

IMAGE IN CARDIOLOGY

Long-term survival in a patient with single ventricle

Sobrevivência a longo prazo num paciente com um único ventrículo

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We report the case of a 58-year-old man with complex congenital heart disease who had undergone a classic Blalock-Taussig (left subclavian artery [SA]-left pulmonary artery [PA]) shunt operation at the age of five. Because of

limiting dyspnea he underwent reoperation at the age of 29. A double-inlet ventricle, transposition of the great arteries, severe pulmonary valve stenosis and a classic Blalock-Taussig shunt with suture stenosis were described in the report of this operation. At that time a modified Blalock-Taussig shunt (Gore-Tex graft between right SA and right PA) was performed. During follow-up a cardiac pacemaker (VVI-R) was implanted. He now presented with exercise intolerance.

The transthoracic echocardiogram (Figure 1) revealed a double-inlet dominant left ventricle, separated from the accessory right ventricle by a hypoplastic intraventricular septum; the atrioventricular connection was discordant. Color Doppler echocardiography showed a small right pulmonary fistula with slow bidirectional flow (Figure 2). Computed tomography angiography revealed pulmonary

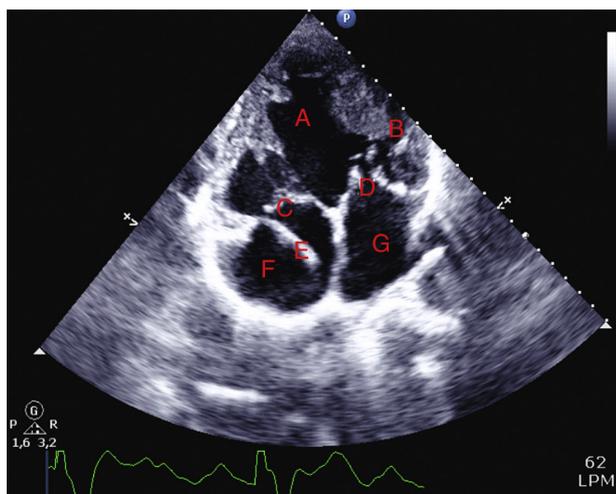


Figure 1 Echocardiogram in apical 4-chamber view showing double-inlet dominant left ventricle (A), separated from the accessory right ventricle (B) by a hypoplastic intraventricular septum. The atrioventricular connection is discordant: mitral (C) and tricuspid (D) valves. Pacemaker electrode (E), right atrium (F) and left atrium (G).

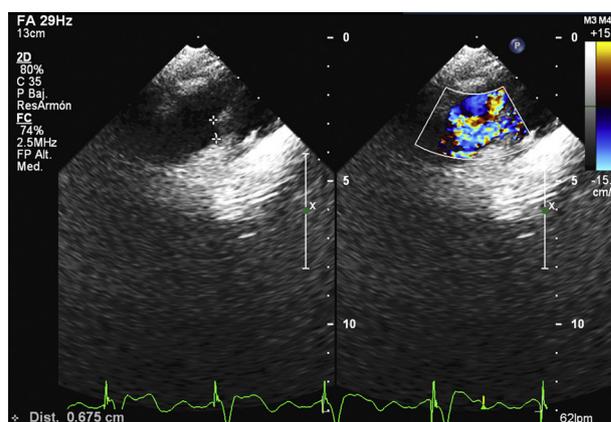


Figure 2 Echocardiogram in color Doppler view showing right pulmonary fistula with slow bidirectional flow.

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Figure 3 Computed tomography angiography showing pulmonary valve stenosis and hypoplastic right ventricular outflow tract.

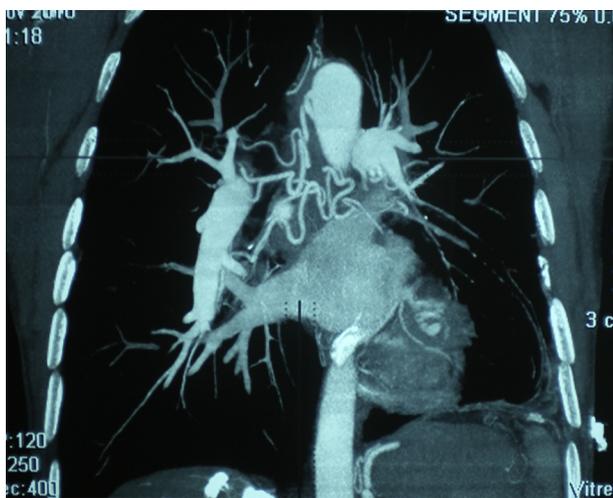


Figure 4 Computed tomography angiography showing systemic-pulmonary collateral circulation via bronchial artery branches.

valve stenosis and a hypoplastic right ventricular outflow tract (Figure 3), as well as systemic-pulmonary collateral circulation via bronchial artery branches (Figure 4).

With these palliative operations, the patient survived with an acceptable quality of life, probably due to the suture stenosis of the first graft and the small caliber second graft. These and the systemic-pulmonary collateral circulation had delayed the appearance of pulmonary hypertension by hyperflow, which is now suspected because of the bidirectional flow in the graft.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that no patient data appear in this article.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

Conflicts of interest

The authors have no conflicts of interest to declare.