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**Clinical presentation and hemodynamics of adults with Eisenmenger syndrome and idiopathic pulmonary hypertension**

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**Background:** ES represents a very specific form of PAH. Unlike patients with IPAH, in Eisenmenger syndrome the clinical and cardiac status is often relatively stable for a years of clinical course

**Methods:** We prospectively recorded with clinical parameters, 6-minute walking test (6MWT), echocardiography, right heart catheterization (RHC), cardiopulmonary exercise test (CPET), laboratory tests of 40 patients (mean age 42±12,2 years, 34 women and 6 men, diagnosed with PH): 20 patients with PAH-CHD and Eisenmenger syndrome (ES) (aged 40±15 years) and 20 patients with IPAH (aged 42±9 years) evaluated in our pulmonary hypertension (PH) centre according to the data of Russian National Registry of Patients With Pulmonary Arterial Hypertension (NCT03707561).

**Results:** Patients were followed for 24 months. The average time from the occurrence of complaints till the final diagnosis was 63,8 months for PAH-CHD and 39,1 months for IPAH. The main complaints were dyspnea (95%) in both groups but in patients with IPAH such complaints as chest pain (65%, p=0,004), dizziness (80%, p=0,004), syncopes (60%, p=0,001), palpitation (85%, p=0,04) were significantly more frequent, and for PAH-CHD the most significant complain was hemoptysis (30%, p=0,008). According to RHC mean PAP and PVR were significantly higher in patients with ES (81±15,2 mm Hg against 58,5±12,3 mm Hg, p=0,0002) and PVR (17,7±8,45 Wood ES had greater systemic cardiac indexes (3,3± 0,4 versus 2,2±0,5 L/min/m2, p <0,005) and lower mean right atrial pressures (4± 2 versus 7± 5 mm Hg, p < 0,0001) than patients with IPAH.
According to CPET load time was significantly higher in a group with IPAH (9,3±4 minutes against 5,7±1,7 minutes, p=0,003). The significant positive correlation between distance in 6MWT and Vo2peak (r=0,67, p<0,05) and significant negative correlation between cardiac index and VE/VCO2 were found (r=-0,68, p<0,05) in the group of PAH-CHD. The VE/Vco2 value had significant positive correlation with mean PAP (r=0,67, p<0,05) in a group with IPAH.

**Conclusions:** Relevant clinical, functional and hemodynamic differences were observed among groups. Our data suggest that adults with ES have more favorable hemodynamic profile which may result in better prognosis than adults with IPAH.