SCIENTIFIC LETTER

ABSENT PULMONARY VALVE IN CORRECTED TETRALOGY OF FALLOT

ABSTRACT

We present the case of a 24-year-old male patient with a history of correction of tetralogy of Fallot when he was 2. He reports presenting 6 syncopal episodes during the last 6 months. Therefore, a transthoracic color Doppler echocardiogram was performed, reporting right ventricular dilatation, mild global deterioration of the right ventricular systolic function, absent pulmonary valve with severe insufficiency, and free passage. A diagnosis of absent pulmonary valve with severe valvular insufficiency was made. Surgical treatment was performed by placing an aortic bioprosthesis (stentless) No. 25, Medtronic Freestyle™, in the pulmonary position. The patient evolved favorably and was discharged without complications on the fourth postoperative day. In addition, a brief bibliographic review of the disease is made.

Keywords: *absent pulmonary valve, tetralogy of Fallot, adult congenital heart disease*

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INTRODUCCIÓN

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease in children and adults. Absent pulmonary valve is a rare variant. In 90% of cases, it is associated with other malformations[1]. Tetralogy of Fallot with an absent pulmonary valve represents 3% to 6% of patients with a rare congenital anomaly characterized by rudimentary ridges or total absence of pulmonary valve tissue and usually with a hypoplastic pulmonary valve annulus[2].

We present the clinical case of a 24-year-old male patient with a pathological history of tetralogy of Fallot that was corrected at two years of age. He evolved well and remained asymptomatic for a long time until 6 months before the consultation, when he began to present episodes of syncope and palpitations. Physical examination showed no particular signs or characteristic facies. During the physical examination of the cardiovascular system, a holodiastolic pulmonary murmur was auscultated. Spirometry was requested, within normal limits, and a 24-hour Holter reported sinus rhythm and frequent ventricular extrasystoles grouped up to 3 beats. A transthoracic color Doppler echocardiogram was performed (*Figure 1*), with right ventricular dilatation, preserved size of the remaining cavities, preserved left ventricular systolic function (Fey 59%), slight global deterioration of right ventricular systolic function, TAPSE 18 mm, and absent pulmonary valve with severe insufficiency with free passage. No significant valvulopathies, no masses, or intracavitary shunt were observed. As a complementary study, coronary angiography was requested, which showed coronary arteries without significant angiographic lesions.

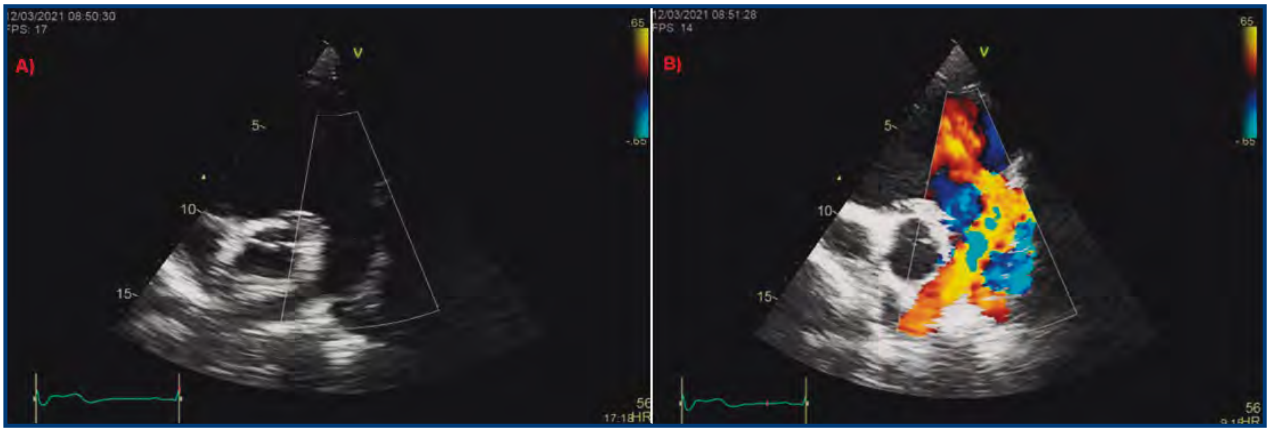

FIGURE 1. Transthoracic color Doppler echocardiogram. Short axis at the level of great vessels. A. Pulmonary valve is absent. B. Color Doppler shows severe pulmonary regurgitation.

With these findings, a diagnosis of absence of pulmonary valve with severe valvular insufficiency was made, and surgical treatment was decided.

Several types of prostheses can be implanted in the pulmonary valve position or surgical correction of the absent pulmonary valve. The types of conduits used are mechanical prosthesis, biological prosthesis, and homografts[3]. Patients with surgically corrected tetralogy of Fallot in pediatric age 5-10% are reoperated over 20-30 years of follow-up, most commonly undergoing surgery for pulmonary valve replacement[4].

A pericardial patch was visualized through a median sternotomy, with extracorporeal circulation, double cava and ascending aorta cannulation, crystalloid cardioplegia, and controlled hypothermia at 28 degrees centigrade. A longitudinal incision was made at the level of the pulmonary artery trunk and infundibulectomy with right ventricular outflow tract enlargement, visualizing the absence of a native pulmonary valve. Aortic bioprosthesis No. 25 biological Medtronic Freestyle (*Figure 2*) was placed in a pulmonary position with a continuous suture of Prolene 4/0 SH, extracorporeal circulation (73 minutes) and aortic clamping (49 minutes).

The patient evolved favorably with extubation at 8 hours postoperatively, hemodynamically stable, without vasopressor or inotropic support, and sinus rhythm; chest drains were removed at 24 hours, leaving the cardiovascular recovery unit on the second postoperative day. A control transthoracic color Doppler echocardiogram was requested, where the prosthesis was observed in pulmonary position, without perivalvular leaks, with maximum velocity 2.7 m/s and mean gradient 25 mmHg of the pulmonary valve, mild tricuspid insufficiency, estimated pulmonary systolic pressure 27 mmHg. On the fourth postoperative day, he was discharged without complications.

Declarations

The authors declare no conflict of interest.

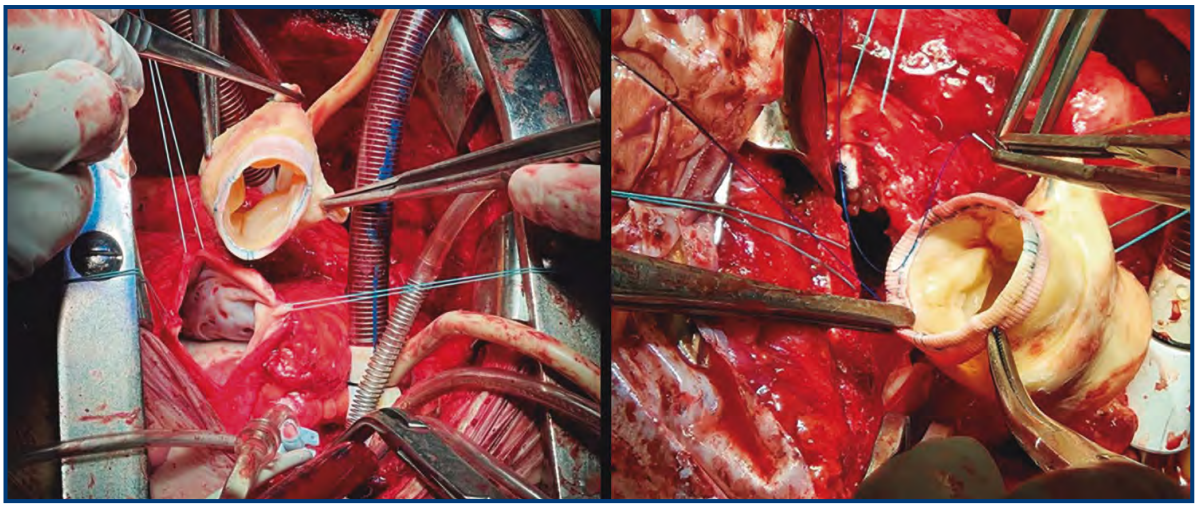

FIGURA 2. Surgical treatment: placement in a pulmonary position of aortic bioprosthesis No. 25 biological Medtronic Freestyle™.

REFERENCES

1. Fallot A. Contribution a l'anatomie pathologique de la maladie bleue (cyanose cardiaque). Marseille Medicale. 1888: 71-
93, 138-158, 207-223, 270-286 y 341-354.

2. Apitz C, Weeb GD, Redington AN. Tetralogía de Fallot. Lancet. 2009; 374: 1462-1471

3. Karl TR, Musumeci F, de Leval M, et al. Surgical treatment of absent pulmonary valve syndrome. J Thorac Cardiovasc Surg. 1986 Apr. 91(4):590-7

4. Snir E, de Leval MR, Elliott MJ, Stark J. Current surgical technique to repair Fallot's tetralogy with absent pulmonary valve syndrome. Ann Thorac Surg. 1991 Jun. 51(6):979-82.