

Adult patient with truncus arteriosus type I. Management and follow-up (RCD code: IV-1C.3b)

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Abstract

Truncus arteriosus is a congenital heart malformation where systemic, pulmonary and coronary circulation is supplied by a single artery arising from the heart. Surgical correction of this disorder remains the treatment of choice. Presenting the case of a 21-year-old male, who underwent surgical correction of the anomaly in youth, we review management and follow-up strategy for adult patients after correction of truncus arteriosus. Conduit failure is the most commonly observed long-term consequence of the surgical correction. We evaluate diagnostic algorithm and indications for conduit replacement. JRC D 2014; 1 (8): 15–18

Key words: grown-up congenital heart disease, single arterial trunk, ventricular septal defect, conduit

Background

Truncus arteriosus (TA) is a rare congenital heart disease occurring in 0.034 to 0.56 per 1000 newborns. It affects 1.4% to 2.8% of all congenital heart disease patients [1]. In this anomaly, a single arterial trunk arises from the heart, overrides the interventricular septum, and supplies systemic, pulmonary, and coronary circulations [2,3]. Without surgical treatment, 80% of the patients die within the first year of life, usually in early infancy [4]. Repair of TA during the neonatal and early infant period has become a standard practice in many centers, with good outcomes [5]. We present a case of a patient who underwent a surgery of TA type I.

Case presentation

A 21-year-old Caucasian man with TA type I was referred to the Centre for Rare Cardiovascular Diseases (CRCD) at the John Paul II Hospital in Krakow, Poland, for cardiac evaluation due to gradual loss of exercise capacity and exertional dyspnea. TA was diagnosed in the second day of life by cardiac echocardiography. The patient underwent a surgical repair in August 1990 with implantation of the pulmonary homograft no. 9 and Dacron conduit no. 12. In 1993, reoperation was performed due to stricture of

pulmonary homograft. Homograft no. 19 pulmonary and Dacron conduit no. 22 were implanted. No complications were observed during either of the procedures.

The patient was referred to our center by his general practitioner in May 2011 with suspicion of pulmonary homograft stricture based on echocardiographic examination. On admission, he was hemodynamically stable with no signs of peripheral or pulmonary edema. His heart rate was 75 beats/min and blood pressure was 135/80 mm Hg. He was considered to be in class I according to the New York Heart Association (NYHA) classification. He complained of minor reduction in exercise capacity. Comorbidities included mild bronchial asthma and recurrent migraine headaches. He had no family history of congenital heart defects. No drug, alcohol, or cigarettes use was reported. In 1990, he underwent pyloroplasty due to inborn pylorus stenosis. A physical examination revealed no significant abnormalities. A biochemical blood analysis showed normal values of complete blood count and no signs of kidney or liver dysfunction.

Transthoracic echocardiography revealed calcifications of the homograft and stricture of the pulmonary conduit. The pressure gradient in the right ventricular (RV) outflow track was 99.9/66 mm Hg (Figure 1). During 24-hours Holter ECG monitoring, no abnormalities were detected. A cardiopulmonary exercise test was completed in 15 min and 2 sec and the result was 11.7 METs, which confirmed

Conflict of interest: none declared.

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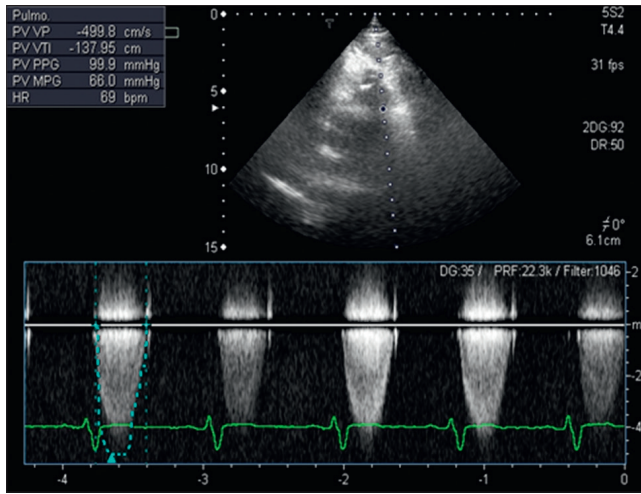


Figure 1. Transthoracic echocardiography. Parasternal short-axis view. The pressure gradient in the right ventricular flow tract was 99.9/66 mm Hg

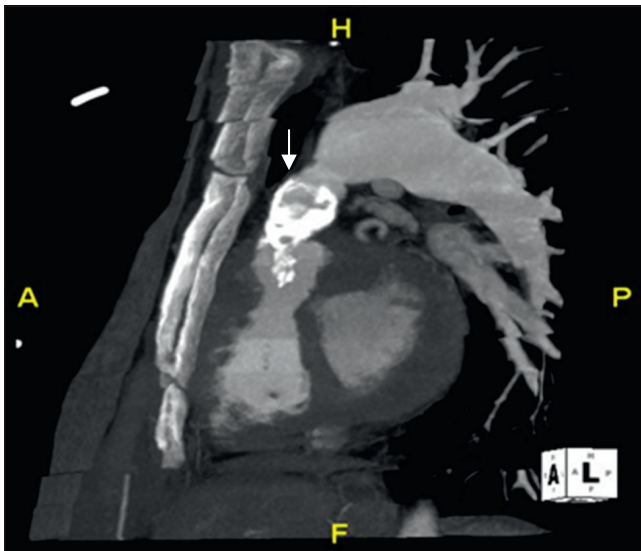


Figure 2. Cardiovascular computed tomography. Two-dimensional reconstruction. Calcification of the homograft (arrow)

that exercise capacity was within the reference range. No chest pain, arrhythmia, or ST-segment deviation was observed during the test. Maximal oxygen consumption reached 23.4 mL/kg/min.

Cardiac computed tomography showed calcified homograft and stenotic Dacron pulmonary conduit (Figure 2, 3). Right heart catheterization revealed high systolic pressure in the RV (100 mm Hg) and severely elevated systolic pressure in the pulmonary graft (up to 100 mm Hg) (Figure 4).

Management strategy

Because the patient is currently asymptomatic and reports only a minor reduction in exercise capacity (NYHA class I), he should

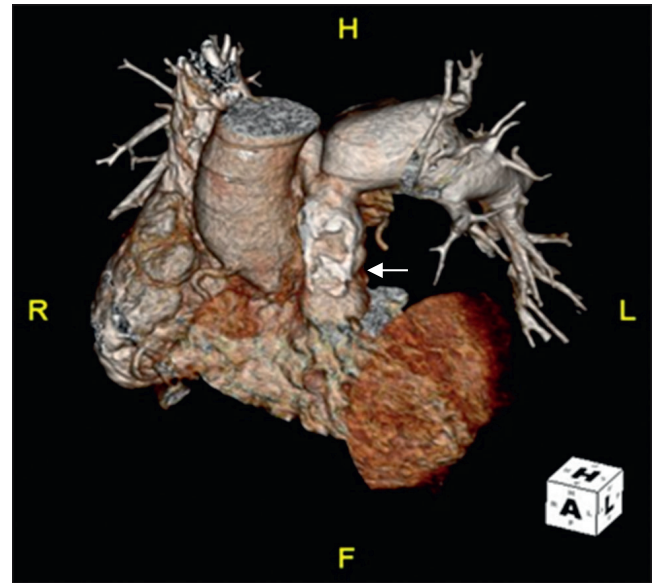


Figure 3. Cardiovascular computed tomography. Three-dimensional reconstruction. Calcification of the homograft and stenosis of Dacron conduit (arrow)

be considered for optimal medical therapy. No surgery is indicated at this point.

Discussion

TA is an uncommon congenital cardiac malformation constituting less than 3% of all congenital heart malformations [1]. TA is characterized by a single great artery arising from the base of the heart, which supplies systemic, coronary, and pulmonary blood flow, together with a ventricular septal defect [2,3]. The two main classification systems used to describe the anatomy of TA are those of Collett and Edwards (1949) and Van Praagh (1965) [6,7]. Without surgical treatment, 80% of the patients die within the first year of life, usually during early infancy [8,9]. The results of physiological repair have improved over the years, but pulmonary hypertensive episodes in the immediate postoperative course are major risk factors [10].

Conduits establish the continuity between the RV and the pulmonary artery in complex defects when the native outflow tract is not amenable to reconstruction.

The types of conduits include valved (pulmonary or aortic homograft, bioprosthetic valves, bovine jugular vein conduits [Contegra]) and nonvalved conduits [11].

There is no ideal conduit. Limited durability implicates early reoperation. Predictors for conduit failure are sterilization/preservation process, smaller conduit, conduit type, younger age at implantation, pulmonary artery stenosis, and diagnosis of transposition.

A 20-year period free from reoperation for conduit failure was reported at the level of 32% to 40% [12]. Complications include outgrowth, progressive obstruction with and without regurgitation, endocarditis, and aneurysms or pseudoaneurysms [13].

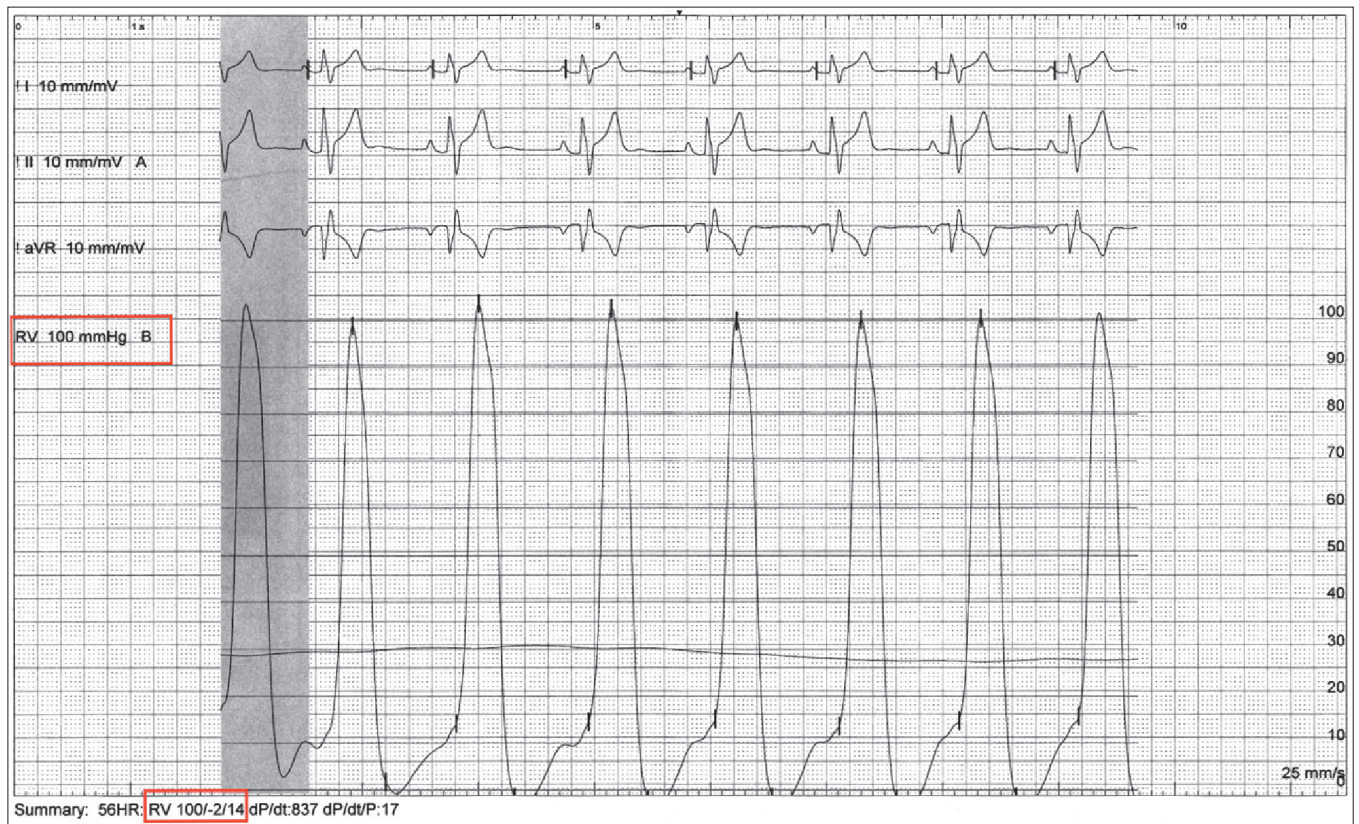


Figure 4. Right heart catheterization. The right ventricular systolic pressure was 100 mm Hg

Clinical presentation may include exertional dyspnea, palpitations, syncope, and sudden cardiac death [14].

In our patient, no clinical symptoms were observed, and the diagnosis of conduit obstruction was made accidentally.

Echocardiography is the first-line diagnostic tool providing providing the measurement of the size and function of both ventricles, pulmonary and tricuspid regurgitation, and associated lesions. Gradients across the conduit may be difficult to measure and not reliable. The RV pressure derived from tricuspid regurgitation velocity should be used to assess conduit stenosis. Cardiac magnetic resonance imaging and computed tomography may be required to image the conduit (level of stenosis), pulmonary artery, and coronary artery for the assessment of the RV and severity of pulmonary regurgitation.

Catheterization with hemodynamic assessment is always required if intervention is considered. Angiography provides information on the level of stenosis, peripheral pulmonary artery stenosis, and coronary anatomy (anomalies/abnormal course).

In our case, we performed the whole diagnostic algorithm including echocardiography, computed tomography, and cardiac catheterization, which allowed us to confirm the diagnosis of conduit stenosis. Owing to the lack of clinical symptoms, we decided against surgery and recommended regular follow-up.

Longitudinal monitoring of the homograft and conduit morphology are more important for timing of reintervention than single measurements. Regular follow-up in a center specializing in grown-up congenital heart diseases is recommended at least every 12 months.

In our case, we decided to perform regular follow-up at least every 3 months. Special attention should be given to exercise capacity (cardiopulmonary exercise testing), RV systolic pressure (conduit gradient), RV function, tricuspid regurgitation, and arrhythmias.

Conclusions

The patient has been scheduled for optimal medical treatment and regular follow-up. Transthoracic echocardiography with careful assessment of the RV function, RV systolic pressure (conduit gradient), and the severity of tricuspid regurgitation is required every 3 months. Periodical cardiopulmonary exercise testing and Holter examination are required. If new symptoms develop and the patient's condition worsens, indications for surgery should be evaluated again.

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