1st Baltic Pulmonary Hypertension Conference

ABSTRACTS

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CONTENT

LEFT MAIN CORONARY TRUNK COMPRESSION BY DILATED PULMONARY ARTERY IN PATIENT WITH SEVERE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Key words: Chronic thromboembolic pulmonary hypertension, left main coronary artery, dilated pulmonary artery, stable angina, extrinsic compression of left main, percutaneous coronary intervention (PCI).

Introduction: Angina-like chest pain is a common symptom in patients with severe pulmonary hypertension. The etiology of these symptoms for this group of patients may be associated not with coronary arteriosclerosis (if patient is too young for cardiovascular risk), but due to the severe dilatation of the pulmonary artery, right ventricular ischemia or the extrinsic compression of the left main trunk by the pulmonary artery. We would like to present the first case in our center of successful stenting of the left main coronary trunk compressed by dilated pulmonary artery.

Case report: 47 years old woman started to complain of breathlessness and chest pain on exertion (walking distance 200 meters) since September of 2012. Heart ultrasound revealed signs of severe pulmonary hypertension.

The patient was sent to our Pulmonary hypertension referral centre for more detailed diagnosis where severe chronic thromboembolic pulmonary hypertension was diagnosed after following examination:

- 6 minutes walking test – 200 meters.
- BNP 332.1 ng/L.
- Multislice computed tomography – A markedly dilated main pulmonary artery 50x55 mm, RPA ~ 38 mm, LPA ~ 33 mm. Chronic thromboembolic disease of small (subsegmental) pulmonary branches. The signs of extrinsic compression of LM by dilated main pulmonary artery.
- Right heart catheterization – pressures: Pulmonary artery 95/40/60 mmHg, Pulmonary artery wedge 13/2/9 mmHg; Cardiac output 6,93 L/min, Resistance 7,39 Wood, 591 dyn•s/cm^5. Conclusion: Severe precapillary pulmonary hypertension.
- Coronarography – revealed moderate-severe stenosis of LM coronary artery compatible with extrinsic compression of LM on CT.

Our team discussed the treatment of this patient. Once the patient wasn't suitable for surgical pulmonary thromboendarterectomy it was decided to perform PCI of LM in order to avoid sudden cardiac death. LM coronary artery was stented by drug eluting stent (DES) Biomatrix 4.0x8 mm 14 A with postdilation of ostium with 4,0x8 balloon 18 A. After PCI of LM clinical signs of stable angina disappeared, 6 minutes walking test improved up to 360 meters. Coronarography was repeated after 6 months of PCI and no signs of in stent restenosis were found.

Conclusions. PCI of the compressed LM is a method to treatment angina and a way to avoid sudden cardiac death in patients with severe pulmonary hypertension.

FACTORS INFLUENCING SURVIVAL IN PATIENTS WITH PULMONARY HYPERTENSION

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Key words: pulmonary hypertension, survival, 6-minute walking test, NT-proBNP, risk factors.

Pulmonary hypertension (PH) - is a pathophysiological disorder with the increase in mean pulmonary arterial pressure (PAPm) ≥25 mmHg at rest as assessed by right heart catheterization (RHC). Although the disease usually progresses and right heart failure occurs rapidly the survival ranges from several months to several years.

The aim of the study: to investigate whether patients gender, age during PH diagnosis, NT-proBNP level, 6-minute walking test (6MWT) result and etiology of PH are associated with survival in patients with PH.

Matherials and methods. Data from the medical charts from the single university hospital were collected and retrospectively analyzed. 61 case of PH (idiopathic pulmonary artery hypertension (IPAH), associated with connective tissue disease, associated with Eisenmenger's syndrome and chronic thromboembolic pulmonary hypertension (CTPH)) confirmed by RHC in 2003 – 2016 and included in the study. There were 42 women and 19 men. Influence of gender, age during diagnosis, NT-proBNP level, 6MWT results and etiology of PH on survival was assessed. All statistical analyses were performed using Statistical Package for the Social Sciences (SPSS), version 20.0. The Kaplan-Meier method with the log-rank test and Cox proportional hazards model were used to calculate survival rates, differences in survival curves and estimate the risk of death, and *P* values of <0.05 were considered to indicate statistical significance.

Results: Median of overall survival of all cases was 65 months (95 % CI 43.1- 86.8). There was no dependence of survival and patients gender (p= 0.699). In patients with age more than 60 years old, risk of death was increased 3.297 times (95% CI Exp(B) 1.282-8.482) (p=0.013) comparing with the group of patients with age less than 60. There was no dependence between survival and 6MWT result. Risk of death was also higher 5.6 times (95% CI Exp(B) 1.368- 22.792) (p=0.017) when NT-proBNP was ≥1500 ng/l, comparing with the group of patients with NT-proBNP < 1500 ng/l. Median survival in IPAH cases was 75, in CTPH- 47, in Eisenmenger's syndrome- 91, in PH associated with connective tissue disease- 31 month. Survival was dependant only on PH associated with connective tissue disease but no other etiologies of PH. There was statistically significant shorter survival in PH associated with connective tissue disease comparing with IPAH (p=0.019) and Eisenmenger's syndrome (p=0.01). It was also found that risk of death was higher for patients with PH associated with connective tissue disease comparing with IPAH (p=0.014) and Eisenmenger's syndrome (p=0.019) and the risk of death was smaller for patients with IPAH comparing with Eisenmenger's syndrome (p=0,014).

Conclusions: Survival in PH and risk of death was associated with age, disease etiology and NT-proBNP level. There was no association between survival and gender, 6MWT.

Summary: The survival in patients with PH decreases and risk of death increases, when PH is diagnosed in older age, when there is higher concentration of NT-proBNP and PH is associated with connective tissue disease. Risk of death is also lower in cases of IPAH.

PULMONARY ARTERIES DIAMETERS MEASURED BY CMR CAN PREDICT VASCULAR ELASTICITY IN PATIENTS WITH PULMONARY HYPERTENSION

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KEY WORDS:
Pulmonary hypertension, pulmonary artery diameter, pulmonary artery elasticity, pulmonary artery relative area change, cardiac magnetic resonance imaging.

INTRODUCTION:
Pulmonary hypertension (PH) characterized by abnormally elevated pressure in pulmonary artery (PA) related to structural changes in the PA – increased diameter and decreased elasticity. Previous studies of cardiac magnetic resonance (CMR) imaging showed that PA elasticity expressed as relative area change (RAC) decrease early in the course of PH and is a predictor of mortality in patients with PH.

AIM
To evaluate PA diameters and RAC in patients with PH based on CMR.

MATERIALS AND METHODS:
CMR was performed to 32 patients with confirmed PH on right heart catheterisation and 10 patients as control group without PH. The diameters of PA's – main PA (MPA), right PA (RPA) and left PA (LPA) were measured at CMR axial white blood images. PA's elasticity defined as relative cross-sectional area change (RAC) was assessed from CMR cine cardiac views. RAC was calculated during cardiac cycle by formula: (maxA – minA)/minA (maxA – maximal area, minA – minimal area) and expressed as a percentage.

RESULTS:
In comparison with control subjects PA's of patients were significantly more dilated (MPA diameter 34,26 ± 0,96 vs 26,20 ± 1,55 mm; RPA diameter 24,05 ± 0,72 vs 16,78 ± 1,24 mm; LPA diameter 23,62 ± 0,60 vs 17,21 ± 0,99 mm, $p<0,001$), more distended (RPA maxA 5,26 ± 0,25 vs 3,72 ± 0,33 cm^2, $p=0.002$; RPA minA 4,39 ± 0,24 vs 2,3 ± 0,26 cm^2, $p<0.001$; LPA maxA 4.93 ± 0,22 vs 3,48 ± 0,33 cm^2, $p=0.001$; LPA minA 4,21 ± 0,21 vs 2,19 ± 0,24 cm^2, $p<0.001$; MPA minA 7,32 ± 0,31 vs 4,92 ± 0,54 cm^2, $p=0.002$) and PA's RAC was smaller (RPA RAC 21,51 ± 2,37 vs 65,43 ± 4,71%; LPA RAC 18.81 ± 2,40 vs 62,74 ± 6.76%; MPA RAC 14,74 ± 2,01 vs 46,14 ± 6.26%, $p<0.001$). PA diameters were directly correlated with their maxA and minA and inversely correlated with their RAC (RPA: r=0.86; 0.9; –0.68, $p<0.001$; LPA: r=0.87; 0.9; –0.69, $p<0.001$, MPA: r=0.8; 0.8, –0.38, $p<0.05$). The linear regression analysis showed that PA's RAC could be predicted based on PA's diameters: model for RPA is $y=100.21-3,05x$ ($r^2=0.46$, $p<0.001$), for LPA is $y=123.12-4.29x$ ($r^2=0.47$, $p<0.001$) and for MPA is $y=56.02-1.08x$ ($r^2=0.15$, $p<0.05$).

CONCLUSIONS:
The PA's diameters, minimal and maximal cross-sectional areas increase and elasticity decrease in patients with PH. PA's diameter could be useful in predicting vascular elasticity.

CONNECTIVE TISSUE DISEASE ASSOCIATED PAH – RETROSPECTIVE STUDY OF REGISTRY OF LATVIA

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Key words. Pulmonary arterial hypertension, Connective tissue disease

Introduction. World Health Organization (WHO) PH classification define 5 main groups with distinct clinical and pathophysiological characteristics. Connective tissue disease associated pulmonary arterial hypertension (CTD-APHA) is within group I, signifies a severe complication of all form of connective tissue disease.

Aim. To identify characteristics and patterns among the population of CTD-APAH in Latvia. **Materials and methods.** We retrospectively analyzed data obtained from the Latvian national registry. All consecutively diagnosed CTD-APH cases (n=24) between February 2008-2015.

All underwent transthoracic echocardiography (TTE) showed right ventricular systolic pressure (RVSP) of ≥40mmHg. PAH was subsequently diagnosed right heart catheterization (RHC). Further evaluation; ECG, chest CT, brain natriuretic peptide (BNP) levels, 6 minute walk test (6MWT), pulmonary functional test.

Inclusion criteria; mean pulmonary arterial pressure (mPAP) of ≥25mmHg at rest, pulmonary capillary wedge pressure (PCWP) ≤15mmHg and pulmonary vascular resistance (PVR) >3 Wood units. Transpulmonary pressure gradient (TPG) ≥15 was applied in borderline (n=3) cases; "out of proportion".

Results. 4 patients were excluded due to lack of data. Population; n=20; 18♀ and 2♂. Mean age and standard deviation $\bar{x}$ = 58.5 ± 14yr. CTD distribution; 55% Systemic Scleroderma, 25% Systemic Lupus Erythematosus, 10% Mixed connective tissue disease and 10% others. At the diagnosis, 85% were in New York Heart Association functional class III or IV. BNP level $\bar{x}$ = 277 ± 211pg/mL (3<50, 12>180). 6MWT distance $\bar{x}$ = 238 ± 87m (4≤165).

mPAP $\bar{x}$ = 39.4 ± 10.6 mmHg. Cardiac index $\bar{x}$ = 2.9 ± 0.9 L/min/m^2. Cardiac output $\bar{x}$ = 4.6 ± 1.3 L/min. PVR $\bar{x}$ = 6.4 Wood units ± 3.67. Out of the 20 patients, only 1 had positive test 5% (CI 95% 0.009 - 0.236).

RVSP predictability of the estimated systolic pulmonary arterial pressure (sPAP); Pearson correlation (r= +.72, SE 4.7mmHg). $p < 0.01$.

1yr survival 85% (n=17) SE 8%. Mortality rate 0.11 deaths/1 person years (95%CI = 0.015-0.21).

Incidence proportion estimates 1.66 cases/Million adult inhabitants and estimates prevalence 12.04 cases/Million adult inhabitants.

Conclusion. In Latvia, the vast majority of CTD-APAH cases are diagnosed in the advanced stages. Earlier recognition and initiation of pathogenic treatment, may improve quality of life and reduce the morbidity. Thus, evaluation of high risk individuals, such as the connective tissue disease patients with available, reproducible tools like TTE and BNP should not be overlooked.

Further work is needed in order to establish the usefulness and cost effectiveness of early assessment among the population of connective tissue disease patients.

EISENMENGER SYNDROME AND RIGHT ATRIAL TUMOUR

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Keywords: Eisenmenger syndrome, pulmonary arterial hypertension, atrial tumour, sinus venosus atrial septal defect.

Introduction: In some cases of uncorrected sinus venosus atrial septal defects severe pulmonary arterial hypertension (PAH) – Eisenmenger syndrome (ES) – may develop. Surgical interventions in patients with ES are extremely high-risk. Surgical removal is recommended for mobile fast-growing atrial tumours.

Aim: We report a case in which two rare and potentially fatal diseases – ES and atrial tumour – are concurrent.

Case report: A 53-year-old male patient was referred to our hospital complaining of shortness of breath and fatigue in 2007. ES and sinus venosus atrial septal defect were diagnosed. Severe PAH was confirmed by right heart catheterisation in 2009. PAH target therapy with Sildenafil 20mg tid was started in 2012. The patent was followed up every 3 months in our pulmonary hypertension centre and improvement of symptoms and physical capacity (6-minute walk test distance increased by more than 70 meters) was observed. A right atrial mass was incidentally found in 2014 during routine echocardiography and confirmed by magnetic resonance imaging.

Right atrial tumour enlarged from 2×2 cm to 5×4 cm during the time period from February 2014 to November 2015. The patient was referred to a cardiac surgeon and high-risk life-saving surgery was recommended. However, the patient categorically refused surgical treatment.

Summary: We present a case report of two very rare and life-threatening diseases.

THE THERAPEUTIC EFFECT AND SHORT-TERM OUTCOME OF PULMONARY ENDARTERECTOMY IN ESTONIA: A NATIONAL DATABASE-DERIVED STUDY

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Key Words: Chronic Thrombembolic Pulmonary Hypertension, Pulmonary Endarterectomy.

Introduction. Pulmonary endarterectomy (PEA) is the treatment of choice in patients diagnosed as having chronic thrombembolic pulmonary hypertension (CTEPH). The outcomes of PEA in Estonia have never been systematically addressed before.

Patients and methods. The data on all patients with CTEPH recorded on the Estonian National database for Pulmonary Arterial Hypertension (PAH) from May 2011 to February 2016, who were deemed operable, were included. The diagnosis of CTEPH conformed to the ESC/ERS guidelines. All patients had undergone right heart catheterization. Survival estimates were calculated and one-month outcomes of PEA were assessed.

Results. Eleven patients (27.3% females) were included at 58.0 (median, IQR 31.0-70.0) years. Of the risk factors, 7 (63.6%) had a non-0 blood group, 5 (45.5%) had previous deep venous thrombosis, 4 (36.4%) had antiphospholipid antibody syndrome, and 2 (18.2%) had used oral hormonal contraceptives. Three patients (27.3%) received preoperative PAH-specific treatment.

PEA was performed in 10 patients, first in Vienna, Austria, in 3 occasions (30.0%, in 2011-2014), thereafter in 2 occasions (20.0%) in Helsinki, Finland (in January and in May 2015, and from September 2015 onward, in Tartu, Estonia (5 patients, 50.0%). One patient never underwent PEA because of inaccessibility of the method in Estonia before 2015 and severe deconditioning. Two patients (18.2%) died during the observation period. The mean survival estimate was 1447±177 days (mean±SEM) for all CTEPH patients and 1567±165 days for those, who underwent PEA. The survival rate was not different between those, who were operated on in abroad and those, who were operated on in Tartu (p=0.32). At 4.8 years of follow-up, it is estimated that 60.0% of our CTEPH patients are still alive.

PEA significantly reduced the WHO functional class from 3 to 1.5 (median) (p=0.015), esPAP) from 95.0 to 29.0 mmHg (p=0.028), mPAP from 54.0 to 32.5 mmHg (p=0.012), serum pro-BNP content from 3224 to 659 pg/mL (p=0.018), and serum creatinine content from 107.0 to 85.0 µmol/L (p=0.028) and increased significantly the 6MWD from 375.0 to 450.5 m (p=0.017) and cardiac index from 2.0 to 2.5 L/(min×m^2) (p=0.036). Reduction of the pulmonary vascular resistance from 11 to 4 mmHg, as well as the drops in serum contents of creatine kinase, urate, and troponin-T did not reach significance.

In 6 patients (60.0%), PEA was associated with complications: pericardial tamponade, atrial fibrillation, pneumonia, and wound complications occurred at 20.0% frequency, whereas hemothorax developed in one patient (10%). One patient died of multiorgan failure on her 4th postoperative day resulting in 20.0% perioperative mortality at our center during the first 6 months of the availability of PEA in Tartu.

Conclusions. This national PAH database-derived analysis represents the first study in Estonia that details CTEPH and indicates that with training and supervisional support during the learning age, CTEPH is acceptably manageable with PEA in a single county with a population below 1.5 million.
Summary: Accepting the learning curve, PEA is a feasible method even in a county with a population below 1.5 million.

INFLUENCE OF LEFT VENTRICULAR DIASTOLIC DYSFUNCTION TO THE DEVELOPMENT OF PULMONARY HYPERTENSION IN PATIENTS WITH SEVERE AORTIC STENOSIS

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Keywords

Pulmonary hypertension, diastolic dysfunction, left ventricular filling pressure, aortic stenosis

Introduction

The development of pulmonary hypertension (PH) in patients with severe aortic stenosis (AS) is associated with clinical deterioration and poor outcomes. Therefore, defining the causes of PH and the clinical markers associated with PH carries significant implications for patient management. Increased LV filling pressure is one of the factors influencing development of PH. The contribution of LV diastolic dysfunction (LVDD) as a determinant of PH in patients with severe AS is not well defined as a tool in clinical practice.

Aim

To evaluate the impact of Doppler-echocardiographic parameters of LVDD (increased LV filling pressure) as determinants of PH in patients with severe AS.

Materials and methods

We evaluated 52 patients, who underwent isolated aortic valve replacement, because of severe aortic stenosis (aortic valve area < 1 cm^2). Patients with documented coronary heart disease, chronic obstructive pulmonary disease, atrial fibrillation and mitral regurgitation were excluded. Pulmonary systolic pressure (PSP), left atrial (LA) volume and parameters of LV diastolic function (peak early (E) and late (A) diastolic velocities of mitral inflow, E/A ratio, transmitral flow velocity (E)/mitral annular diastolic velocity (E') ratio) were evaluated on preoperative echocardiographic protocol. Pulmonary hypertension was defined as an estimated PSP > 35 mm Hg. Statistical analysis was performed using SPSS version 21.0.

Results

We enrolled 52 patients, the mean age was 68,7 ± 7,42 years. There were 25 men (48,1 %) and 27 women (51,9 %). Among these patients, 35 (67,3%) had PH (mean systolic PAP was 43,17 ± 9,35 mmHg) and 17 (32,7%)- hadn't PH (mean systolic PAP was 31,29 ± 3,18 mmHg). Frequent of comorbidities (arterial hypertension, diabetes) were equal in both groups. Patients with PH had statistically significant worse LV diastolic function parameters, compared with group without PH (E/E' ratio- 14,95 ± 7,15 vs. 10,89 ± 3,09 (p=0,04); average of LA volume- 95,94 ± 32,28 vs. 80,06 ± 25,28ml (p=0,05), accordingly). This study have demonstrated a significant correlation between the level of PSP and E/A ratio (r=0,23, p=0,02), E/E' ratio (r=0,3, p=0,03), LA volume (r=0,32, p=0,01).

Conclusions

Increased left ventricular filling pressure and increased left atrial volume are cumulatively linked in the prediction of pulmonary hypertension in patients with severe aortic stenosis.

REASONS FOR DELAYED DIAGNOSIS OF PULMONARY ARTERIAL HYPERTENSION

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Key words: Pulmonary hypertension, pulmonary arterial hypertension

Introduction: Symptoms of pulmonary hypertension (PH) are non-specific, so PH and it's most severe forms – pulmonary arterial hypertension (PAH) and chronic thromboembolic pulmonary hypertension (CTEPH) – are often diagnosed at an advanced stage (World Health Organization functional class functional class (WHO-FC) III-IV). Early precise diagnosis and appropriate treatment is essential for improving symptoms and prolonging survival.

Aim: To find out the possible reasons of delayed diagnosis and treatment in our patients with PAH.

Materials and methods: 199 consecutive patients managed at our hospital's Pulmonary Hypertension Referral Center (PHc) till 2015.12 were found in the database. Their medical records were retrospectively reviewed and most common symptoms, details of referral to our center, time to right heart catheterization (RHC) and receiving precise diagnosis as well as starting on PAH target therapy were analyzed. Delayed diagnosis was considered: >1,5 years duration between onset of symptoms and referral to PHc and/or RHC was performed >6 months after first visit in PHc.

Results: The most common symptoms in our patients were: dyspnoe – 86,43%, fatigue – 33,17%, arrhythmia – 31,16%, angina pectoris – 25,63%, peripheral edema– 17,59%, syncope - 11%. Most of the patients (68%) were referred by cardiologists. PAH was diagnosed in 96, CTEPH in 55, mixed in 37, other forms of PH in 11 patients. The mean duration between onset of symptoms and referral to our PHc was >1,5 years (71,43% patients). 82,78% patients were in WHO-FC III-IV. RHC was performed at <1 month (64,29%) after first visit in PHc. (For 18 (64,29%) patients diagnosis was established during ≤1 month, 5 (17,86%) - 1-3 months, 3 (10,71%) - 3-6 months, 1 (3,57%) - 6-12 months, 1 (3,57%) - >12 months).
In PAH group the mean age during admission was 45,73 (SD=19,89 / 8-85) years and the most common form was PAH due to congenital heart disease. Target PAH therapy was started in 71 (73,96% of PAH patients) at mean ≤1 month (43,66% patients) after RHC. The most common reasons for delayed diagnosis and treatment in PAH were delayed referral to PHc and patient's refusal of RHC.
Conclusions: Most often patients were sent for the consultation to PHc overdue, in FC III-IV. In our PHc most of the patients had their diagnosis established in time. WHO- FC III-IV patients were examined faster.
Summary: Early precise diagnosis and appropriate treatment is essential to improving symptoms and prolonging survival in patients with PAH and CTEPH. Data of our PH center were retrospectively reviewed and the reasons of delayed diagnosis and treatment to our patients were analyzed in the study.

THE EFFECT OF SILDENAFIL IN PULMONARY HYPERTENSION DUE TO FAMILIAL RESTRICTIVE CARDIOMYOPATHY

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Key words: familial restrictive cardiomyopathy, sildenafil.

Introduction: Familial restrictive cardiomyopathy (RCM) is a very rare form of heart muscle disease that is characterized by restrictive filling of the ventricles and early onset of pulmonary hypertension (PH). The long-term prognosis for patients with RCM varies depending on the symptoms at the time of diagnosis and the presence of PH. Irreversible PH is the major risk factor associated with poor outcome.

Aim: To present oral sildenafil effect in management of PH in a patient with familial RCM.

Material, methods and results (A case report): A 25-year-old man was admitted to our hospital with symptoms of dyspnea, fatigue and signs of congestive heart failure. Familial RCM was diagnosed in childhood. His father died after heart transplantation at the age of 36 years. A diagnosis of mixed PH (precapillary and postcapillary) with very high pulmonary vascular resistance ((PVR) - 9,8 WU) and diastolic pressure gradient ((DPG) 17 mmHg) was confirmed by right heart catheterization (RHC). Other reasons for precapillary PH were not found. A cardiopulmonary exercise test revealed indication for heart and lung transplantation. Specific PAH therapy - oral sildenafil 20 mg twice a day was added to his heart failure (HF) treatment and patients condition improved. After 196 days he was hospitalized with a worsening of congestive HF. RHC was repeated and decreased PVR (5 WU) was found. The patient was placed on a waiting list for a heart transplantation. Left ventricular assist device (LVAD) was recommended as a bridge to a heart transplantation in case of worsening.

Conclusions: Sildenafil could be effective in reducing an elevated PVR in restrictive cardiomyopathy and might allow to select heart transplantation alone instead of heart and lung complex transplantation. Adequate admission of diuretics is important to avoid fluid retention and worsening of heart failure.

Summary: The presence of irreversible PH is a contraindication for a heart transplantation - the only known effective treatment for RCM. Oral Sildenafil could be effective in reducing pulmonary vascular resistance and could enable patients with high PVR to receive heart transplants.

VILNIUS EXPERIENCE OF TREATING PULMONARY ARTERIAL HYPERTENSION WITH TREPROSTINIL

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Key words: Pulmonary arterial hypertension, subcutaneous treprostinil.

Introduction: Pulmonary arterial hypertension (PAH) is a severe condition, characterized by progressive pulmonary arterial vasoconstriction and remodelation that causes right ventricular failure and poor survival. Continuous prostanoid infusion is recommended for patients with advanced PAH. Subcutaneous (SC) prostacyclin analogue treprostinil showed good results in reducing symptoms, increasing exercise capacity and improving cardiopulmonary hemodynamics.

Aim: The aim of this study was to present our centre experience in PAH treatment with SC treprostinil.

Materials and methods: 9 consecutive patients treated with SC treprostinil from October 2011 till March 2016 were identified from our hospital Pulmonary hypertension referral centre database. The patients medical records were retrospectively reviewed. Treatment efficiency was assessed after 1, 3 months and 1 year.

Results: The mean age of the patients was 56±18 years, all except 1 were adults (4 diagnosed with PAH associated with systemic connective tissue disease, 5 patients with idiopathic PAH). SC treprostinil was started as a part of complex PAH therapy, when treatment with 2 oral medications (sildenafil and endothelin receptor antagonist and /or inhaled iloprost) was inadequate. 4 of the patients felt pain at the site of the infusion, for 3 of them pain was controlled by analgesics and non-pharmacological methods. SC treprostinil was discontinued for 2 patients due to severe infusion site pain after 7 and 9 days of treatment. For the rest of the patients mean treprostinil dose was 15.6±5.9 ng/ml/min. The mean duration of treatment was 179±214 days. 4 World Health Organization functional class (WHO-FK) IV patients died after 10.4±8.9 months. 5 of the patients improved clinically temporarily, 6-minute walk test (6-MWT) distance improved in 3 patients, B-type natriuretic peptide (BNP) level significantly decreased in 5 patients ($p<0.05$) after first month of the treatment.

Conclusions: SC treprostinil could be an efficacious PAH treatment option even in WHO-FK IV patients, resulting in improved clinical condition and may prolong survival.

Summary: Our experience with first 9 patients with SC treprostinil from October 2011 till March 2016 was analysed. SC treprostinil could be effective in decreasing symptoms and improving survival even for WHO-FK IV patients. Infusion site pain might limit the treatment.