Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

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CASE SUMMARY

Thirty one year old female with a complex congenital heart defect (CHD) involving common truncus arteriosus (CTA) and ventricular septal defect (VSD) and systemic to pulmonary artery shunts, was referred to the department of cardiology with acute decompensation of chronic heart failure to NYHA class IV accompanied by respiratory tract infection and the suspicion of infective endocarditis. CHD was diagnosed in the neonatal period and subsequently confirmed by means of cardiac catheterization at the age of two years. Past medical history was positive for infective endocarditis in 2002. Physical examination revealed arterial blood pressure of 90/60 mmHg, regular heart rhythm 85 per minute, central cyanosis, clubbed fingers, systolic heart murmur. Transthoracic echocardiography (TTE) denoted CTA with diameter of 50 mm and tricuspid valve fraught with mild insufficiency, as well as preserved global and regional left ventricular systolic function (ejection fraction of 65%), myocardial hypertrophy (posterior wall thickness 14 mm, inter-ventricular septum 16 mm, right ventricular wall thickness 12 mm) and VSD with a diameter of 20 mm in the membranous part of inter-ventricular septum. Performed cardiac magnetic resonance (CMR), which enabled to visualize VSD concerning both membranous and muscular part of inter-ventricular septum, and common truncus arteriosus originating from above the VSD with a maximal diameter of 55 mm combined with tricuspid valve (orifice size 52x55 mm), as well as physiological origin of cephalic arteries. Computed tomography angiography (CT angiogram) was conducted so as to evaluate the anatomy of pulmonary arteries and to exclude the possibility of pulmonary artery stenosis. CT angiogram confirmed previous TTE/CMR findings related with the morphology of CHD and additionally revealed anomalous origin of pulmonary arteries from descending aorta (predominantly) and the suspicion of anomalous origin and number of coronary arteries, whereas no pulmonary

artery stenosis was documented. Subsequently, an attempt to assess pulmonary arterial pressure during heart catheterization led to the discovery of venous system anomaly consisting of venous conglomerate draining to vena cava inferior. Aortography performed through right femoral access showed wide common truncus arteriosus with physiological branching of aortic arch and two pulmonary arteries originating from descending aorta below the aortic arch. Selective catheterization of pulmonary arteries was not amenable to perform due to unfavorable branching from aorta documented after unselective contrast medium administration to descending aorta. Invasive hemodynamic measurements allowed for evaluation of blood pressure in truncus arteriosus (114/55/78 mmHg) and femoral artery (118/54/77 mmHg). The patient was discharged from the department on hitherto pharmacotherapy (bisoprolol 2.5mg q.d., spironolactone 25 mg q.d.)

DISCUSSION

The median survival of patients with truncus arteriosus is five weeks of age with rare cases reaching adulthood (1). Surgical correction is the treatment of choice. Truncus arteriosus babies are treated by closure of VSD along with right ventricle to pulmonary artery conduit. Feasibility, safety and effectiveness of treatment of cyanotic CHD with currently available medical, transcatheter and surgical methods are well established and should be performed at an appropriate age in order to prevent damage to cardiovascular structures (2). Bosentan is recommended for symptomatic patients with Eisenmenger syndrome due to simple congenital lesions. However, its long-term efficacy and safety in patients with pulmonary arterial hypertension (PAH) associated with complex congenital heart disease (CHD) is unknown. Baptista R et all shown that Bosentan was safe and was associated with improved exercise capacity in patients with PAH and complex CHD. This improvement was sustained for up to four years and the safety profile was similar to simple CHD patients (3).

EXPERT'S OPINION

] Given the complex inoperable cyanotic congenital heart defect highly suspected of pulmonary hypertension, a consideration should be given to vasodilator treatment with bosentan under National Healthcare Provider programme

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