Long-term experience with CTEPH treatment in the Czech Republic

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Background. Chronic thromboembolic pulmonary hypertension (CTEPH) is caused by chronic obstruction of pulmonary vessels by organized thromboembolic material. Majority of CTEPH patients are potentially candidates of surgical therapy – pulmonary endarterectomy (PEA). Final diagnosis, operability assessment and therapy of CTEPH should be concentrated in specialized centres with experienced CTEPH team.

Patients and methods. We studied the charts from all consecutive patients with CTEPH between 2001 and 2013 who were referred to our centre from the Czech Republic and Slovak Republic for consideration of PEA. Operability was assessed based on V/Q scintigraphy, hemodynamic parameters, pulmonary angiography, CT angiography and clinical data. PEA program was initiated in 2004. Residual/recurrent pulmonary hypertension was assessed by echocardiography at month 6 during follow-up (PASP estimation $\geq$ 40 mmHg). Medical therapy in inoperable patients or in patients with residual/recurrent pulmonary hypertension was used only when patients were included in the clinical trials with investigational drugs. Standard Kaplan-Meier methodology was applied in the analysis of patients survival.

Results. The CTEPH population included 401 patients ranging in age from 19 to 82, the mean age was 62.1 years, female:male ratio was 0.87, mean pulmonary artery pressure, cardiac index and pulmonary vascular resistance were 49.3±12.02 mmHg, 2,2±0,51/min/m2, and 9,6±4,3 Wood units, respectively. 75.8 % of patients were considered as operable, and 55.1 % patients underwent surgery with early mortality rate 8.1 %. Following PEA at month 6, there were significant improvements in NYHA class (pre 2.9 vs. post 1.4, P <0.001), right ventricular systolic pressure (pre 84.4±37.4 mmHg vs. post 37.4±19.76 mmHg, P <0.001), 6-minute walk distance (pre 310.0±121.8 m vs. post 506.9±228.0 m, P <0.001). Residual/recurrent pulmonary hypertension was detected in 34.4 % of patients after PEA. Specific vasodilatation therapy was used in 30.0 % of inoperable CTEPH patients and in 23.7 % of patients with residual/recurrent pulmonary hypertension after surgery. The 1-, 3-, 5- and 8-year survival rates after PEA were 88 %, 85 %, 80 % and 76 %, respectively. The 1-, 3-, 5- and 8-year survival rates in inoperable patients were 89 %, 76 %, 68 % and 57 %, respectively.

Conclusions. PEA is curative therapy for majority of CTEPH patients. Residual pulmonary hypertension rate is relatively low and long-term survival of operated patients is excellent in experienced centre.