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Diagnostics and clinical patterns of idiopathic pulmonary hypertension in Russia. A two-center study.

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Objective: to identify clinical, functional, haemodynamic, and biochemical characteristics of idiopathic pulmonary arterial hypertension (iPAH) in children.
Methods: 90 pts with PAH (52% female) aged 4 months - 17 years studied in Jan 2011 – Dec 2013. Tests included routine cardiological examination; CT angiography & scintigraphy; HIV, thyroid, liver function tests; ruling out lupus, scleroderma, rheumatoid arthritis; endothelin & NT-proBNP measurements; six-minute walk test (6MWT – age 7+ only); right heart catheterization (RHC - in 76%).
Results. iPAH confirmed in 21 (23%, thereof 71% female). At first examination 24% had functional class (FC) II, 47.5% - FC III; 28.5% - FC IV. 76% had dyspnea aggravated by physical effort. Syncope registered in 4 pts; recurrent hemoptysis in 3 pts. Median duration of symptoms was 23 (2-48) months. Dilatation of the right heart chambers, elevated RV wall thickness, dilated main PA, short acceleration time of RV ejection, and abnormal end-diastolic septal curve found in 86%. RV ejection fraction decreased in 57% of pts (29.5±3.2%). Tricuspid annular plane systolic excursion (TAPSE) constituted 14.2±4.7 mm. Marked variability of mean pulmonary artery pressure (32 to 98 mmHg) and pulmonary vascular resistance (8.5 to 29.3 UW) were detected. Cardiac index was 1.9±0.5 L/min/m² and median arterial oxygen saturation was 91.5±2.7%. Elevated values of endothelin (0.43±0.18 pmol/ml) and NT-proBNP (64.8±18.6 pg/ml) were found. Two pts were treated with sildenafil, 13 pts - with bosentan and 6 pts - with a combination of the two drugs. In 5 pts the atrioseptostomy was performed. Median duration of follow-up was 36 months (2 to 42 months). Reduction of dyspnea and improvement of functional characteristics observed in 11 (52%) pts. 6MWT distance increased by 156.2±12.7 m. RV ejection fraction reached 32.1±4.5%, pulmonary vascular resistance decreased by 8.9±3.4 UW. Five IV class pts and two III class pts have died.
Conclusions. Late diagnostics of iPAH in children is common and was observed in 71% of pts. FC and hemodynamic parameters are the strongest predictors of survival in children with iPAH. Reduction of dyspnea and increased distance on the 6MWT are the main markers of a positive response to therapy.