Pulmonary artery atresia with ventricular septal defect, segmental pulmonary hypertension, major aortopulmonary collaterals (MAPCAs) and giant MAPCA aneurysm

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A 32-year-old woman with pulmonary artery atresia with ventricular septal defect (VSD) was admitted because of New York Heart Association (NYHA) class worsening from II to III during the last 6 months. The patient presented resting cyanosis, short 6-minute walk test (6MWT) distance (370 m with desaturation – baseline: 84%, after 6MWT: 67%). N-terminal prohormone of brain natriuretic peptide (NT-proBNP) level was 194–262 pg/ml. She also suffered from significant kyphoscoliosis with stable chronic ventilation disturbances: forced vital capacity (FVC) = 1.38 l, forced expiratory volume in 1 s (FEV_1) = 0.94 l, FEV_1/FVC = 68%.

Imaging methods (transthoracic echocardiography, transoesophageal echocardiography, magnetic resonance imaging – MRI, angio-computed tomography – CT) confirmed the presence of VSD (Figures 1 A, B), enlarged ascending aorta up to 49 mm above VSD and moderate aortic valve insufficiency. Angio-CT and MRI revealed occurrence of major aortopulmonary collaterals (MAPCAs) (Figure 1 C) with a giant aneurysm of one of them (Figure 1 D) and no pulmonary artery. Coronarography showed abnormality of the left artery descdending (LAD) orifice from the right coronary artery. We simultaneously excluded coronary and mammary artery to MAPCA shunts. The aortography followed by selective catheterization showed five MAPCAs: three on the left side (the one that supplied the left lower pulmonary lobe was stenosed in the proximal part) and two on the right side, among others a MAPCA creating a giant (6 cm) aneurysm (Figure 1 E). Each individual part of the lungs is supplied from a single source. In catheterization using a Swan-Ganz catheter we obtained the following outcomes: the mean collateral pressure was high in the MAPCAs that were not stenosed (69 mm Hg) and there was no response to iloprost (Figure 1 F); the mean collateral pressure was quite low in the stenosed MAPCA (14 mm Hg). According to single case reports and limited research [1, 2] and our hemodynamic outcomes, we prescribed bosentan, with quite a good clinical effect. During bosentan therapy the patient's NYHA class decreased from III to II. The patient was disqualified from surgical treatment. In the literature we have found some reports of large MAPCA aneurysm diagnosed *in vivo* but not surgically treated [3] and one case concerning successful invasive treatment [4]. During 2-years follow-up we increased the dose of bosentan to 125 mg twice daily and we obtained a very good result. The patient is still in NYHA functional class II without any progression of pulmonary hypertension symptoms. There still remains the question whether the above described MAPCA aneurysm should be percutaneously occluded or surgically treated in prevention of aneurysm rupture and sudden death. Taking into account coexisting kyphoscoliosis and ventilation disturbances, the risk of interventions seems to be very high.

Conflict of interest

The authors declare no conflict of interest.

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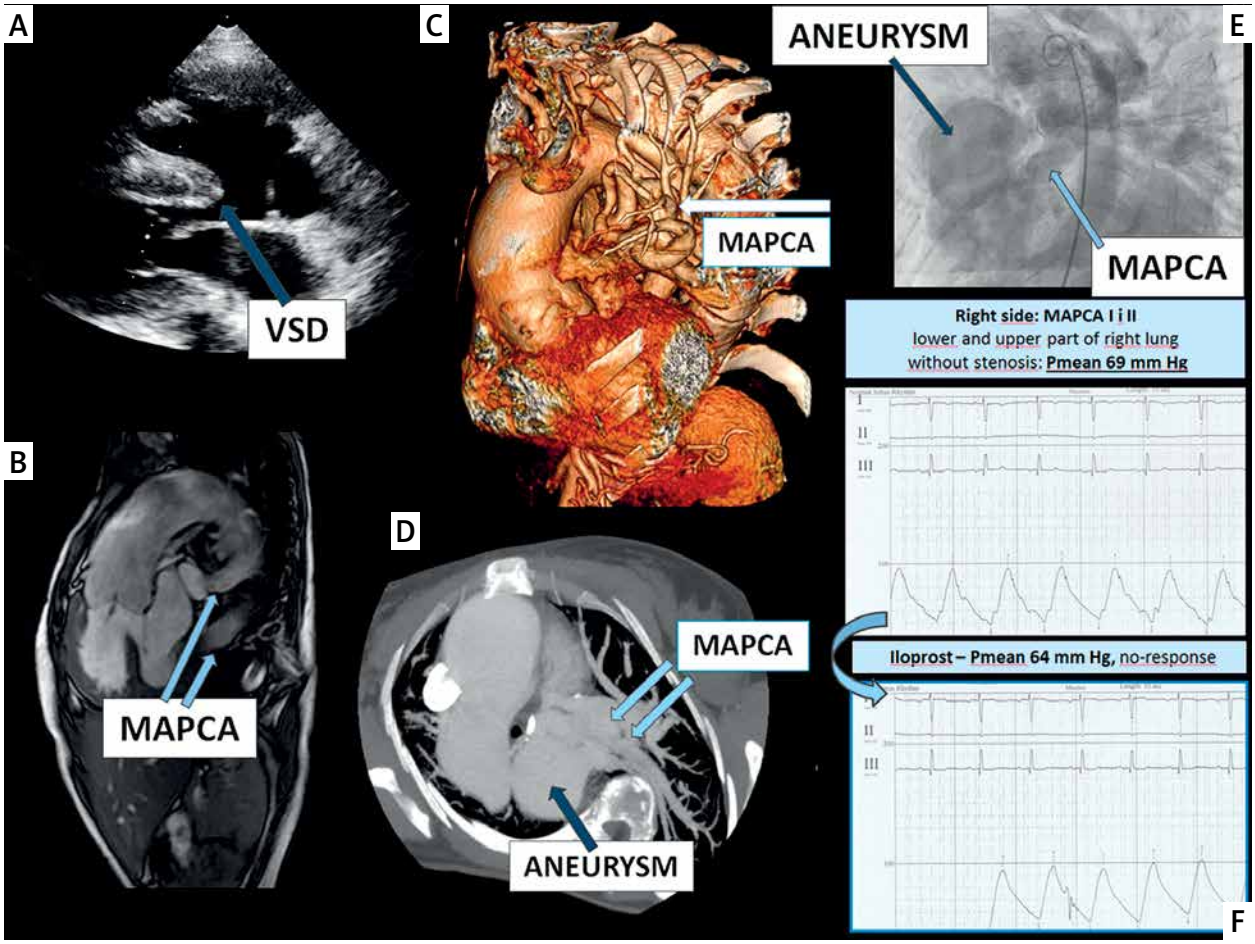

Figure 1. Multimodality imaging: **A** – transthoracic echocardiography – parasternal long axis view, arrow: large VSD; **B** – CT scan, arrows: MAPCAs; **C** – CT scan, 3D reconstruction, arrow: MAPCA; **D** – CT scan, arrows: MAPCAs, arrow: large aneurysm; **E** – angiography, arrows: MAPCA, aneurysm; **F** – vasoreactivity test with no response for iloprost

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