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Congenitally corrected transposition of the great arteries and participation in competitive sport

Wrodzone skorygowane przełożenie wielkich pni tętniczych a możliwość uprawiania sportu wyczynowego

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Abstract

Congenitally corrected transposition of the great arteries is a rare anomaly, where the systemic circulation is supported by the morphological right ventricle. We present a 43 year-old asymptomatic male, a former competitive short-distance runner, with recently diagnosed congenitally corrected transposition of the great arteries. To our knowledge this is the first report of such a case.

Key words: congenitally corrected transposition of the great arteries, transesophageal echocardiography, athlete's heart

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INTRODUCTION

Congenitally corrected transposition of the great arteries (ccTGA), first described by Von Rokitansky in 1875 [1], is characterised by atrioventricular and ventriculoarterial discordance. It is a rare anomaly, accounting for less then 1% of congenital heart disease, and is usually accompanied by other cardiac defects including ventricular septal defect, pulmonary or subpulmonary stenosis, and left atrioventricular (morphological tricuspid valve) anomalies [2]. Congenital complete heart block occurs in 5% of cases. Patients with ccTGA as a lone anomaly do not require surgical repair and the diagnosis is often discovered accidentally during adulthood. We present a case of an asymptomatic adult, a former competitive short-distance runner, with newly diagnosed ccTGA without any other cardiac abnormalities.

CASE REPORT

A 43 year-old male was referred to the Congenital Heart Disease Department due to abnormal chest X-ray found during a standard check-up at work. The patient was completely asymptomatic and reported excellent exercise tolerance, allowing him to regularly run, play football and do other physical activities with no restrictions. He was formerly an athlete and competed at local, regional and national levels as a short-distance runner (with some significant achievements at national level as a juvenile competitor).

On physical examination, no signs of heart failure were found. Only soft systolic murmur (grade 1/6) was audible at the left sternal border. A 12-lead ECG showed normal sinus rhythm (QRS duration 106 ms) with non-specific ST-T wave abnormalities.

Because the transthoracic echocardiographic study was insufficiently clear, due to mesocardia, transesophageal echocardiography was performed. In two-dimensional study, the right atrium entered the morphological left ventricle (LV) and the left atrium communicated with the morphological right ventricle (RV) (Fig. 1). The echocardiogram also revealed aorta rising from the left-sided morphological RV with tricuspid valve (Fig. 2). The morphological LV with mitral valve laid on the right side and was connected to the pulmonary artery. The RV wall was thickened, but the ventricular function was well preserved. Colour Doppler imaging showed mild systemic atrioventricular (tricuspid) and pulmonary valve regurgitation (Fig. 3). No additional cardiac defects were found.

The patient was allowed to continue his sporting activities and was submitted to routine clinical follow-up evaluation with no pharmacological therapy.

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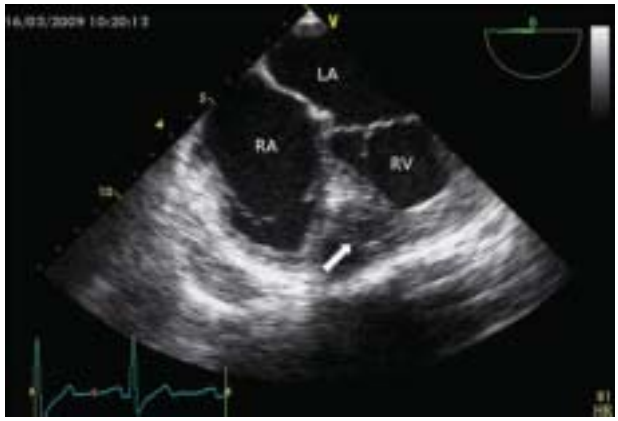

Figure 1. Transesophageal echocardiogram (four-chamber view, transverse plane). The left-sided morphological right ventricle (RV) with tricuspid valve communicates with the left atrium (LA) and the morphological left ventricle with mitral valve (laid on the right side) — with the right atrium (RA). Hypertrophy of the RV wall and thickened moderator band can be noticed

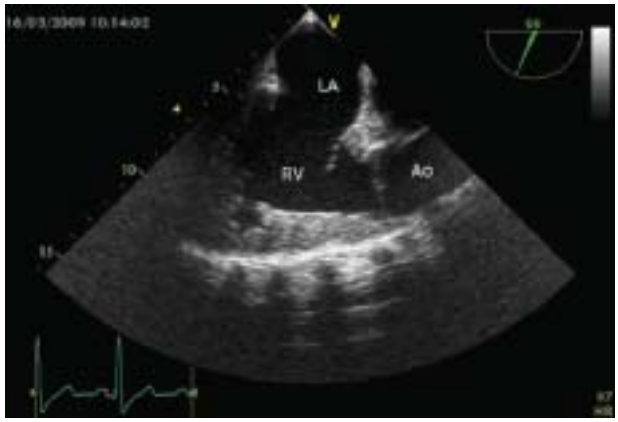

Figure 2. Transesophageal echocardiogram (long axis ascending aorta, 66° rotation). Aorta (Ao) rises from the morphological right ventricle (RV). Discontinuity between systemic atrioventricular (tricuspid) valve and aortic valve can be observed; LA — left atrium

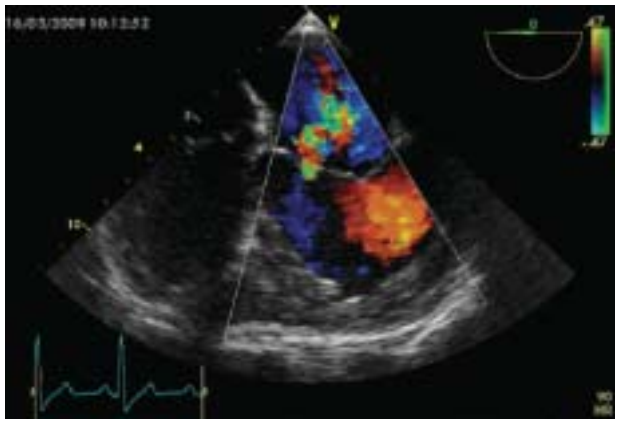

Figure 3. Transesophageal echocardiogram (four-chamber view, 0° rotation). Colour Doppler imaging. Mild systemic atrioventricular (tricuspid) valve regurgitation

DISCUSSION

Patients with ccTGA may reach adulthood being considered as entirely healthy. Echocardiography remains the crucial diagnostic tool in the proper diagnosis and follow-up of patients with ccTGA. Patients with this condition often require transesophageal study, because of poor echo window (particularly when dextrocardia or mesocardia are present), to assess ventricular function, atrio-ventricular valve regurgitation and concomitant abnormalities.

The absence of other cardiovascular disorders associated with this anomaly characterises numerous survivals until the sixth or seventh decade of life. However, the lifespan of individuals with ccTGA is limited by the onset of the failure of the RV, that supports systemic circulation over decades.

Fredriksen et al. [3] demonstrated that aerobic capacity in patients with ccTGA is severely diminished, achieving only 30–50% of the results observed in healthy subjects. Although normal values of right ventricular and left ventricular ejection fraction were found, the systemic right ventricular ejection fraction did not increase > 5% from rest to exercise, as was observed in healthy subjects. Bos et al. [4] showed that asymptomatic middle-aged patients with ccTGA have RV dysfunction quantified by Doppler tissue/strain echocardiography. A retrospective multi-institutional study revealed that, even without associated malformations, more than one in three patients with ccTGA develop heart failure by the age of 40 [5].

However, our case shows that a patient with isolated ccTGA may present an excellent clinical condition. According to the Bethesda guidelines, asymptomatic patients with ccTGA and no other cardiac abnormalities may participate in class IA and IIA sports (e.g. golf, diving) if no systemic ventricle enlargements or arrhythmias are found on examination [6]. They should however be periodically evaluated to detect arrhythmias, systemic ventricular dysfunction or systemic atrioventricular valve regurgitation. Sports with a large static component (classes IIIA, IIIB, IIIC — e.g. weightlifting, skiing) are not recommended. Our described athlete had significantly exceeded the permitted level of activity for individuals with ccTGA practising sports with moderate static and moderate/large dynamic load.

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