

Management of plastic bronchitis after Fontan operation with resolution of symptoms

Postępowanie w przypadku plastycznego zapalenia oskrzeli po operacji Fontana z ustąpieniem objawów

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Plastic bronchitis (PB) is characterised by formation of bronchial casts, leading to airways obstruction and pulmonary failure, and is sometimes reported after Fontan procedure. The proper diagnosis is usually made late and the treatment is still challenging. A four-year-old girl with hypoplastic left heart syndrome palliated at the age of 2.5 years with Fontan procedure (extracardiac conduit) was referred to our hospital. One and a half years after procedure she developed cyanosis, dyspnoea, and intensive cough. After three months of ineffective therapy, she expectorated a yellowish gelatinous bronchial cast, which allowed diagnosis of PB. She was treated with ambroxol, N-acetylcysteine, ipratropium bromide, salbutamol, fenoterol, nebulised saline (3%), and intensive respiratory rehabilitation. A chest X-ray showed emphysema in peripheral parts of both lungs. Echocardiography revealed a stenosis of the left pulmonary artery (LPA 4.2 mm) and good function of the systemic right ventricle. Cardiac catheterisation revealed a gradient of 2–3 mm Hg through the hypoplastic LPA, transpulmonary gradient of 5 mm Hg, and pressure in the Fontan conduit of 9 mm Hg. An 8 × 18-mm stent was implanted into the LPA. The child was discharged with inhalations of 5% NaCl, salbutamol, dornase alfa, and heparin. Almost two years later, she remains free from any symptoms and currently receives only inhalations of heparin twice a day. A 12-year-old girl with mitral atresia and ventricular septal defect was admitted with PB diagnosis. When she was 3.5 years old, she underwent fenestrated Fontan procedure (extracardiac conduit). Interventional closure of fenestration was done 1.5 years later. Seven months before admission she developed persistent cough diagnosed as asthma or infection. After 1.5 months she started to expectorate large solid casts (10–12 cm) and yellowish liquid, which suggested the PB diagnosis. For 5.5 months she was treated with nebulised tissue plasminogen activator, switched to aerosolised heparin. That led to her bringing up smaller casts instead of large and solid (Fig. 1) with a reduction in the amount. She received sildenafil, nebulised dornase alfa, salbutamol, and steroids but without improvement. At admission she presented good general condition with oxygen saturation of 93%. Cardiac catheterisation revealed mean pressure in the Fontan conduit of 10 mm Hg and transpulmonary gradient of 6 mm Hg, and a re-fenestration of Fontan conduit was made with an 8 × 18-mm stent implantation. The saturation after intervention decreased to 87%. She was discharged with aldactone, enalapril, warfarin, sildenafil, and inhalations of heparin. Two years later she remains free from symptoms, with saturations around 86–90%. Patients with PB after Fontan procedure should have detailed cardiological evaluation. Both interventional and pharmacological treatment should target cardiac output improvement. Fenestration of the Fontan conduit or stent implantation into a narrow pulmonary artery might decrease central venous pressure, augment ventricular filling, and improve cardiac output.



Figure 1. A. Cast directly after expectoration; B. Cast after a few minutes in water; C. Casts after treatment with tissue plasminogen activator

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