

## CLINICAL VIGNETTE

# Oligosymptomatic cyanotic heart disease in an adult with a univentricular heart

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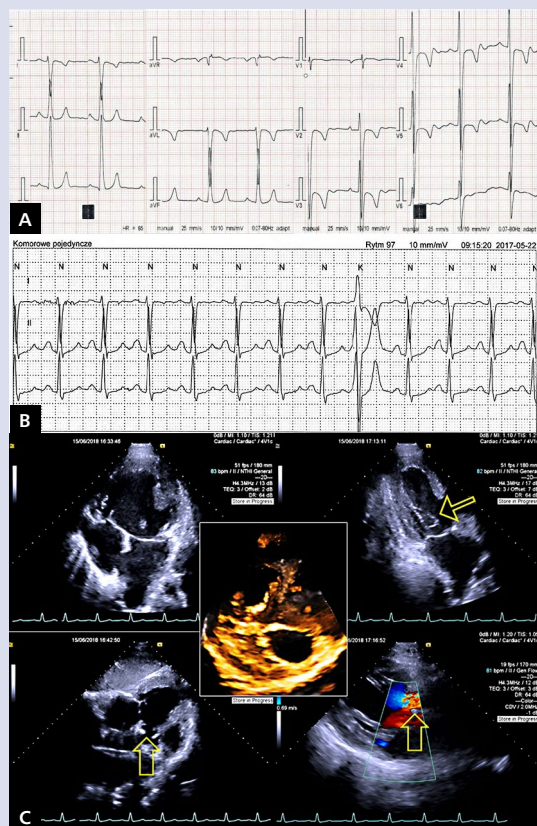
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A 27-year-old, non-smoking, normotensive man in New York Heart Association functional class I with moderate central cyanosis and loud systolic murmur at Erb's point presented for a check-up in an outpatient department. He was a physical worker with no reported exercise limitation and had no history of palpitations, peripheral oedema, dyspnea, syncope, or chest pain. Resting electrocardiogram (ECG) revealed normal sinus rhythm (SR) of 67 bpm, right axis deviation, biventricular hypertrophy (Katz-Wachtel sign), as well as PR interval of 0.2 s, QRS of 0.12 s, and QTc of 0.415 s (within reference range) (Fig. 1A). Ambulatory (Holter) ECG monitoring showed SR of 75 bpm, with 31,000 monomorphic ventricular extrasystoles forming long periods of ventricular bigeminy and trigeminy, and 170 ventricular couplets, without ventricular tachycardia. After initiation of 25 mg metoprolol succinate per day, ECG monitoring showed only 37 premature ventricular beats (Fig. 1B). The most relevant recent laboratory test results included N-terminal pro-B-type natriuretic peptide (180 pg/mL), haemoglobin (16.5 g/dL), haematocrit (47.4%), and red blood cell count ( $5.1 \times 10^6/\mu\text{L}$ ), without other abnormalities. For personal reasons, the patient refused to undergo cardiopulmonary exercise testing, but he reported optimal exercise tolerance and was able to regularly cover a 30-km distance on a bicycle. On imaging, posteroanterior chest X-ray showed prominent hila with features of increased pulmonary flow. Echocardiogram revealed a severe structural anomaly: a univentricular heart. Two separate atrioventricular valves (with trace regurgitation only) were connected with a double inlet common chamber with left ventricular morphology (DILV), with a typical rudimentary right ventricle connected via nonrestrictive 20-mm ventricular septal defect (VSD). The great arteries were transposed with aorta in anterior and rightward orientation (type II DILV) [1] and there was coexisting pulmonary valve stenosis (with possible fibrotic subpulmonary component) with a peak pressure gradient of approximately 64 mmHg — thus we considered the patient's pulmonary pressures as moderately elevated (Fig. 1C). The term “univentricular heart” refers to a group of congenital heart defects in which the inflow into the dominant chamber originates from both atria, and the second chamber is usually severely hypoplastic, connected by nonrestrictive VSD (sometimes called “foramen bulboventriculare”). In our patient, the anatomy could be more completely described as situs solitus: double inlet single ventricle of left ventricular morphology with outlet chamber and transposition of the great arteries. Patients with unoperated univentricular heart have a 10%–30% chance to survive one year [2], and the median survival is 14 years. Until 2016 only 14 cases of unoperated univentricular heart surviving over 50 years of age were identified; all of them with DILV, usually with transposition of great arteries [3]. The longest described survival was 77 years. In the context of the Mayo Clinic group report, the decision to treat the patient conservatively seems justified because the average survival in persons reaching adulthood without surgery is

55 years, and the probability of surviving without a heart transplant is 65% at the age of 50 years. A typical age-related problem is supraventricular arrhythmia, however, unusually, our patient had ventricular ectopy, with good response to  $\beta$ -blocker therapy. Our report describes a rare case of a “haemodynamically optimal” version of a severe congenital heart defect, emphasising the individual approach to monitoring for arrhythmia and heart failure development. Importantly, our patient showed all features favouring a mild clinical course: DILV morphology, transposition, atrioventricular valve competence, and moderate stenosis of the right ventricular outflow tract, preventing excessive cyanosis but also inadequate chamber overload.

## References

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**Figure 1.** A. Resting electrocardiogram showing normal sinus rhythm and biventricular hypertrophy (Katz-Wachtel phenomenon with bidirectional QRS > 50 mm in precordial leads); B. Holter monitoring before initiation of metoprolol, showing ventricular ectopy; C. Trans-thoracic echocardiography demonstrating double inlet common chamber with left ventricular morphology and two separate atrioventricular valves in apical four-chamber view (top left) and in two-chamber view (top-right); accessory mitral tissue indicated by the arrow; subcostal view shows stenotic right ventricular outflow tract with dysplastic pulmonary valve (arrow, bottom left) causing flow acceleration and turbulence, recorded from parasternal long axis view (bottom right); three-dimensional view presents adjacent atrioventricular valves seen from mid-left ventricular cavity (centre)

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## Conflict of interest: none declared

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