

2nd BALTIC & 3rd LITHUANIAN

PULMONARY HYPERTENSION C O N F E R E N C E

An aerial photograph of Vilnius, Lithuania, showing a dense urban landscape with historic buildings, red-tiled roofs, and a river. The image is partially obscured by a large white circular graphic element that frames the text.

**ONLINE
ABSTRACT
BOOK**

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ONLINE ABSTRACT BOOK

The content of the abstracts presented is the responsibility of their authors and co-authors.
The abstracts are arranged in sequence according to the conference moderated session programme.

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MODERATED POSTER SESSION ABSTRACTS

I. Survival and complications in patients with pulmonary hypertension long-term after correction of congenital heart defects

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Introduction

Shunt related pulmonary hypertension (PH) could develop in patients with congenital heart disease (CHD) (1). For some patients pulmonary arterial hypertension (PAH) persists despite early surgical correction of CHD (2). In that case they have worse outcomes compared to other clinical subgroups of PAH related to CHD and may suffer from lifelong complications, such as; residual lesions leading to re-interventions, arrhythmias, heart failure and thromboembolisms (3–5).

Aims and Objectives

The aim of our study was to assess long-term outcomes of PH after surgical correction of congenital systemic-to-pulmonary shunts.

Materials and methods

In this retrospective cohort study, we aimed to identify all patients with CHD who underwent a cardiac catheterization in the Vilnius University Hospital Santaros klinikos during 1985 – 2007. Only patients with PH (mean pulmonary artery pressure ≥ 25 mmHg) were selected. For the final long-term outcome evaluation, patients who underwent surgical correction of CHD and had regular follow-up data were included.

Results

Of the 4118 cardiac catheterizations 388 paediatric patients were identified with CHD and PH. Surgical correction of CHD was performed in 160 patients of which 88 patients were finally included with available follow-up data. Median age at CHD and PH diagnosis was 0.8 [0.6 - 3.0] years and at CHD surgery 1.1 [0.6 - 3.9] years (50% females). Residual PH was observed in 30.7% (n=27). After a median follow-up of 21 [15 – 24] years, 9.1% of the patients died. Kaplan-Meier survival analysis revealed significantly higher mortality in the residual PH group (p=0.035). Defect recanalization was observed in 27.3% (n=24) of the patients and subsequently almost all of them 87.5% (n=21) underwent repeated surgery. Only two patients (2.3%) had thromboembolic events, both in the non-residual PH patients' group. Arrhythmic complications presented in 14.8% (n=13) of individuals, in most cases supraventricular arrhythmia were registered (61.5%, n=8). Eight patients (9.1%) underwent pacemaker implantation. There was no significant difference related to complications during follow-up between residual and non-residual PH groups.

Conclusions

In almost one third of the patients PH persisted after surgical correction. Residual PH was associated with higher mortality in patients with corrected CHD. Complication rate during long-term follow-up was similar in patients with persistent PH and without PH in our cohort.

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II. Evaluation of functional class, quality of life and presence of Eisenmenger syndrome depending on congenital heart disease type in pulmonary arterial hypertension patients

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Introduction

Pulmonary hypertension (PH) is a debilitating and life-threatening disease which can occur as a result of various medical conditions. PH is classified in 5 groups. Group 1- pulmonary arterial hypertension (PAH) is one of the most studied forms of PH (Sean H. Oldroyd et al, 2018). Approximately 3 to 10 percent of adults with congenital heart disease develop PAH (PAH-CHD). Early diagnosis and repair of CHD has decreased the number of adults with CHD, however, the overall number of PAH-CHD patients is increasing as more patients with complex and palliated CHD survive to adulthood (Heidi M. Connolly et al, 2017). Progression of pulmonary arteriopathy in PAH-CHD patients can result in the reversal of the intracardiac shunt manifesting as the most advanced form of PAH, known as Eisenmenger syndrome (ES).

Aims and Objectives

The aim of the study was to evaluate functional class, quality of life and presence of ES depending on CHD type in PAH-CHD patients.

Materials and methods

During a time period from 17.07. - 27.08.2018 adult patients with confirmed PAH-CHD were interviewed at Pauls Stradiņš Clinical University Hospital regarding their quality of life (EQ 5-D scale was used, scored 0 (poorest possible health) to 100 (best possible health)). A 6 minute walking test (6MWT) was performed to evaluate NYHA functional class (FC). CHD type and presence of ES were obtained from in patients' medical records. Statistical analysis was performed using IBM SPSS 22.0 and results were considered significant if $p < 0.05$.

Results

Twenty-one PAH-CHD patients were included in present study- 67% were female (N= 14), 33% were male (N= 7). 52% (N= 11) of patients had atrial septal defect (ASD), 38% (N= 8) had ventricular septal defect (VSD) and 10% (N= 2) had atrioventricular septal defect (AVSD). 19% (N= 4) of patients had corrected CHD- three of them had ASD, one patient had VSD. The mean age in ASD, VSD and AVSD groups was 69 ± 13 , 45 ± 13 and 37 ± 1 years, respectively. The mean pulmonary artery pressure (mPAP) in these groups was 41 ± 9 , 79 ± 25 and 73 ± 5 mmHg, respectively.

When analysing patients who were able to perform 6MWT (N= 18), of eight patients with ASD 12% were FC I (N= 1), 25% were FC II (N= 2) and 63% were FC III (N= 5). Among eight patients with VSD 37% were FC II (N= 3) and 63% were FC III (N= 5). Two remaining patients with AVSD were FC III.

33% (N= 7) of all patients had progressed to ES. ES had developed in 9% (N= 1) of patients with ASD, 63% (N= 5) of patients with VSD, and in 50% (N= 1) of patients with AVSD. Among patients with ASD average quality of life score was 65, with VSD- 57 and among those with AVSD- 45.

Conclusions

There were no significant differences in patient distribution by FC among patients with ASD, nor among those with VSD. However, there was a significant correlation between presence of ES and CHD type (Fisher's Exact Test $p = 0.036$) - ES was more common in patients having VSD compared with patients having ASD. Patients with ASD had significantly lower mean mPAP compared to VSD patient group. There were no significant differences in mean mPAP values between ASD and AVSD groups, probably due to the small sample size. In addition, there were no significant differences between quality of life among patients with ASD and VSD.

III. The right ventricle under pressure: mechanics and shape changes in pulmonary hypertension. A 3-dimensional echocardiography study

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Background

Right ventricular (RV) adapts to pressure overload (PO) by changing size, function and shape. To date, alterations in interventricular septum shape have been studied more extensively than shape changes occurring in other regions of the RV in PO. Moreover, despite RV shows a quite complex mechanics, conventional echocardiography usually explores mainly its longitudinal function only. Therefore, the relative contribution of the different components of RV wall displacement (e.g. longitudinal and radial) to global RV ejection fraction (EF) in PO conditions remains to be clarified.

Purpose

To evaluate RV shape and mechanics changes in patients (pts) with PO.

Methods

We obtained 3D transthoracic echocardiography from 31 pts with pre-capillary pulmonary hypertension (pPH) (mean age 57±14 years, 77% women, mean systolic pulmonary artery pressure (PAP) 57.7±15.2 mmHg,) and 30 age and gender matched healthy volunteers. Dedicated 3D full-volume data sets of the RV were obtained using multibeam acquisitions. RVEF was measured using commercially available dedicated software packages. The longitudinal (LEF) and radial (REF) displacements of the RV walls and their relative contribution to global RV ejection fraction (LEF/GEF and REF/GEF) were obtained from 3DE data sets using custom software. For shape analysis RV surfaces were reconstructed at end-diastole (ED) and end-systole (ES) and the RV was segmented in 4 parts: inflow (RVIT) and outflow tracts (RVOT), apex and body (both divided into free wall and septum) and their curvatures were calculated using custom software. Zero curvature defines a flat surface, whereas positive or negative curvature indicates convexity or concavity, respectively.

Results

pPH pts showed significantly lower 3D RVEF and larger 3DRV volumes comparing with normals (Table 1). Comparing with control group, pressure overloaded RV showed flatter RV free wall and RVIT during all cardiac cycle (Table 2). Also, pPH pts had more convex (rounder) IVS (especially in the apex) during all cardiac cycle and RVOT at ES than healthy volunteers (Table 2). Moreover, PO group demonstrated significantly lower REF/GEF than control group, but similar LEF/GEF (Table 1).

Conclusions

Chronic PO leads to RV dilatation and function deterioration in pPH. Under pressure, the septum becomes more convex and bulges more into the left side during all cardiac cycle (especially at ES) with a more flattened free wall and RVIT comparing with healthy volunteers. Also, the increased afterload in pPH leads to remodeling of the RVOT into a more circular shape. Finally, in PO the relative contribution of the radial RV wall displacement to global RVEF is impaired more than its longitudinal shortening.

Clarifying RV mechanical and shape adaptations to pressure overload may improve our understanding of the mechanisms leading to RV failure in pPH.

Table 1. Demographics, right ventricular function and mechanics in pPH and in controls

RV parameters	pPH (N=31)	Control group (N=30)
Age (years)	57±14	57±11
Women (%)	77	73
Systolic PAP (mmHg)	58±15**	20±4
End-diastolic volume (ml/m ²)	95±22**	58±13
End-systolic volume (ml/m ²)	61±20**	24±6
Ejection fraction (%)	36±8**	59±7
LEF/GEF	51±12	46±7
REF/GEF	35±11**	48±7

*p<0.05, **p<0.0001

Table 2. Right ventricular volumes and curvature in pressure overload and in control group

Parameters	End-diastole		End-systole	
	pPH (n=31)	Control group (n=30)	pPH (n=31)	Control group (n=30)
Volume index (ml/m ²)				
Total volume	95±23	66±14**	62±21	31±8**
Apex	15±4	10±2**	9±3	4±1**
Body	36±9	25±5**	24±8	12±3**
RVIT	27±6	18±4**	17±6	9±2**
RVOT	18±4	12±3**	12±4	6±1**
Curvature (indexed to volume)				
RVIT	1.19±0.1	1.32±0.3**	1.16±0.1	1.25±0.1**
RVOT	1.49±0.1	1.40±0.1	1.43±0.1	1.31±0.1**
General FW	1.36±0.05	1.42±0.09**	1.29±0.04	1.35±0.06**
Free wall body	1.09±0.1	1.16±0.1**	1.01±0.1	1.09±0.1**
Free wall apex	2.02±0.3	2.34±0.16**	1.85±0.2	2.29±0.2**
General septum	0.70±0.11	0.59±0.08**	0.76±0.12	0.58±0.1**
Septal body	0.32±0.1	0.18±0.1**	0.46±0.1	0.2±0.1**
Septal apex	0.98±0.3	0.38±0.2**	1.19±0.5	0.46±0.3**

*p<0.05, **p<0.001

IV. A complex clinical case of pulmonary hypertension due to lung diseases

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Introduction and aim

The shortness of breath (dyspnea) is the main symptom of pulmonary hypertension (PH). Severe PH due to lung disease is uncommon. The etiology of dyspnea is multifactorial in about one-third of patients. In some patients it might be difficult to determine whether PH is due to lung disease or whether the patient suffers from two diseases (pulmonary arterial hypertension and chronic lung disease). The aim of this presentation is to introduce a case of PH due to lung diseases with complex identification of diagnosis.

Case report

A 61 years-old man complaining of progressing dyspnea since 2012 and was referred to our center in 2013. From anamnesis, it was known that he was former smoker and had acute infection of cytomegalovirus in 1999. Right empyemectomy and decortication was performed due to right side pneumonia and pleural empyema in 2005, following with pleurostomy of the right lung. After two years left side pneumonia with haemothorax developed. Pericardial calcification was found in 2008 and partial pericardectomy was performed. Since 2011 he was anticoagulated and treated with amiodarone due to paroxysmal atrial fibrillation.

Investigation according to PH algorithm was performed in April 2013. Hypoxemia was found in arterial blood gases test. Pulmonary function test (PFT) showed reduced lung volumes (TLC 2.4 L (33%), VC 1.68 L (36%)) and diffusing capacity (DLCO 34%) with no signs of obstruction. Echocardiography revealed dilatation of the right heart and signs of PH (systolic pulmonary artery pressure (PAP) 52 mmHg). Computed tomography (CT) and CT pulmonary angiography showed no signs of pulmonary thromboembolism, but pneumofibrosis was evident. Precapillary PH (mean PAP 33 mmHg, mean pulmonary arterial wedge pressure (PAWP) 14 mmHg) with slightly elevated pulmonary vascular resistance (PVR 3.39 WU) and positive acute vasoreactivity test were found during right heart catheterization (RHC). Therefore home oxygen therapy and high dose of calcium channel blocker (diltiazem) were additionally prescribed.

After one month the patient presented with severe peripheral edema and fluid retention in the left pleural cavity. Diltiazem was discontinued and repeated pleural punctures were performed. In 2015 he underwent thoracoplasty and myoplasty due to chest pain and defect of the right chest wall. Progressive deterioration: severe dyspnoea, very low exercise capacity, signs of heart failure and further decrease of DLCO (28%) were found in 2016. RHC and coronary angiogram showed a slight decline in PAP (mean PAP 27 mmHg, mean PCW 5 mmHg, PVR 3, 38 WU) and no stenosis in the coronary arteries. In April 2016, after multidisciplinary team discussion low dose of sildenafil (10 mg 3 times per day) was prescribed, but discontinued after two weeks by the cause of severe drop of arterial saturation despite oxygen therapy. Patient was referred for lung transplantation. In 2017 due to progression of dyspnea, recurrent hypercapnia and hypoxemia in arterial blood gas tests (pCO₂ 77,1 mmHg, pO₂ 54,8 mmHg) and after exclusion of neuromuscular diseases, noninvasive night time ventilation bi-level regimen simultaneously with oxygen therapy was prescribed due to hypoventilation syndrome. Slight improvement was achieved (pCO₂ 77,1->57,9, pO₂ 54,8->90,6). Afterwards, the patient was re-hospitalized several times for correction of noninvasive ventilation and oxygen therapy parameters.

Discussion

Well known that restrictive pulmonary diseases and sleep disorders could lead to PH. However some patients could have several diseases and overlapping representation. In such cases results of diagnostic tests could be misleading and could be difficult to establish accurate diagnosis and prescribe appropriate treatment.

Conclusions

Careful examination of the patient is necessary to establish the accurate diagnosis. Supportive treatment with noninvasive ventilation and long-term oxygen therapy remains the main option of treatment for patients with PH due to restrictive pulmonary diseases and sleep disorders.

V. Portopulmonary hypertension in a Patient with Liver Cirrhosis

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Introduction and aim

According to European Society of Cardiology (ESC) guidelines, Portopulmonary hypertension (PoPH) is defined as Pulmonary arterial hypertension (PAH) associated with Portal hypertension with or without the presence of liver disease. However PoPH is one of the most common complications associated with liver cirrhosis and remains one of the biggest challenges in patients undergoing liver transplantation.

The diagnosis of PoPH is made by:

1) Right Heart Catheterization (RHC): mean Pulmonary arterial pressure (mPAP): mild >25 mmHg, moderate > 35-46 mmHg, severe >45 mmHg, Pulmonary capillary wedge pressure (PCWP) ≤15 mmHg and Pulmonary Vascular Resistance (PVR) >3 Wood Unit.

2) Hepatic venous pressure gradient (HVPG): mild Portal hypertension (HVPG > 5 but < 10 mm Hg) and clinically significant Portal hypertension HVPG ≥10 mm Hg.

We report a case of a patient with liver cirrhosis and severe PoPH undergoing vasodilator therapy with sildenafil and treprostinil prior to liver transplantation.

Case report

Our patient – female, born in 1968, diagnosed with Hepatitis C in 2000. The treatment of hepatitis C with interferons was stopped due to severe side effects. In 2009 liver cirrhosis associated with Portal hypertension was diagnosed, and due to the oesophageal bleeding, TIPS (Transjugular Intrahepatic Portosystemic Shunt) was performed.

In 2015, due to the progression of symptoms of hepatic insufficiency, the patient was referred to Tartu University Hospital for consultation for liver transplantation (not possible in Latvia in 2015). The echocardiography done in Tartu University Hospital showed dilation of the right heart, right ventricular overload and elevated right ventricular systolic pressure (70 mmHg) with normal left side of the heart. Due to suspicion of Pulmonary hypertension patient was sent back to Riga for further examinations.

The patient was admitted to the Cardiology department of Pauls Stradins Clinical University Hospital for the 1st time on the 22.11.2015. The patient presented with: dyspnoea on exertion, palpitations, fatigue. BP: 116/44. ECG: SR, 74 bpm. Biochemistry: ALAT: 248 U/L, ASAT: 323 U/L, Total bilirubin: 51 µmol, Creatinine: 58 µmol, Glucose: 3.7 mmol/L. On 24.11.2015 the 1st RHC was performed: mPAP – 70 mmHg, PCWP – 15 and PVR 11 Wood units, HVP: 15 mmHg. The diagnosis of Portopulmonary hypertension was confirmed and the patient was started on therapy with sildenafil 20 mg x3 PO, propranolol 10 mg x2 PO, spironolactone 25 mg x1 PO in the morning. On 03.01.2016 patient visit hospital for control: BP: 114/43. ECG: SR, 65 bpm. Biochemistry: ALAT: 213 U/L, ASAT: 333 U/L, Total bilirubin: 46 µmol, Creatinine: 54 µmol, Glucose: 5.4 mmol/L. 2nd RHC: mPAP – 69 mmHg, PCWP – 15 mmHg, PVR 9.5 Wood units. Propranolol was discontinued. On 19.01.2016 treprostinil 20 ng/kg/min s/c was added. Echocardiography on the 29.01.2016: TAPSE: 26 mm, RV diameter 47 mm, EF (Simpson) 60%. The patient underwent 3rd RHC on the 24.02.2016: mPAP – 62 mmHg, PCWP – 15 mmHg, PVR 4.7 Wood units. This therapy was maintained and on the 16.05.2016 the last RHC was performed: mPAP – 54 mmHg, PVR 4.5 Wood units.

Patient was sent home and continued therapy but died on the 01.02.2017 due to progressive right heart failure without achieving the optimal values for liver transplantation.

Discussion

Approximately 5-8% of candidates to liver transplantation in United States have PoPH. Although time of diagnosis is a major criteria many patients lack proper diagnosis of PoPH until later stages of liver disease. Without medical treatment and liver transplantation the survival of patients with PoPH is very low. According to the European Respiratory Society Task Force, patients with mild PoPH can safely undergo liver transplantation, patients with mPAP of 35-45 mmHg should receive vasodilator therapy preoperatively, but in patients with mPAP > 45 mmHg the liver transplantation is contraindicated.

According to the ESC guidelines, phosphodiesterase-5 inhibitors, soluble guanylate cyclase stimulators and prostacyclin analogues may be used in the treatment of PoPH.

In our case we started with sildenafil, a drug used extensively in the treatment of PAH. Sildenafil is a safe and well-tolerated therapy in PoPH with possible clinical, functional and haemodynamic improvements after 3 months and stable response over one year either as monotherapy or in combination with prostanoids. In our case we used treprostinil together with sildenafil and achieved the decrease of mPAP from 69 to 54 mmHg in approximately 4 months of therapy.

Beta-blockers that are usually used for the treatment of Portal hypertension tend to worsen haemodynamics in patients with PoPH and therefore propranolol was discontinued.

Conclusions

Most patients with PoPH are not included in therapeutical studies that have been done and therefore the adequate therapy is yet to be discovered. Although we did all recommended steps from the European guidelines with our patient, they were insufficient in providing an effective way for the treatment of PoPH. The failure in lowering the mPAP and providing the opportunity to our patient to undergo liver transplantation showed that is crucial to improve our knowledge in this field in order to increase life expectancy for patients diagnosed with PoPH.

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VI. Pulmonary hypertension in patients with hematological disorders

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Introduction

Pulmonary hypertension (PH), which defined as an elevated mean pulmonary artery pressure ≥ 25 mmHg at rest, could be a major complication of several haematological disorders. Haematological disorders, such as chronic myeloproliferative disease (CMPDs) associated with PH are included in group five of the most recent clinical classification, corresponding to PH with an unclear and/or multifactorial aetiology [1]. Three major distinct forms of PH have been described in patients with CMPDs: chronic thromboembolic PH (CTEPH), pulmonary arterial hypertension (PAH) associated with CMPDs without other risk factors of PAH and drug-induced PAH [2,3]. Pulmonary veno-occlusive disease (PVOD) can be an uncommon cause of PH as well. CTEPH is the most common pulmonary complication in patients with haematological disorders [4].

Aims and Objectives

To analyse our institutional experience in PH and haematological disorders.

Materials and methods

A retrospective analysis of the 400 patients, managed in Pulmonary Hypertension centre of Vilnius University Hospital Santaros Klinikos, during 2010-2018 year, was performed. Patients with PH and myeloproliferative disease were selected. The demographic and clinical data (gender, age, hematological disorder, right heart catheterization data and etc.) were analyzed.

Results

Overall, ten patients (2,5%) were identified with PH and myeloproliferative disease and were included to review. Male and female ratio was 1:1 (aged 52 ± 17 years). During median 2.4 (± 0.5) years follow up period, three (30%) patients died. Overall, 90 % (n=9) of the patients were in NYHA functional class more than II. CTEPH was diagnosed in 4 patients (three patients with essential thrombocythemia and one with chronic myelogenous leukaemia). Four patients were in clinical PH group 5 (one with polycythaemia vera, one with acute myelogenous leukaemia and two with monoclonal gammopathy). For one patient, combined PH (PH group 5 due to polycythaemia vera and PH due to left heart disease) was confirmed. The last one patient, 34 years old female was diagnosed with chronic myelogenous leukaemia and recurrent drug induced PAH, treated with tyrosine kinase inhibitors.

On the whole, brain natriuretic peptide levels were increased in 80 % of patients: 749.4 (\pm 423.7 ng/l). Right heart catheterization was performed in 80 % of patients. The mean pulmonary artery pressure was 48 (\pm 9.8) mmHg, pulmonary vascular resistance – 8.8 (\pm 7.7 WU), pulmonary artery wedge pressure – 11.9 (\pm 5.3) mmHg, cardiac output – 5.3 (\pm 2.1) l/min and cardiac index – 3.0 (\pm 1.3) l/min/m². Half of the patients, were on advanced therapy.

Conclusions

PH associated with CMPDs is a rare heterogeneous group of diseases with different hemodynamic and clinical presentation. The large multicentre prospective studies to determine the prevalence, pathological mechanisms, causes and therapies options of haematological disorders and PH, is essential.

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VII. Impaired left ventricular performance as a prognostic marker in patients with precapillary pulmonary hypertension: cardiac magnetic resonance feature tracking study

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Introduction

Pulmonary hypertension (PH) causes right ventricle (RV) dysfunction. Cardiac magnetic resonance (CMR) derived RV ejection fraction (EF) has prognostic value in this condition. However, left ventricle (LV) function and mechanics could be affected in patients with RV failure as expression of ventricular interdependence. Despite the fact that the prognostic merit of specific imaging findings is still far from robust, conventional and the novel CMR technology – feature tracking (FT) LV parameters - could be an important additional prognostic value for survival outcomes.

Aims and Objectives

The aim of this study was to evaluate conventional CMR and FT parameters of LV mechanics in patients with precapillary PH (pPH) and to define the prognostic markers for adverse clinical outcomes.

Materials and methods

CMR cine imaging data of 43 patients with confirmed pPH diagnosis on right heart catheterization (mean pulmonary artery pressure (mPAP) 55.91 \pm 15.87mmHg, pulmonary capillary wedge pressure <15 mmHg) were retrospectively analyzed. Patients were divided in two groups according to survival outcomes (survival and non-survival) during the follow-up and treatment period. Patients with documented coronary and valvular heart disease also with atrial fibrillation were excluded from the study. LV and RV EF, indices of RV end diastolic and end systolic volumes (EDVi and ESVi) were evaluated using conventional CMR software (syngo.via; Siemens Healthcare). LV global longitudinal and circumferential strains (LV-LS, LV-CS) were assessed in long (two, three, four chambers) and short axes (basal, mid and apical) views using CMR FT software package (Medis Suite QStrain 2.0; Medis Medical Imaging Systems bv). The average of global LS and CS were calculated. Independent Samples Mann-Whitney U test was used to compare the distribution across categories and Spearman's correlation was performed.

Results

The distribution of age and gender ($p>0.05$) as well as mPAP (56.51 ± 16.27 mmHg vs 54.45 ± 15.6 mmHg, respectively $p=0.818$) did not differ in survival and non-survival patient groups. RV EF was lower (32.17 ± 12.85 vs $41.48\pm12.01\%$, respectively $p=0.088$) and RV ESVi was increased (68.58 ± 27.25 vs 51.32 ± 21.42 ml/m², respectively $p=0.05$) in non-survival group compared to survival patients. LV EF was decreased in non-survival group (48.50 ± 14.05 vs $57.90 \pm 9.64\%$, $p=0.042$). The global LV-LS was reduced in non-survival pPH group compared to survival group (-13.27 ± 6.34 vs -18.90 ± 4.67 , respectively $p=0.009$).

Decreased RV EF and increased RV ESVi significantly correlated with reduced global LV longitudinal (LV-LS) ($r=-0.544$, $p<0.05$, $r=-0.534$, $p<0.05$) and circumferential strain (LV-CS) parameters ($r=0.506$, $p<0.05$, $r=0.558$, $p<0.05$), also with reduced LV EF ($r=0.532$, $p<0.05$ and $r=-0.553$, $p<0.05$, respectively).

Univariable analysis using the Cox model showed that severely reduced global LV-LS $>-14.2\%$, with good sensitivity (66.7 %) and high specificity (93.5 %) indicated an increase of the risk of death by 11-fold; increased RVESVI >60 ml/m², despite its limited sensitivity (58.3%), but good specificity (77.4%) indicated an increase of the risk of death by 3-fold; decreased RVEF $<25.5\%$ and LVEF $<52\%$ with sensitivity (41.7% and 66.7 %) and good specificity (92.3% and 71%) indicated an increase of the risk of death by 3.5 and 3.3-fold, respectively.

Conclusions

Combination of CMR and the novel CMR technology FT allows more accurate evaluation of left ventricle mechanics and function that could help to predict the adverse clinical outcome in patients with precapillary PH.

VIII. Effects of Exercise and Respiratory Training on Exercise Capacity and Quality of Life in Precapillary Pulmonary Hypertension: primary results of A Prospective, Randomized Controlled Study in Lithuania

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Introduction

Pulmonary hypertension (PH) is a rare, progressive and devastating disease, which leads to reduced patient's physical activity and a poor quality of life (QOL). Different studies revealed, that a supervised rehabilitation program in specialized centers can lead to improvement in patient's QOL and exercise capacity. However, the effect of exercise and respiratory training (ET) in PH patients has never been examined in Lithuania.

Aims and Objectives

To assess efficacy of exercise and respiratory training as add-on to medical therapy in patients with precapillary pulmonary hypertension.

Materials and methods

Stable patients with invasively confirmed precapillary PH were randomly assigned to training and control groups. Training group patients received ET in-hospital for 10-21 days and continued at home for 12 weeks. Control group patients continued their daily lifestyle. Efficacy parameters have been evaluated at baseline and after 15 weeks. Exercise capacity was measured by six-minute walk test (6MWT) and cardiopulmonary exercise test (CPET). QOL was evaluated by 36-item short form survey (SF-36).

Results

Twelve consecutive patients were included in the study from February 2017 to June 2018. Control group consisted of 5 patients, while training group consisted of 7 patients. Demographic data between training and control groups, accordingly: age (54.00 ± 15 vs 55.60 ± 11), female predominance of gender (100% vs 71.42%). Patients in training group improved the mean distance in 6MWT (change, $+18.17 \pm 29.06$ vs -2.20 ± 10.5 meters, $p=0.174$). QOL, measured by SF36, became higher in training group than in control group, respectively: physical summary score (change, $+19.14 \pm 18.07$ vs $+4.25 \pm 19.23$, $p=0.200$), mental summary score (change, $+11.93 \pm 15.48$ vs -0.048 ± 17.69 , $p=0.241$). There were no major improvements in CPET parameters in both groups. The results are summarized in the table **Table 1**.

Conclusions

The primary data of our study showed an increase in physical exercise capacity parameters, measured by 6 minute walking test, and health-related quality of life assessment. Cardiopulmonary exercise test did not show an improvement in physical exercise capacity parameters 15 weeks after exercise and respiratory training.

		Training group (N=7)			Control group (N=5)			P value
		Baseline	After 15 weeks	Change	Baseline	After 15 weeks	Change	
6MWT	Distance (m)	458 ± 51	477.5 ± 68	+18.2 ± 29.1	378 ± 147	375.8 ± 147	-2.2 ± 10.5	0.174
	SPO ₂ after the test (%)	87.4 ± 9	89.8 ± 9	+2.0 ± 10.2	84.8 ± 13	82.2 ± 18	-2.6 ± 5.7	0.396
SF-36	Physical functioning	45 ± 12.6	57.9 ± 17.3	+12.6 ± 15.5	42 ± 16.8	40 ± 18.4	-2.0 ± 13.0	0.112
	Physical role functioning	14.3 ± 24.4	50 ± 38.2	+35.7 ± 34.9	25 ± 35.4	40 ± 28.5	+15 ± 57.5	0.453
	Bodily pain	62.6 ± 21.5	80 ± 25.3	+17.1 ± 23.3	54 ± 9.5	52 ± 20.2	-2.0 ± 16.5	0.149
	General health perceptions	39.3 ± 13.7	50.7 ± 21.3	+11.4 ± 16.5	45 ± 11.2	51 ± 9.6	-2.0 ± 16.5	0.527
	Vitality	60 ± 7.1	62.1 ± 8.1	+2.1 ± 4.9	53 ± 2.7	51 ± 6.5	-2.0 ± 8.4	0.307
	Social functioning	73.2 ± 28.4	78.6 ± 25.7	+5.4 ± 12.2	55 ± 28.8	60 ± 28.5	+5.0 ± 18.96	0.969
	Emotional role functioning	33.3 ± 47.1	66.6 ± 43.1	+33.3 ± 47.2	46.7 ± 50.6	46.7 ± 44.7	+0.008 ± 62.4	0.315
	Mental health	69.1 ± 8.2	76 ± 11.6	+6.9 ± 10.8	64.8 ± 15.9	61.6 ± 13.7	-3.2 ± 13.7	0.183
	Physical summary score	40.5 ± 12.9	59.6 ± 20.9	+19.1 ± 18.1	41.5 ± 11.5	45.8 ± 11.6	+4.3 ± 19.2	0.200
	Mental summary score	58.9 ± 17.9	70.9 ± 17.3	+11.9 ± 15.5	54.9 ± 21.3	54.8 ± 17.5	-0.048 ± 17.7	0.241
CPET	Workload max (W)	75.4 ± 12.5	76 ± 13.2	+0.57 ± 8.1	62.2 ± 12.4	66.4 ± 9.8	+4.2 ± 5.1	0.400
	Peak VO ₂ /kg (ml/kg/min)	14.4 ± 2.8	14.6 ± 2.2	+0.21 ± 0.7	11.5 ± 2.6	12.2 ± 2.8	+0.74 ± 1.6	0.615
	Peak VO ₂ (%)	56.3 ± 14.8	58 ± 18.8	+1.71 ± 7.2	49.4 ± 14.7	51.4 ± 13.7	+2.0 ± 7.5	0.948
	METS	4.1 ± 0.8	4.3 ± 0.8	+0.23 ± 0.8	3.3 ± 0.7	3.5 ± 0.8	+0.2 ± 0.5	0.943

IX. Pulmonary hypertension in patient with end stage pulmonary sarcoidosis: challenges in treatment

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Introduction and aim

Pulmonary hypertension (PH) is recognised as a serious complication of pulmonary sarcoidosis. The frequency of PH largely depends on the severity of the disease and is associated with poor outcomes. Current sarcoidosis associated pulmonary hypertension (SAPH) treatment recommendations suggest optimisation of underlying lung disease and use of oxygen when hypoxemia is present, however, there is still insufficient evidence to recommend advanced therapies.

Here we present challenges in treatment of a woman with end stage pulmonary sarcoidosis and severe SAPH to demonstrate the effect of oral vasodilator therapy while on the list for urgent lung transplantation.

Case report

A 40-year-old lady with history of sarcoidosis presented to our clinic due to worsening respiratory insufficiency. Physical exam was notable for cachexia, tachypnoe, cyanosis, peripheral edema, and ascites. On presentation the patient was put on supplemental oxygen and diuretic therapy. Due to severe dyspnoe 6 minute walk test could not be performed. Following a thoracic CT scan, transthoracic echocardiography, and right heart catheterization (Table 1 A,B) she was determined to have stage IV sarcoidosis with severe pulmonary fibrosis and cystic degeneration, SAPH, and associated right ventricular failure. Due to poor functional status (NYHA IV stage), severe pulmonary fibrosis, as well as severe PH, she was determined to have a bad prognosis according to available scientific data. The patient was included to the Urgent Lung Transplant list and Sildenafil was started increasing dosage from 10 to 20 mg x 3 times a day alongside optimal pharmaceutical heart failure treatment.

The clinical improvement was noticed immediately after the initiation of specific PH therapy and has remained stable for a year. 2D echocardiographic parameters didn't change significantly. The patient is doing better from a clinical stand-point, the objective parameters are listed in the table below (Table 2).

Discussion

PH is a known complication of sarcoidosis and is associated with increased mortality (1,2). With the advent of specific therapies for PH, there has been a resurgence of interest in the pathophysiology, diagnosis, and treatment of SAPH. SAPH may be the consequence of complex underlying mechanisms including granulomatous fibrosis, sarcoid-induced occlusive venopathy and granulomatous inflammation of pulmonary arteries, as well as left ventricular failure, as shown in pathological studies (3).

Given the high mortality risk of patients with SAPH, scarcity of data regarding the pharmacological treatment of SAPH, as well as the temporary nature of improvement seen in most studies, an early referral for lung transplantation is recommended (4). Nonetheless, following the results of prior studies (5,6) and in-line with the recommendations of Fifth World Symposium on Pulmonary Hypertension, an off-label therapy with pulmonary arterial hypertension (PAH)-specific agent may be considered. There is, however, no recommendations regarding the choice of most favorable agent.

Conclusions

Our case suggests that specific PH therapy with phosphodiesterase-5 inhibitor can improve the clinical status of patients with end-stage SAPH and right ventricular failure.

Tables 1 A and B. Results of the right heart catheterization showing proof of severe pre-capillary pulmonary hypertension with reduced cardiac output (1A) and result of echocardiographic study (1B) at baseline.

Table 1A

RHC parameters	Result
Systolic pulmonary arterial pressure, mmHg	75
Mean pulmonary arterial pressure, mmHg	58
Pulmonary vascular resistance, Wood units	17
Cardiac output, L/min	2.53
Cardiac index, L/min/m ²	1.81
Wedge pressure, mmHg	14

Table 1B

Echo measurements	Result
LV end diastolic diameter, mm	31
LV ejection fraction, %	40
RV end diastolic diameter, mm	50
RA diameter, mm	55
TAPSE, mm	12
S', cm/s	9
FAC, %	9,5
PASP, mmHg	85
TV regurgitation grade	III - IV
Pericardial fluid	+

Table 2. Results of the main blood tests at baseline and after 1 year

	2017 09	2018 09
NT pro BNP, ng/l	9917	7060
Arterial blood gas (ph, P _a O ₂ , SaO ₂),	7.5/ 51.1/ 85	7.45/71.4/ 94.7
Uric acid, μmol/l	670	511
Creatinine, μmol/l	95	60
Liver transaminases (AST/ALT/ALP/GGT)	21/ 16/ 72/ 132	20/ 17/ 81/ 70
HGB, g/l	189	156

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X. One in a million: a rare case of pulmonary veno-occlusive disease

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Introduction and aim

Pulmonary veno-occlusive disease is a rare cause of pulmonary arterial hypertension (PAH) (about 10% of idiopathic cases). The lowest estimated incidence of PVOD is <1 case/million. Therefore, recognition of the disease is difficult. We want to present a rare and complex case of pulmonary veno-occlusive disease (PVOD).

Case report

In December 2016, a 50-year-old woman was hospitalized to regional hospital due to complaints of progressing dyspnea during exercise from 05/2016. There were no signs of pulmonary embolism, connective tissue diseases, significant pulmonary diseases (COPD, interstitial), left side and congenital heart diseases were excluded, pulmonary hypertension (PH) was suspected on echocardiography. After acute worsening due to infection (pneumonia suspected) the patient was transferred from the regional hospital to our intensive care unit in 01/2017. The patient was hemodynamically unstable (on sympathomimetics), intubated, sedated. Her blood pressure was 90/56 mmHg, heart rate 89 b/min, oxygen saturation 95%.

Echocardiography showed right heart overload and dilatation, signs of severe pulmonary hypertension (systolic pressure 80 mmHg), TV II – III regurgitation, TAPSE 1.6 cm, mild pericardial effusion. Thoracic computed tomography (CT) and pulmonary angiography showed a slight pulmonary artery dilatation; no signs of pulmonary embolism at large and medium branches, pulmonary interstitial-perivascular changes, ground-glass image, lymphadenopathy of mediastinum.

Right heart catheterization (RHC) was postponed due to a very poor condition of the patient and sepsis with multiple organ dysfunction. After multidisciplinary team discussion diagnosis of severe idiopathic PAH functional class (FC) IV was made and PAH target treatment was started with oral Sildenafil and inhaled Iloprost. After 1 week Iloprost was switched to subcutaneous Treprostinil. RHC was performed after hemodynamic stabilization and treatment of infection, showing mean pulmonary arterial pressure (mPAP) 60 mmHg, mean pulmonary capillary wedge pressure (mPCWP) 9 mmHg, pulmonary vascular resistance (PVR) 10.4 Wood, 838 Dyn.

After 1 month (2017-03) the patient came for out-patient medical check-up at PH referral center. Her condition remarkably improved to FC III: increased exercise tolerance, decreased dyspnea. Lung function test was performed showing reduced diffusing capacity (DLCO 39%). During ventilation/perfusion lung scan no signs of thromboembolism were observed.

After 3 month (2017-06) the patient suffered bronchitis, her condition deteriorated: increased dyspnea, decreased exercise tolerance. The woman was hospitalized and RHC repeated showing increased mPAP (60 -> 69 mmHg), mPCWP (9->14 mmHg), PVR (10.4 -> 21 Wood). Additional PAH target treatment with Ambrisentan was added. Due to severe precapillary PH, hypoxemia, reduced diffusing capacity (DLCO < 40%), rapid deterioration on PAH target treatment and findings on thoracic CT (interstitial-perivascular changes, ground-glass image, lymphadenopathy of mediastinum), PVOD was suspected. The patient was discussed with PVOD experts from Paris PH group and it was decided that the most likely diagnosis is PVOD. The woman was immediately referred for lung transplantation and in early 2018 was included to the waiting list.

Discussion

The diagnosis of PVOD for this patient was based on combination of case history, physical examination and radiological findings. As lung biopsy was a gold standard to confirm a histological diagnosis of PVOD, nowadays it is no longer recommended in most cases. The presence of triad: septal lines, centrilobular ground glass opacities and mediastinal lymphadenopathy, are 100% specific for PVOD. This patient was treated with a combination of PAH target therapy due to severe PH with rapid deterioration, though it is recommended to use vasodilators with caution. As there is no established medical therapy for PVOD and poor prognosis, lung transplantation is the only curative therapy.

Conclusions

Severe and atypical PAH, specific findings in instrumental examination can help to identify a rare disease and provide proper treatment strategy.

XI. Changes of right ventricular size and function in severe aortic stenosis with pulmonary hypertension: Cardiac magnetic resonance study

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Introduction

The development of pulmonary hypertension (PH) and right ventricular (RV) failure worsens the prognosis of severe aortic stenosis (AS) undergoing aortic valve surgical or interventional treatment. Therefore diagnostic RV imaging is of great importance to improve the risk stratification and the outcomes in this group of patients.

Aims and Objectives

To investigate changes of RV size and function in severe AS with PH using 2D echocardiography (2D echo) and cardiac magnetic resonance imaging (CMR).

Materials and methods

30 patients with isolated severe AS (aortic valve area <1 cm²) underwent 2D echo and CMR before surgical aortic valve replacement. According to systolic pulmonary artery pressure (sPAP) patients were divided into two groups: I group – sPAP ≥ 45 mmHg (n = 7), II group – sPAP < 45 mmHg (n = 23). Indices of RV volumes, ejection fraction (EF) were analysed by CMR. End diastolic RV diameter and indices of RV free wall longitudinal function (TAPSE and s') were analysed by 2Decho and compared between groups. Echocardiographic parameters of RV size and function were compared with CMR parameters. Statistical analysis was performed using SPSS version 23.0.

Results

There was no significant difference between groups in 2D echo derived end diastolic RV diameter (39 ± 3.4 vs 37 ± 3.5 mm, p = 0.14). However, CMR indices of RV end-systolic volume (55.9 ± 21 vs 29.9 ± 10 ml/m², p = 0.005) and RV end-diastolic volume (94.0 ± 19 vs 64.1 ± 16 ml/m², p = 0.005) were significantly larger in PH group patients and they did not correlated with 2Decho derived RV size (p > 0.05). Indices of RV free wall longitudinal systolic function, estimated by 2Decho in both groups were within normal values, although TAPSE and s' were lower in PH group patients (TAPSE 19.8 ± 3.6 vs 23.5 ± 3.0 mm, p = 0.045, s' 10.5 ± 3.6 vs 13.3 ± 3.7 cm/s, p = 0.018). CMR derived RV EF was significantly lower in PH group patients (41 ± 17 vs $56 \pm 9\%$, p = 0.037) and did not correlated with RV free wall longitudinal function parameters, assessed by 2D echo (p > 0.05).

Conclusions

Higher right ventricular volumes and lower systolic function was found in patients with severe aortic stenosis and pulmonary hypertension. Cardiac magnetic resonance is more accurate to assess changes of right ventricular size and function in this group of patients.

XII. Demographics, clinical characteristics and treatment of chronic thromboembolic pulmonary hypertension patients in Latvia

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Introduction

Chronic thromboembolic pulmonary hypertension (CTEPH) is a type of pulmonary hypertension (PH) that is a complex, debilitating, pulmonary vascular disease, characterised by obstruction or occlusion of pulmonary arteries by postembolic fibrotic material. (Wilkens et al., 2018). Patients with CTEPH can lack a history of acute pulmonary embolism (APE) or deep vein thrombosis in 19-63% of cases (Klok et al., 2018)

Aims and Objectives

To perform a descriptive analysis about the patient demographics, clinical and treatment characteristics of CTEPH in the Latvian population.

Materials and methods

A nationwide, prospective, observational cohort study of the national PH registry was conducted. A total of 55 patients with newly diagnosed CTEPH were enrolled between January 1 2014 and September 30 2018. The diagnosis for all patients was confirmed after at least 3 months of effective anticoagulation by measurement of mean pulmonary artery pressure (mPAP) ≥ 25 mmHg and pulmonary artery wedge pressure ≤ 15 mmHg in right heart catheterization, thrombi in the pulmonary arteries were detected by multidetector CT angiography. Statistical analysis was performed using IBM SPSS 20.

Results

Mean age at diagnosis for CTEPH patients was 69.8 [95% CI = 66.7-72.9; SD = ± 11.8] years. Females made up 67% [54-78] (n=37) of the patients [p=0.008]. The mean body mass index (BMI) was 28.6 [26.8-30.4; ± 6.7] kg/m² ranging from 19.5 to 44.3 kg/m². Of all patients 69% [56-80] (n=38) had BMI ≥ 25 kg/m². The mean baseline mPAP was 43.5 [40.1-46.9; ± 12.7] mmHg. A history of APE was reported for 89% [78-95] (n=49) of the enrolled patients. Pulmonary endarterectomy was done in 11% [5-22] (n=6) of the patients. PH-specific medical therapy after the diagnosis was received by 89% [78-95] (n=49) of patients: 80% [66-89] (n=39) were treated with phosphodiesterase type 5 inhibitors (PDE5i), 12% [6-24] (n=6) received endothelin receptor antagonists (ERA), and 8% [3-19] (n=4) received initial combination therapy (PDE5i+ERA). All patients after CTEPH diagnosis were treated with anticoagulants. 31% [20-45] (n=15) of non-operated patients received direct oral anticoagulants (DOACs), while the remaining received warfarin. At the time of diagnosis, New York Heart Association (NYHA) functional class I-II was observed in 33% [22-46] (n=18), class III in 47% [35-60] (n=26), class IV in 20% [12-32] (n=11).

Conclusions

Females and patients with high BMI are predominant in the CTEPH population. More than 10% of patients with CTEPH had no history of APE. Almost a third of CTEPH patients received DOACs. The elevated mPAP in combination with a high proportion of NYHA class III/IV indicates already progressed disease at the time of diagnosis.

XIII. Great collaboration – case series of successful pulmonary endarterectomy abroad

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Introduction and aim

Chronic thromboembolic pulmonary hypertension (CTEPH) is a potentially curable disease. Pulmonary endarterectomy (PEA) is a first – line effective treatment for patients with surgical accessibility of thrombi in the main, lobar or segmental pulmonary arteries. Patients who do not undergo PEA or suffer from persistent or recurrent PH after PEA (post-PEA PH) face a poor prognosis. PEA is a technically challenging operation currently

successfully performed in experienced centers around the world. The aim was to report case series of PEA performed outside Lithuania at CTEPH expert center in Germany.

Case report

Two young men: patient A and patient B were admitted to our hospital's PH center due to dyspnea after episodes of acute pulmonary artery thromboembolism (PE) and >3 months of appropriate anticoagulation. A diagnosis of CTEPH was confirmed for both. The patients' data are summarised in **Table 1**.

Table 1. Baseline characteristics*:

Before PEA	Patient A	Patient B
Age (y/o)	24	22
Symptoms	severe dyspnea from 2015	exercise dyspnea from 2014
WHO functional class	III	I-II
Thromboembolic events	acute PE and DVT in 2015	DVT in 2013, PE in 2014 recurrence in 2016
Risk factors	protein C deficiency antiphospholipid antibody syndrome splenectomy	antiphospholipid antibody syndrome
Concomitant disease	chronic thrombosis of portal vein; portal hypertension with oesophageal varices (several episodes of gastrointestinal bleeding from oesophageal veins, ligation of veins) 2 stomach ulcers	undifferentiated connective tissue disease
BNP (ng/l)	432,4	14.2
6MWT (m)	350	690
Echocardiography	signs of PH	signs of PH
CT pulmonary angiography	bilateral thrombi in lobal and segmental branches	bilateral thrombi in S4, S9 segmental branches
RHC: mPAP (mmHg)	52	51
mPACW (mmHg)	5	17
PVR (WU)	9,4	4,5
CI (l/min/m ²)	2,32	3,68
Pulmonary angiography	bilateral thrombosis of large and medium branches	
Treatment	Fraxiparine 0,9 ml s/c b.i.d Bosentan 125mg b.i.d -> Ambrisentan 5 mg q.d.**	Warfarine

*when diagnosis was confirmed.

**treatment changed due to elevated liver transaminase levels.

The patients were estimated as technically operable with non-acceptable risk/benefit ratio. When reimbursement from National Health Insurance Fund was received the patients were referred to a specialised Kerghoff clinic, Germany for consultation of prof. E. Mayer. Successful complete bilateral PEA was performed for both patients in March of 2017 and 2018. Improvement of subjective symptoms, functional class and right ventricular function were confirmed (**table 2**).

Table 2. Post – operative characteristics:

Post PEA	Patient A	Patient B
Dyspnea	no	no
WHO functional class	II	I
BNP (ng/l)	35.6	-
6MWT (m)	545	660
Echocardiographic signs of PH	minimal	no
RHC: mPAP (mmHg)	27	-
mPACW (mmHg)	15	-
PVR (WU)	2	-
CI (l/min/m ²)	3,42	-
Treatment	Rivaroxaban 20mg q.d.	Warfarine

Discussion

The guidelines ^[1, 2] states that eligibility for surgery is determined by multiple factors that can not easily be standardised. So treatment of patients with CTEPH depends largely on the subjective assessment of operability, risk/benefit ratio. A controversial aspect of guidelines is the removal of a recommendation for a second surgical

opinion ^[2]. Lithuanian National Health Insurance Fund can reimburse a patient's consultation or the required treatment in all European Union countries when such healthcare service is unavailable in Lithuania or a team of specialists recommends treatment in high-volume centers.

Conclusions

The practice of collaboration between expert centers and selective referral of patients to hospitals with high – volume of procedures is crucial in treatment of a rare disease.

¹Galie N, Humbert M, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension, Eur Heart J (2016) 37 (1): 67-119.

²Konstantinides S.V, Torbicki A, et al. 2014 ESC Guidelines on the diagnosis and management of acute pulmonary embolism, Eur Heart J (2014) 35, 3033–3080.

XIV. Outcomes of pulmonary endarterectomy for patients with chronic thromboembolic hypertension in Latvia

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Introduction

Chronic thromboembolic pulmonary hypertension (CTEPH) is classified as group 4 of pulmonary hypertension. It is a hemodynamic state characterized by chronic obstruction in pulmonary circulation, that develops when unresolved emboli in pulmonary arteries undergo fibrotic transformation. The associated remodelling and dysfunction of the pulmonary microvasculature are believed to be significant factors contributing to pathology of CTEPH.

The treatment of choice is pulmonary endarterectomy (PEA) for all patients who are considered operable by an experienced multidisciplinary CTEPH team. For the patient to be eligible for the surgery, sufficient amount of surgically accessible thromboembolic material is required, with a proportional PVR indicating the absence of extensive secondary vasculopathy. The estimated amount of patients who are operable, ranges from 50 to 70%.

Aims and Objectives

The goal of this study was to review experience of the Latvian pulmonary hypertension center, evaluating outcomes and baseline characteristics of patients who have underwent pulmonary endarterectomy procedure.

Materials and methods

This is a prospective, observational, single-centre study of the Latvian CTEPH registry. We analysed the registry data in the timeframe between September 1, 2007 and December 31, 2016 and all of the patients who underwent pulmonary endarterectomy were included.

The patients were classified in accordance to the New York Health Association (NYHA) functional class and assessed by 6-minute walk test (6MWT).

If possible, a follow-up evaluation which consisted of RHC, TTE and 6MWT was done at 12 months after the surgery to reassess the state of disease and to evaluate patient's functional state.

The data is presented as mean ± standard deviation for continuous variables and as a number or percentage for categorical variables. Statistical analysis was performed using SPSS version 23 software (SPSS Inc., Chicago IL, USA) with p values less than 0.05 considered significant.

Results

PEA was done for 7 patients. The in-hospital mortality was 0%. The 3-year survival rate was 86%. The procedure restored pulmonary blood pressure to normal values for 3 of the patients (42%). The remaining four (57%) had persistent pulmonary hypertension (mPAP>30 mmHg), which required continuous therapy. Six of the patients (86%) had a comprehensive follow-up evaluation 12 months after PEA. The postoperative results can be seen in Table 2. Mean mPAP and RAP decreased from 53.4±14.4 and 12±7.3 mmHg, to 44.3±30.0 (p>0.05) and 11.3±8.7 mmHg (p>0.05), respectively. Mean preoperative PAWP value was 11.6±4 mmHg (p>0.05) with mean PVR of 8.2±2.4 Wood units. After the surgery the mean PAWP had increased to 12.7±10 (p>0.05), however PVR was reduced to 6.8±5.8 Wood units (p>0.05). There were no significant changes in cardiac output and cardiac index values, as they were 4.8±0.4 l/min and 2.4±0.4 l/min/m², post surgery, respectively.

The mean 6MWD at the time of follow-up was significantly improved at 421.8±113.3 metres (p<0.01 compared to baseline). Two of the patients (29%) improved their NYHA functional class from III to II, and two (29%) were in NYHA class I.

There was a significant reduction in BNP, the mean value decreased from 524.5 ± 88.3 pg/ml to 181.3 ± 101.3 pg/ml ($p < 0.01$).

Only 16% of all prevalent latvian CTEPH patients have underwent PEA in the course of nine years, despite it being the treatment of choice for CTEPH.

As PEA and other emerging treatment options, such as balloon pulmonary angioplasty, can only be done in expert centres, numerous organizational, logistical and economical issues arise for patients of smaller countries, where such centers have not yet been created due to lack of experience and limited amount of patients.

Conclusions

This study shows that PEA can be done with limited risk and it significantly improves both hemodynamic and functional state for patients with CTEPH.

XV. Factors influencing survival in patients with pulmonary hypertension: a brief review from single university registry data

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Introduction

Pulmonary hypertension (PH) is a rare pathophysiological disorder that embraces a diversity of diseases with the increase in mean pulmonary arterial pressure (PAPm) ≥ 25 mmHg at rest as assessed by right heart catheterization. Usually progressive pulmonary vascular remodelling and increased right ventricular afterload lead to right heart failure. Regardless of new medications and interventional treatment possibilities the survival of PH patients ranges from several months to several years.

Aims and Objectives

The aim of this study was to investigate the possible factors (gender, age during PH diagnosis, etiology, baseline N-terminal B-type natriuretic peptide (NT-proBNP) level, heart ultrasound data and six-minute walk test (6MWT)) affecting the mortality and survival of PH patients from single PH center.

Materials and methods

Data from the medical charts from the single university hospital PH center were retrospectively analyzed and 59 cases of adult patients diagnosed with PH confirmed by right heart catheterization during the period 2010-2017 were included into the study. There were 18 men and 41 women. The statistical analysis was performed using Statistical Package for the Social Sciences (SPSS), version 20.0. P values of < 0.05 were considered to indicate statistical significance.

Results

Mean overall survival of all selected PH patients was 50 months (95 % CI 40-59).

The shorter survival was seen in PH associated with CTD group comparing with IPAH and Eisenmenger's syndrome group (31 months (95 % CI 18-44) vs. 56 months (95 % CI 41-71), $p = 0.028$ vs. 77 months (95 % CI 59-96), $p = 0.008$ accordingly). Meanwhile mean survival between IPAH, CTEPH and Eisenmenger's syndrome patients did not differ. The risk of death was found to be three times lower for patients with IPAH ($p = 0.039$) and six times lower for patients with Eisenmenger's syndrome ($p = 0.017$), comparing with PH associated with CTD.

Our data also showed that mean survival was higher in group with NT-proBNP ≤ 1500 ng/l (70 months (95 % CI 60-80)) compared to group with NT-proBNP > 1500 ng/l (38 months (95 % CI 24-51)) ($p = 0.002$). Risk of death was five times higher (95% CI Exp(B) 1.6-14.4) in patients with NT-proBNP > 1500 ng/l, comparing with the patients with NT-proBNP ≤ 1500 ng/l ($p = 0.006$).

When analysing echocardiographic data of our selected PH patients, only right ventricular (RV) enlargement and severity of tricuspid regurgitation (TR) were found as independent predictors of survival ($p = 0.001$ and $p = 0.025$ accordingly). Surprisingly we did not found any correlation between the left ventricular ejection fraction, tricuspid annular plane systolic excursion (TAPSE), pulmonary velocity acceleration time (PVAT), pericardial effusion existence and survival. However mean survival was higher in group of patients with RV < 45 mm compare to the patient group with RV > 45 mm (62 months (95 % CI 50-74) vs. 26 months (95 % CI 20-32), $p = 0.001$). Also we observed the higher survival of patients with I-II° TR compared to the patients with III-IV° TR (56 months (95 % CI 46-74) vs. 36 months (95 % CI 26-46), $p = 0.025$).

Furthermore, the gender and the age at the diagnosis as well as baseline 6MWT result according to our data did not have any impact on PH patient survival.

Conclusions

According to single university hospital PH center data, PH associated with CTD, higher baseline NT-proBNP concentration and severe RV enlargement as well as greater TR are associated with worse survival and higher risk of death in PH.

XVI. Relationship between pulmonary function test parameters and heart failure markers in Eisenmenger syndrome

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Introduction

Eisenmenger syndrome (ES), a severe form of pulmonary arterial hypertension, occurs in patients with congenital heart defects if large shunts are not closed in time. It has been reported that up to 60 % of patients with chronic heart failure have ventilation and diffusion abnormalities with reduction of lung volumes on lung function testing. The diagnosis of chronic obstructive pulmonary disease in patients with heart failure is challenging because of both conditions overlap and the absence of usual etiologies for obstructive lung disease. There is controversy about changes in lung function in pulmonary hypertension (PH), especially in patients with ES.

Aims and Objectives

To assess the relationship between lung function and heart failure parameters in patients with Eisenmenger syndrome.

Materials and methods

The retrospective analysis of PH registry in Vilnius University hospital Santaros klinikos was performed. Patients with diagnosis of Eisenmenger syndrome (ES) who underwent comprehensive pulmonary function test (PFT), including spirometry, body plethysmography and gas diffusion evaluation (DLCO), were included in to the study. According to presence of bronchial obstruction ($FEV_1/FVC < 70\%$ and $<$ lower limit of normal (LLN)) patients were divided into 2 groups: with bronchial obstruction and without. Congenital heart disease (CHD) severity was distributed into simple and combined groups. Type of CHD, blood gases, hemoglobin (Hgb), B-type natriuretic peptide (BNP), six-minute walk test (6 MWD), invasive hemodynamic parameters (pulmonary vascular resistance (PVR), mean pulmonary artery pressure (PAP)) and outcomes were compared between the groups.

Results

More than 400 patients with PH were analyzed and 25 with ES were included to the final study. Airway obstruction was found in 7 patients (35 %), they were older than those without obstruction ($p=0.001$). There was not found any significant difference in other pulmonary function test parameters (FVC, FEV_1 , TLC, VC and DLCO) between the groups. DLCO correlated with BNP level ($r=0.623$, $p=0.017$). There was no relationship between bronchial obstruction and DLCO, PVR, PAP, 6 MWD, Hgb and blood gasses.

Patients with combined CHD were younger than patients with simple defects ($P=0.007$). SpO₂ and BNP levels were worse but PVR was lower in the combined CHD group ($p=0.026$ and $p=0.048$, respectively). Increased residual volume (RV) was associated with the worse functional class of NYHA and higher mortality ($p=0.00414$ and $p=0.00266$, respectively).

Conclusions

Bronchial obstruction and diffusion abnormalities were quite common finding in our ES patients group. Lower DLCO was associated with worse heart failure parameters. Higher residual volume was associated with worse NYHA functional class and higher mortality of ES patients. The absence of link between bronchial obstruction, cardiac failure and hemodynamic parameters suggests that bronchial obstruction does not depend on heart failure, but additional multicenter studies with larger patients' number are needed to prove it.

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