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A successful pulmonary artery banding in an infant with idiopathic dilated cardiomyopathy and severe heart failure

Short title: Pulmonary artery banding in dilated cardiomyopathy and severe heart failure

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Childhood dilated cardiomyopathy (DCM) remains a challenge of paediatric cardiology. The 5-year survival free of heart transplant is around 50% with highest death rate in children presenting <1 year of age [1]. High waitlist mortality and shortage of smaller donors led to the seek for alternative solutions. Pulmonary artery banding (PAB) in DCM was firstly described in 2007 [2]. Since then, the method proved to be effective, especially in infants. However, it is still not commonly used worldwide. Less than 20 centres presented their results until 2022 [3]. The largest study published so far showed promising results with 34/70 patients experiencing complete left ventricle (LV) functional recovery [4]. The hemodynamic changes induced by PAB result from positive right and left ventricle interactions. The increase in right ventricle

wall stress results in leftward shift of intraventricular septum and LV reshaping that further leads to improvement in diastolic and systolic function [5]. However, the exact mechanism on cellular level remains unknown. It is assumed that changes induced by PAB have the potential to reactivate proliferative potential of cardiomyocytes [3].

We report a case of successful PAB in an infant with idiopathic DCM, despite severe end-stage heart failure (HF) prior to the procedure. The boy was born after 38th week of gestation with birth weight of 2700 grams. He remained healthy until the age of six months old, when the symptoms of HF developed and he was referred to Pediatric Cardiology Clinic. Physical examination showed pale skin, severe work of breathing, tachycardia, hepatomegaly. Echocardiography revealed severely enlarged LV with ejection fraction (EF) of 10% and moderate mitral regurgitation. CT scan excluded coronary artery abnormalities. No other specific cause of cardiomyopathy could be found, including carnitine deficiency, most common metabolic diseases and viral infections. Magnetic resonance imaging showed enlarged LV with EF of 8%. The study excluded non-compaction cardiomyopathy, but showed areas of late gadolinium enhancement characteristic for DCM.

The clinical state of the boy deteriorated, despite intensive treatment. He was qualified on the heart transplant waiting list. However, due to further deterioration, the patient was qualified by the heart team for PAB operation. Final pre-surgical echocardiography showed EF of 11% and global longitudinal LV strain of -4% (Figure 1A-C; Supplementary material, *Videos S1* and *S2*). At the age of 9-months old the child underwent PAB. The pulmonary artery diameter was reduced by 50% resulting in right ventricular pressure increase to 55% of systemic arterial pressure. After the operation the clinical state of the patient slowly improved. Two months postsurgery the intravenous drugs were withdrawn and after another two months the boy was discharged home with oral HF treatment.

During one year of follow up we observed outstanding improvement in LV size and function. Echocardiography at the age of 20 months showed EF of above 60% and global longitudinal strain of –19% (Figure 1D–F; Supplementary material, *Videos S3* and *S4*). This improvement was also confirmed by magnetic resonance imaging. The boy developed well, with no signs of HF and was withdrawn from heart transplant waiting list.

Supplementary material

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

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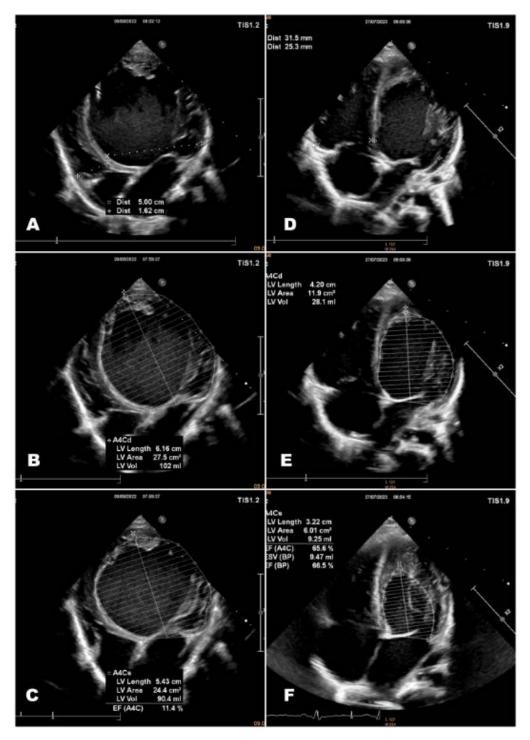


Figure 1. Echocardiographic apical 4-chamber view. A, B, C. Examination prior PAB. D, E,
F. Examination 12-months post PAB. A, D. Right and left ventricle end diastolic diameters. B,
E. LV end diastolic volume. C, F. LV EF

Abbreviations: A4Cd, apical 4-chamber view, diastole; A4Cs, apical 4-chamber view, systole; BP, assessed in biplane (4-chamber and 2-chamber view); Dist, distance; EF, ejection fraction; ESV, end systolic volume; LV, left ventricle; Vol, volume