Epidemiology of Pulmonary Hypertension Associated with Congenital Heart Disease (PH-CHD) in Latvia

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Introduction. Pulmonary hypertension is a serious complication in patients with congenital heart disease. The defect can work as a shunt by connecting the left and the right side of the heart. Most commonly the defect is a ventricular septal defect (VSD), but it can also be an atrial septal defect (ASD), a patent ductus arteriosus (PDA) or a combination of any of the previously mentioned defects. According to the current guidelines, in patients with WHO-FC II and III, first line monotherapy treatment is Ambrisentan, Bosentan or Sildenafil. The recommended treatment goal is a 6MWD result of ≥ 440 m.

Aim, Material and Methods. The aim of the research was to analyse the haemodynamic charasteristics, ECHO data, laboratory data, complications, and the efficacy of the treatment among the PH-CHD patients in Latvia. We retrospectively analysed the collected clinical data of 41 patients with CHD-PAH. A 6-minute walking test was done and information about complications was obtained from 30 patients. We included patients with a known existing or surgically corrected intra- or extra-cardiac shunt with a mean pulmonary artery pressure (mPAP) of 25 mmHg and more.

Results. Data of 41 patients were analysed, 30 female and 11 male with mean age of 54.63 ± 17.90 with the youngest being 22 and the oldest – 80 at the time of the study. Mean mPAP was 57.15 ± 22.58 mmHg, range 25–106 mmHg. Mean systolic PAP was 87.80 ± 32.14 mmHg (range 37–145 mmHg). Mean PVR result was 10.31 ± 7.46 WU with results varying from 1.36 to 22.60 WU. Mean RAP was 12.43 ± 7.50 mmHg, range 3–32 mmHg. Mean 6MWD result was 358.50 ± 111.06 metres (range 148–543 m). Seven patients achieved a result of ≥ 440 m and two – ≤ 165 m. The mean BNP result was 251.27 ± 320.69 ng/l (17.6–1601 ng/l). Mean bilirubin level was 22.86 ± 16.72 μmol/l (6.2–90.00 μmol/l). DLCO2 results were not available for most patients, thus they were discarded.

Conclusions. Most (48.8%) patients were WHO-FC III (150–425 m in 6MWD). Seven patients achieved a 6-minute walking distance result of over 440 metres; only one could do less than 150 m. All of the patients that achieved a 6MWD of ≥ 440 m had taken either Sildenafil or Ambrisentan.