Two decades of follow-up of a 60-year-old cyanotic patient with an unoperated univentricular heart

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Univentricular heart is a rare congenital heart malformation, and a life span of over 60 years is very unusual in this population [1]. A 60-year-old woman with this congenital heart defect due to non-restrictive ventricular septal defect (VSD) with consequent cyanosis (oxygen saturation = 75%), transposition of the great arteries (TGA), and pulmonary stenosis has been followed up for 20 years in our outpatient center. She was stable on her first visit. The examination revealed central cyanosis and a systolic murmur in the second left intercostal space and parasternal on the right side. On ECG, she had regular sinus rhythm, atrioventrivular (AV) block I, and right bundle branch block. Transthoracic echocardiography showed a heart with single ventricular physiology (82 mm) resulting from bidirectional VSD, ventricular wall — 13 mm, TGA with pulmonary trunk stenosis (max gradient 102 mm Hg), and its post-stenotic widening (Figure 1). The function of both ventricles was moderately impaired. Pulmonary function was preserved with no future of restriction in spirometry.

During the first 5 years of follow-up, the patient's condition was stable (New York Heart Association [NYHA] class II), and there were only single ventricular and supraventricular extrasystoles. The first severe clinical complication appeared in 2008, with a well rehabilitated ischemic stroke. Aspirin 75 mg/day was used. Subsequent hospitalization took place in 2019 due to worsening heart failure (HF) and pre-syncope, which resulted from significant posthemorrhagic anemia caused by massive epistaxis during home oxygen therapy, aggravated by thrombocytopenia. Aspirin was discontinued. I.e. a further incident leading to decompensation bleeding occurred after tooth extraction in 2020. In both cases, clinical improvement followed monitored HF treatment (low doses of diuretic, beta-blocker, aldosterone antagonist), blood transfusion, and iron supplementation. Two hospital admissions in 2022 were caused by HF exacerbation due to the deterioration of ventricular systolic function. Due to unsatisfactory improvement after diuretic treatment, levosimendan was added, making the patient's general condition better with decreased N-terminal prohormone of brain natriuretic peptide (NT-proBNP). The last hospitalization (2023) occurred because of aggravation of the hemodynamic conditions. Apart from diuretics, the patient required catecholamines and intravenous iron supply. The patient, treated with these preparations orally, remains in home therapy under our supervision, in serious but stable condition.

Unoperated univentricular patients reach adulthood only when there is a hemodynamic balance between two-way communication between the systemic and pulmonary circulation, which ensures sufficient oxygen supply within pulmonary stenosis and protects against severe pulmonary hypertension [2]. In our patient, over the years, inevitable complications have appeared due to leakage between the cavities (increasing pulmonary resistance, impaired ventricular function) and cyanosis, mainly in the form of thromboembolic complications, which are typical of these patients, due to an increase in blood viscosity resulting from chronic hypoxia [3]. The main management strategy is to provide medical care that will not disturb their fragile pathophysiological balance. It is important to use anticoagulants carefully and only in exceptional situations. Hemoglobin should be maintained at a higher level than the normal range, using iron supplementation for this purpose. Heart failure should be treated with

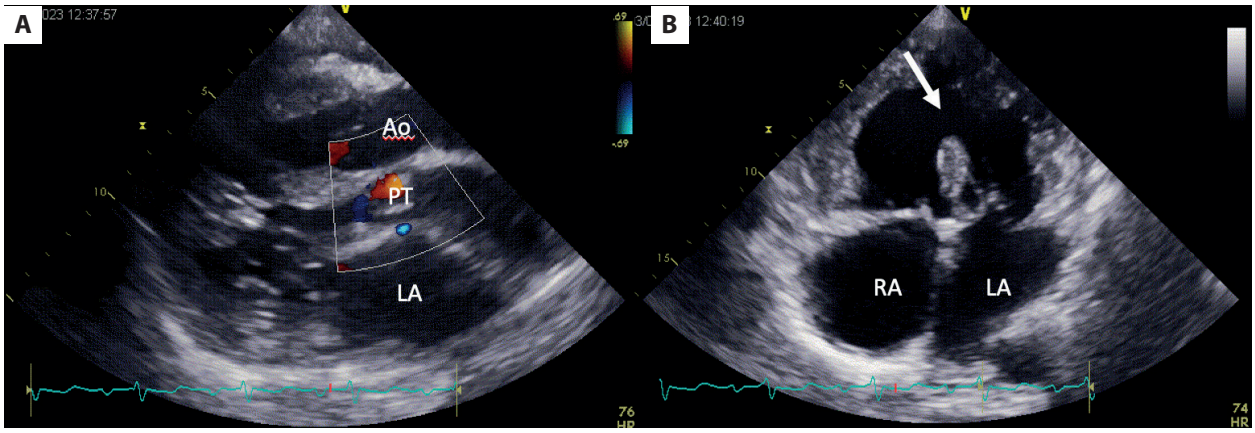

Figure 1. A. Echocardiography, parasternal long axis view. **B.** Echocardiography, four-chamber apical view; arrow indicates the ventricular septal defect

Abbreviations: Ao, aorta; LA, left atrium; PT, pulmonary trunk; RA, right atrium

all possible and available drugs. However, excessive reduction of preload (diuretics, angiotensin-converting enzyme inhibitors) and high doses of negative inotropes should be avoided [4]. These patients should have close contact with an adult congenital heart disease center [5].

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/kardiologia_polska.

Article information

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REFERENCES

1. Gesuete V, Fabi M, Bonvicini M. A 74-year-old unoperated univentricular heart: the oldest reported survival. Cardiol Young. 2016; 26(4): 805–807, doi: 10.1017/S1047951115001973, indexed in Pubmed: 26508431.
2. Rao PS. Diagnosis and management of cyanotic congenital heart disease: part I. Indian J Pediatr. 2009; 76(1): 57–70, doi: 10.1007/s12098-009-0030-4, indexed in Pubmed: 19391004.
3. Trojnarska O, Szczepaniak-Chichel L, Gabriel M, et al. Arterial stiffness and arterial function in adult cyanotic patients with congenital heart disease. J Cardiol. 2017; 70(1): 62–67, doi: 10.1016/j.jjcc.2016.09.003, indexed in Pubmed: 27756510.
4. Cordina RL, Celermajer DS. Chronic cyanosis and vascular function: implications for patients with cyanotic congenital heart disease. Cardiol Young. 2010; 20(3): 242–253, doi: 10.1017/S1047951110000466, indexed in Pubmed: 20416139.
5. Baumgartner H, De Backer J, Babu-Narayan SV, et al. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2021; 42(6): 563–645, doi: 10.1093/eurheartj/ehaa554, indexed in Pubmed: 32860028.