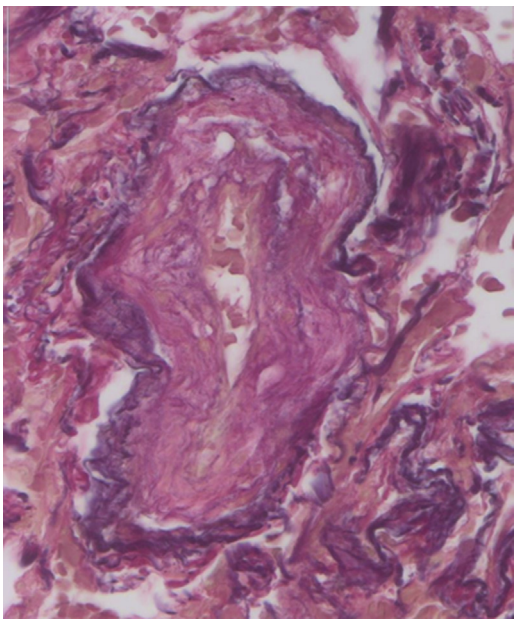
Autopsy showed atrial situs solitus, anterior and right-sided aorta, and left juxtaposition of the atrial appendages (1) (Figure 1A). Concordant atrioventricular connection was present (Figure 1B) and the aorta originated from a hypoplastic RV and the pulmonary artery from the left ventricle (LV) (discordant ventriculo-arterial connection) in keeping with complete TGA. A common atrium was noted, due to total absence of the interatrial septum (Figure 1B). A large perimembranous ventricular septal defect (VSD) was found as well as a subpulmonary stenosis due to a big anterolateral muscle band associated with a fibrous ring (Figure 1C). Moreover, the RV was hypoplastic compared with the LV (Figure 1D). The pulmonary valve was bicuspid, stenotic and dysplastic (Figure 1E). At histological examination of the lungs, the pulmonary small arterial vessels and arterioles presented hypertensive vascular lesions grade III according to Heath-Edwards classification (2) (Figure 2). The final diagnosis was complete TGA, common atrium, left juxtaposition of the atrial appendages, and VSD with pulmonary stenosis and hypoplasia of the RV.

FIGURE 1 Results From the Autopsy



(A) Anterior view of the heart with anterior and right-sided aorta (Ao). Note the presence of left juxtaposition of atrial appendages (right atrial appendage [RAA] and left atrial appendage [LAA]). **(B)** At the opening of the atria, a common atrium is present with the total absence of the atrial septum. Note the mitral (M) and tricuspid valves (T) in the setting of atrioventricular concordance and the entrance of RAA and LAA. **(C)** View of the left ventricle (LV) with a wide ventricular septal defect (VSD) and a subpulmonary fibrous ring **(arrow)**. **(D)** View of right ventricle (RV) with hypoplasia and hypertrophy of the trabeculae. **(E)** A bicuspid, stenotic, and dysplastic pulmonary valve (PV) is seen. CT = cresta terminalis.

FIGURE 2 Histological Examination of the Lungs



Pulmonary small artery with concentric, obstructive fibrous thickening (grade III of Heath-Edwards classification). Weigert van Gieson stain; 320x magnification.

DISCUSSION

In patients with complete TGA (concordant atrioventricular and discordant ventriculo-arterial connections) the systemic and pulmonary circulations are in parallel. The survival after birth is accomplished by prostaglandin to maintain patency at the ductus arteriosus or by the creation of an adequate mixing between the two circulations (Raskind or Blalock-Hanlon procedures), followed by atrial or arterial switch total repair.

TGA is a severe congenital heart disease, incompatible with extrauterine life because pulmonary and systemic circulations are in parallel after birth (3,4).

Before planning total repair, either atrial or arterial switch, palliative emergency procedures are mandatory to guarantee blood mixing for oxygenation.

In 1950, Alfred Blalock and Rollins Hanlon invented the surgical removal of the atrial septum (atrial septectomy known as Blalock-Hanlon technique) (5) and in 1966 William Rashkind and William Miller created an atrial septal defect with balloon catheter through foramen ovale without thoracotomy (atrioseptostomy known as Rashkind palliation) (6).

The peculiarity of this report is the long-term survival (68 years of age) of this patient affected by complete TGA without any previous percutaneous or surgical interventions. A common atrium, due to absence of the interatrial septum, allowed a large blood mixing at atrial level and permitted a long natural survival. The anatomy of the heart with two ventricles and a single atrium once was called "cor triloculare biventriculare" (7). Oddly enough, despite the presence of pulmonary stenosis, obstructive pulmonary vascular disease occurred.

Pulmonary vascular disease is a multifactorial disease; it develops in congenital heart defects with pulmonary hypertension (PH) for hemodynamic reasons. In our case, there was a pulmonary stenosis characterized by both hypertrophic muscular band and fibrous ring that should have protected the patient from the development of severe pulmonary vasculopathy. In adult patients we classified 5 phenotypes of PH (8). Our case could be categorized as hypertension with CHD at old age. As suggested by some authors, PH could also be considered to be coincidental and up to 10% of patients who undergo the Mustard or Senning procedure in the first few weeks of life still can develop advanced PH (9). Our patient developed grade III pulmonary vasculopathy, according to Heath-Edwards classification. In this very intriguing case, some associated anomalies, in particular the pulmonary stenosis, should have protected the lung from hypertension. Of great value would be the assessment of adequacy of ventricle chamber size for the repairing of malformation. In this case, the presence of hypoplastic RV and a common atrium, a large VSD with shunt left to right into the aorta, can account for the long survival with cyanosis.

CONCLUSIONS

In our case, the common atrium resolved "naturally" the blood missing and the huge interventricular septal defect favored the blood flow into the aorta. If the pulmonary hypertension increased in old age, the serial defects could have guaranteed the oxygenated blood flow into the aorta. A wide communication between the two atria may be congenitally present, if the atrial septum does not develop, not requiring palliative emergency procedures.

AUTHOR DISCLOSURES

This study was supported by the Veneto Region Registry. All authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS **complete transposition of the great arteries, congenital heart disease**