

# Potts shunt in child with suprasystemic pulmonary arterial hypertension

Zespolenie Potts'a u dziecka z ponadsystemowym tętnicznym nadciśnieniem płucnym

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Pulmonary arterial hypertension (PAH) still remains a progressive disease with unfavourable prognosis. In cases of pharmacotherapy failure, lung transplantation is the only therapeutic option. In selected cases, palliative treatment — Potts shunt, may be performed. This surgical procedure results in the right-to-left shunt from left pulmonary artery to descending aorta and leads to decompression of the failing right ventricle, but causes a decrease in oxygen saturation in the lower part of the body. The patient described herein is the first child in Poland with idiopathic PAH (IPAH) in whom a Potts shunt was performed. The 2-year-old female was admitted to the Paediatric Cardiology Department of the Children's Memorial Health Institute because of chest pain and shortness of breath. On examination, cyanosis with blood oxygen saturation ( $\text{SaO}_2$ ) 87%  $\text{HbO}_2$  and symptoms of right heart failure (World Heart Organisation Functional Class [WHO-FC] II/III) were observed. N-terminal pro-B-type natriuretic peptide (NT-proBNP) level exceeded 9000 pg/mL. Echocardiography revealed no congenital heart disease and the probability of pulmonary hypertension. Based on right heart catheterisation (RHC), IPAH with suprasystemic pulmonary arterial pressure was diagnosed. Treatment with bosentan, sildenafil, and acenocoumarol was introduced. The patient remained stable at WHO-FC II until 15 months after diagnosis, when she developed loss of appetite, abdominal pain, and irritability. On echocardiography, pericardial effusion was present, left ventricle diastolic dimension decreased to 59% of normal value (Fig. 1), and estimated pulmonary arterial systolic pressure (PASP) was 95 mm Hg (systemic pressure 73/50 mm Hg). NT-proBNP level was 5888 pg/mL. Treatment with digoxin, diuretics, and then iloprost was added. Nevertheless, a few days later, during viral respiratory tract infection, shortness of breath, cyanosis ( $\text{SaO}_2$  80–85%  $\text{HbO}_2$ ), chest pain, ventricular arrhythmia, and low cardiac output symptoms (WHO-FC IV) were observed. Iloprost was changed to an epoprostenol IV. Because of considerable deterioration of the patient's general condition, RHC was not performed. Based on previous diagnosis, clinical status, and non-invasive exams, the patient was qualified to Potts shunt. The procedure was performed in the University Children's Hospital of Krakow. Eight days after surgery, epoprostenol was stopped. The patient was discharged 18 days after surgery with WHO-FC II and still on bosentan, sildenafil, acenocoumarol, and diuretics.  $\text{SaO}_2$  on the upper limb was 98%, on lower limb 66%  $\text{HbO}_2$ , and the NT-proBNP level was 1411 pg/mL. Fourteen months later, the patient is in WHO-FC I.  $\text{SaO}_2$  on the upper limb is 100%, on the lower 95%  $\text{HbO}_2$ , and NT-proBNP is 60 pg/mL. On echo, the left ventricle dimension is at normal value, without pericardial effusion, but estimated PASP remains suprasystemic (Fig. 1). In the presented patient, Potts shunt is a treatment allowing longer survival and better quality of life. Echo images illustrate very well the haemodynamic status just before and after surgery.

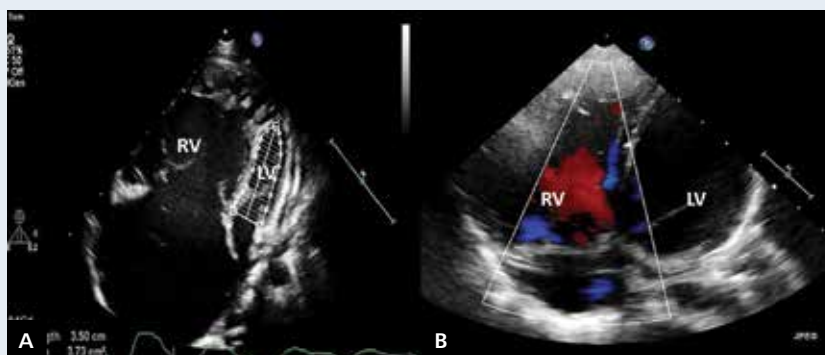


Figure 1. Echocardiography — four-chamber view; **A.** Before Potts shunt; **B.** Eight months after; RV — right ventricle; LV — left ventricle

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