

Ukrainian Center for Pulmonary Arterial Hypertension in Children: Achievements and Problems.

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Introduction. Pulmonary arterial hypertension (PAH) in children is a hemodynamic and pathophysiological state characterized by increasing of mean pressure in the pulmonary artery above 25 mm Hg in patients elder than age of 3 months, and is estimated by the data of the catheterization of the right heart. In 2015 the Ukrainian Center for Pulmonary Arterial Hypertension (UCPAH) in Children was created at the Ukrainian Children's Cardiac Center. The aim of our study was to analyze the demographic and diagnostic spectrum of children in Ukraine with pulmonary hypertension.

Methods. The assessment of our patients included evaluation of clinical symptoms and anamnestic data, roentgenography of the chest, electrocardiography, investigation of the function of the lungs and gases of arterial blood, a 6-minute walk test (6 MW), echocardiography, catheterization of the right heart and a study of vasoreactivity, a high resolution computed tomography, a CT with contrast and pulmonary angiography, a MRI of the heart, blood tests (serology, immunology and genetics), an ultrasonography of the abdominal cavity. Unfortunately, a ventilation perfusion scanning of lungs is not available in Ukraine.

Results. During 2 year 156 patients with PAH were registrered in UCCC. There were 84 female and 72 male. All patients were with diagnosis of idiopathic PAH (iPAH) - 31 (19,8%) patients and PAH – associated with congenital heart disease (PAH-CHD) – 125 (80,2%) patients. In group of PAH-CHD 29 (18,5%) was after total repair of CHD, 52 (33,3%) – after palliative operations, 44 (28,4%) – non operative. During our investigation we analyzed their clinical condition, current medications and operability (in case of PAH-CHD). 57 patients with different CHD underwent of total repair, 32 – palliative procedures.

Conclusions. Creation of UCPAH in children helps in creation of a general register of patients with a pediatric group with PAH, carrying out of highly specialized diagnostics of PAH, optimisation of medical therapy, monitoring its effectiveness and assessing the dynamics of patients' clinical condition, providing cardiac surgery, provision of emergency care to patients with complications of PAH in ICU, ilncrease the level of knowledge about the diagnosis and treatment of children with PAH.