**Article\***

"Development of the European Network in Orphan Cardiovascular Diseases"

„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych”

**Title:** Clinical assessment of patients with pulmonary hypertension associated with congenital heart disease

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Initial evaluation of a patient with pulmonary hypertension associated with congenital heart disease includes history, physical examination, ECG, and echocardiography however a definitive diagnosis requires invasive hemodynamic study. Additional tests include: exercise testing, chest radiography, ventilation/perfusion scan, computed tomography angiography, cardiovascular magnetic resonance, laboratory tests, and pulmonary function tests.

Typical symptoms of pulmonary hypertension include exertional dyspnea, fatigue, hemoptysis, angina, presyncope, and syncope. No special symptoms are specific for patient with pulmonary hypertension associated with congenital heart disease. Physical examination may reveal a parasternal lift, highpitched tricuspid regurgitation murmur, widely split S2, pulmonary ejection sound, or loud P2.

Pulmonary hypertension is usually first suspected based on echocardiography. In patients with tricuspid regurgitation right ventricular systolic pressure can be estimated. Additionally performance of right ventricle can be evaluated.

Symptomatic patients with congenital heart disease who are suspected for pulmonary hypertension should undergo cardiac catheterization. It is essential to confirm pulmonary hypertension and to understand the pathophysiology of the disease. To determine cardiac output the Fick method is recommended with direct measurement of O2 uptake at the time of catheterization. The procedure should be performed by a specialist experienced in pulmonary hypertension associated with congenital heart disease.

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