162. Clinical characteristics of patients with pulmonary hypertension

P1512

Right ventricular dyssynchrony measured using speckle tracking strain as a predictive factor in pulmonary hypertension patients

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Introduction: RV function is a major prognosis factor in pulmonary hypertension patients. TAPSE has been described as a good predictive factor in this population. Right ventricular speckle tracking strain analysis can quantify regional contraction and identify RV dyssynchrony.
Objective: To test whether RV dyssynchrony exists in PH patients and can predict RV function.
Design: Prospective study.
Results: 42 simultaneous measurements were prospectively obtained using pulmonary artery catheterization and a TTE: hemodynamics, TAPSE, RVEDA/LVEDA. The speckle tracking analysis was used to generate 6 segmental RV strain curves. Time to peak strain from each of 6 time-strain curves was determined with dyssynchrony defined as the difference between earliest and latest segments. Global radial strain was calculated.
MPAP was 38±9 mmHg, PAOP was 12±4 mmHg, cardiac index was 3.0±1.1 L/min/m^2. TAPSE was 17±7, RVEDA/LVEDA was 1.17±0.64. RV global strain was decreased: -12.7±5.7% and RV had dyssynchrony 155±93 msec.
RV dyssynchrony was significantly correlated to RV dilation: y= 75x + 68, r2=0.25, p<0.001 but not correlated to TAPSE: y = 5.77x + 247, r2=0.12, p=0.056.
Conclusion: In our PH patients, we found RV dyssynchrony diagnosed using echocardiographic speckle tracking strain analysis while usual indices of RV function as TAPSE are still normal. This result should be confirmed in a larger population and ask the question. Assessment of RV dyssynchrony could be used for the follow-up of PH patients: predictive factor and non-invasive hemodynamic monitoring. RV resynchronisation therapy could be considered to improve the prognosis of PH patients.

P1513

Clinical evaluation of RV wall stress in pulmonary arterial hypertension: A follow-up study using magnetic resonance imaging

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Background: In pulmonary arterial hypertension (PAH) survival is strongly associated with right ventricular (RV) function and its ability to adapt to the increased pulmonary artery pressure (PAP). RV remodeling to the increased load is often characterized by dilatation, and hypertrophy. RV wall stress is a simple parameter that contains the effects of PAP, dilatation and hypertrophy. Therefore this study aims to evaluate RV wall stress in patients during follow-up.
Methods and results: At baseline 53 patients underwent magnetic resonance imaging (MRI) and right heart catheterization (RHC). In all patients RV end-systolic wall stress (RVESWS) was calculated using the law of Laplace. Eight patients died during the first year of follow-up and therefore 45 patients underwent MRI and RHC after 1-year follow-up. During a median long term follow-up of 57 months another 10 patients died. At baseline, RVESWS appeared to be similar in survivors and non-survivors (n=53, p=0.765). In contrast, change of RVESWS during the 1-year follow-up differed significantly (n=45, p=0.014) between survivors and non-survivors. Survivors showed a decrease in RVESWS during 1-year follow-up, whereas non-survivors showed an increase of RVESWS during 1-year follow-up. Kaplan-Meier analysis showed a higher mortality rate in patients with an increase of RVESWS>17 mmHg than in patients with an increase of RVESWS<17 mmHg or a decrease of RVESWS during follow- up (n=45, p<0.001).
Conclusion: Progressive RV failure is characterized by an increase of RVESWS.

P1514

Ventilation perfusion lung scan in pulmonary veno-occlusive disease

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Introduction: Pulmonary veno-occlusive disease (PVOD) is a rare form of pulmonary arterial hypertension (PAH) that remains poorly understood and is both difficult to diagnose and treat. Histological proof is required for a definitive diagnosis of PVOD; however, this approach is hazardous in patients with pulmonary hypertension and therefore surgical lung biopsy is not recommended. In recent conjoint ERS/ESC guidelines, ventilation and perfusion (V/Q) lung scan was recommended to look for chronic thromboembolic pulmonary hypertension and it has been suggested that unmatched perfusion defects may suggest PVOD.

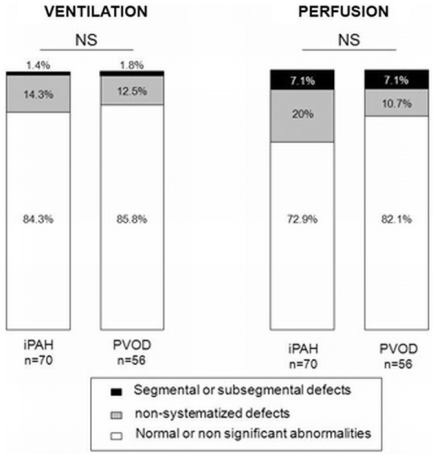

Figure 1. Comparative evaluation of the V/Q scans between iPAH and PVOD groups.

Aim of the study: To evaluate the interest of V/Q lung scan in the non-invasive approach to screen PVOD patients.
Methods: V/Q lung scans from 70 patients with idiopathic PAH and 56 patients with confirmed or highly probable PVOD, were reviewed in double blind.
Results: The vast majority of V/Q lung scan were normal or with no significant abnormalities in both group of patients. No differences in ventilation or perfusion lung scan were observed between idiopathic PAH and PVOD patients (all p values >0.05). No differences were observed between confirmed (n=31) or highly probable PVOD (n=25). Unmatched perfusion defects were found in 7 (10%) idiopathic PAH patients and 4 (7.14%) PVOD patients (p>0.05).
Conclusion: Unmatched perfusion defects were rarely observed in idiopathic PAH or PVOD and V/Q lung scan may be not useful to discriminate PVOD.

P1515
Current practice for determining pulmonary capillary wedge pressure predisposes to serious errors in the classification of patients with pulmonary hypertension
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Background: Accurate measurement of left ventricular filling pressure (LVEDP) is important to distinguish between Group 1 pulmonary arterial hypertension (PAH) and Group 2 pulmonary hypertension from diastolic heart failure (PH-HFpEF).
Methods: We prospectively performed cardiac catheterization on 62 patients referred for evaluation of PH and compared the LVEDP at end-expiration to: a) the pulmonary capillary wedge pressure manually determined at end-expiration (PCWP-EXP) and b) the PCWP determined electronically (PCWP-digital).
Results: The PCWP-EXP was a more reliable measure of the LVEDP (mean 13mmHg vs 12.4mmHg, p=NS) than the PCWP-digital (mean 8mmHg vs 12.4mmHg, p<0.05). Bland-Altman analysis of PCWP-digital and LVEDP revealed a mean bias of -4.4 mmHg (95% limits of agreement -11.3mmHg to 2.4mmHg). Bland-Altman analysis of PCWP-EXP and LVEDP revealed a mean bias of 0.7mmHg (95% limits of agreement -5.5mmHg to 7mmHg). If the PCWP-digital were used to define the LVEDP, 11 patients (18%) would have been misclassified as having PAH rather than PH-HFpEF. Interestingly, 8 of these 11 patients had either morbid obesity or hypoxia. In contrast, no patients were misclassified as PAH instead of PH-HFpEF using the PCWP-EXP to define LVEDP.
Conclusions: The common practice of using PCWP-digital measurements instead of PCWP-EXP measurements results in significant underestimation of the LVEDP. In our study, this translated to almost 20% of patients being misclassified as having PAH rather than PH-HFpEF. Thus, reliance on PCWP-digital measurements should be avoided as this may lead to the inappropriate use of pulmonary vasodilators.

P1516
Increased renin-angiotensin-aldosteron system activity in lungs of patients with idiopathic pulmonary arterial hypertension
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Studies have reported over-activation of the sympathetic nerve system (SNS) in patients with idiopathic Pulmonary Arterial Hypertension (iPAH). Since the Renin-Angiotensin-Aldosteron-System (RAAS) is closely related to SNS and the lungs are the major site for angiotensin 2 formation, we hypothesized that RAAS-activity is increased in iPAH.
Pulmonary endothelial cell (P-EC) cultures were generated from lung specimens of iPAH-patients (n=5) and controls (n=5) to assess angiotensin converting enzyme (ACE) activity (Fig. 1A). We determined angiotensin 2 production upon incubation with angiotensin 1 alone and in combination with ACE-inhibitor enalapril. Subsequently, protein levels of the angiotensin 2 receptor type 1 (AT1R) and tyrosine kinase SRC-activity (downstream target of AT1R) were evaluated in pulmonary arteries (PA) homogenates.
P-EC of iPAH-patients produced significantly more angiotensin 2 upon angiotensin 1 incubation, compared to control. Interestingly, enalapril normalized angiotensin 2 production (Fig. 1B). In addition, pulmonary arteries of iPAH-patients exhibited increased AT1R expression and accentuated SRC-activity (Fig. 1C,D)
Conclusion: This study demonstrates increased RAAS-activity in lungs of iPAH-patients, illustrated by elevated ACE-activity and AT1R signalling. Future studies will focus on the effects of chronic inhibition of RAAS on pulmonary vascular remodelling in iPAH.

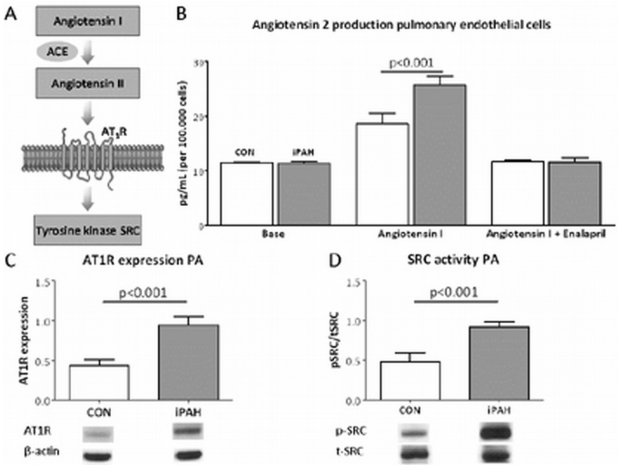

Figure 1

P1517
Changes in right ventricular mass are unrelated to changes in pulmonary pressures in pulmonary arterial hypertension
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Background: In pulmonary arterial hypertension (PAH), chronic pressure overload leads to right ventricular (RV) dilatation, hypertrophy and ultimately RV failure and death. Yet, the relevance of changes in RV mass and the interaction with changes in pulmonary pressure have never been investigated.
Objectives: To assess the relationship between changes in pulmonary pressure, changes in RV mass and survival in PAH patients under PAH targeted therapies.
Methods: 45 patients underwent right heart catheterization to measure mean pulmonary artery pressure (mPAP) and cardiac magnetic resonance to assess RV mass before and after 12 months of therapy. RV mass was indexed for body surface area. During long-term follow-up of 59±27 months, 11 patients died.
Results: At baseline, survivors and non-survivors showed a similar mPAP (p=0.43) and RV mass index (p=0.06). RV mass index correlated to mPAP (R=0.51, p<0.01). During 1-year follow-up, changes in mPAP did not differ between survivors (-8±28%) and non-survivors (3±30%) (p=0.86). Overall, RV mass increased by 11±26% (p=0.03). Survivors showed an increased RV mass (17±25%; p<0.01), non-survivors showed an unchanged RV mass (-7±11%; p=0.16). Changes in RV mass were unrelated to age (p=0.50) or gender (p=0.44). Changes in RV mass were unrelated to changes in mPAP (R=0.14; p=0.37).
A decreased RV mass was associated with mortality (HR: 0.95; 95%-CI 0.91-0.99; p=0.02) whereas changes in mPAP did not relate to mortality (HR: 1.01; 95%-CI 0.98–1.02; p=0.25).
Conclusions: In PAH, changes in mPAP were not followed by changes in RV mass. An increased RV mass during follow-up was associated with a favorable prognosis.

P1518
Systemic hypoxia contributes to pulmonary hypertension in heart failure
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Background: The classical mechanism for pulmonary hypertension (PH) in heart failure (HF) is related to a passive rise in pulmonary venous pressure secondary to an increase in LV filling pressure. However, much of the PH in HF is reversed by vasodilators. This suggests a reactive component to the elevated pulmonary vascular resistance (PVR) that is likely related to dysregulation of vascular smooth muscle tone. Although the cause of this dysregulation is unclear, hypoxia may play a role. We examined whether variations in resting systemic O_2 levels influence PVR in HF.
Methods: Thirty-nine patients (54±9 yr, LVEF 20±6%, NYHA class I-III) undergoing right heart catheterization for pre transplant assessment were studied. PVR was derived from pulmonary arterial and wedge pressures measured via Swan Ganz catheter and cardiac output measured via direct Fick. Mixed venous and arterial blood was drawn from the pulmonary and radial arteries for measurement of PaO_2, PvO_2, SaO_2, SvO_2 and endothelin-I (ET-I).
Results: Group mean PVR, PaO_2, PvO_2, SaO_2 and SvO_2 were 303±215 dynes/s/cm^5, 72±12 mmHg, 32±4 mmHg, 94±4% and 57±11%, respectively. PVR was negatively correlated with PaO_2, PvO_2, SaO_2 and SvO_2 (r= -0.53, -0.62, -0.48, -0.67, respectively, all P<0.01). Multiple linear regression suggested that SvO_2 was the strongest predictor of PVR. In combination, PaO_2, PvO_2, SaO_2 and SvO_2 accounted for ~60% of the variance in PVR. In addition, ET-I was related to both SvO_2 (r= -0.75) and PVR (r=0.50) (P<0.01).
Conclusion: Systemic hypoxia, particularly a low SvO_2, appears to play a role in PH and elevated PVR in HF. We suggest a hypoxia-mediated increase in the release of the vasoconstrictor ET-I as a likely mechanism. NIH HL71478

P1519

Atrial arrhythmias in pulmonary hypertension

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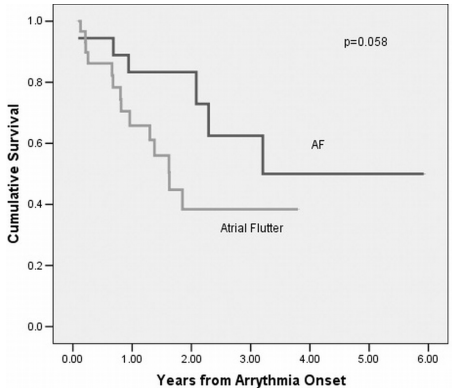
Introduction: There are few data regarding the frequency, importance and management of atrial arrhythmias in pulmonary hypertension (PH).
Methods: A six-year retrospective analysis was conducted of >1000 newly diagnosed patients with PH.
Results: There were 264 pre-existing cases of atrial fibrillation (AF) including 29% pulmonary arterial hypertension (PAH), 53% PH due to left heart disease (PH-LHD), and 7% chronic thromboembolic PH (CTEPH). PAH and CTEPH were more common (45% and 24%) than PH-LHD (17%) as the underlying cause of PH in the 29 patients who had atrial flutter (flutter) diagnosed prior to PH diagnosis ($p<0.05$). 49 new diagnoses of flutter or AF were made at PH diagnosis (n=6) or during follow-up (n=43). ~7% of all PAH patients developed an atrial arrhythmia. Baseline age, exercise capacity and pulmonary haemodynamics were not significantly different between the flutter and AF group but initial mixed venous oxygen saturations were lower in patients developing flutter (58% *v* 64%, p=0.02). Management including chemical or DC cardioversion and/or ablation therapy maintained 59% of patients in sinus rhythm (SR). Failure to maintain SR was not associated with prognosis (p=0.9). Survival was worse in patients developing flutter rather than AF (figure).



Conclusion: Atrial arrhythmias are common in PH. AF is more commonly associated with PH-LHD and developing flutter was associated with poorer survival. Further analysis of optimal management strategies is required.

P1520

Estimation of right ventricular isovolumic pressure in experimental pulmonary hypertension from a single ejecting beat

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In pulmonary hypertension (PH), assessment of right ventricle (RV) contractility from end-systolic pressure-volume relationships (ESPVR) is difficult due to the requirement of multiple-beats by alterations in cardiac preload. Several single-beat (SB) methods have been proposed to calculate ESPVR, mostly based on estimation of maximum isovolumic pressure (P_{max}) using arbitrary wave shapes of isovolumic pressure. The purpose of this study was to derive experimentally the isovolumic pressure wave shape of the RV and to use this curve to compute $P_{max(SB)}$. Isovolumic pressure curves were determined by clamping the pulmonary artery in 4 control, 3 stable PH and 3 progressive PH rats. PH was induced by monocrotaline (MCT) 40 and 60 mg/kg, respectively. All curves were normalized in amplitude and duration to obtain the typical isovolumic pressure wave shape. This curve was used to estimate $P_{max(SB)}$ in 9 control, 7 MCT40 and 7 MCT60 rats from ejecting RV pressure curves. $P_{max(SB)}$ values were compared to values obtained from multiple pressure-volume loops by vena cava occlusion [$P_{max(VCO)}$]. Three SB methods from literature were included for comparison. With our method close correlations were found between $P_{max(SB)}$ and $P_{max(VCO)}$ ($r^2=0.85$, $p<0.001$). The other methods had r^2 values of 0.74, 0.82, and 0.85; $p<0.001$. The latter two methods significantly underestimated $P_{max(VCO)}$ in MCT60 rats, and all three methods resulted in less realistic shapes of isovolumic pressure waves compared to our experimental curves. In conclusion, with our method realistic isovolumic pressure curves can be obtained providing accurate estimates of Pmax over a wide range of RV contractility.

P1521

Contribution of increased afterload to right ventricular ejection fraction in pulmonary hypertension

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Background and aim: Right ventricular (RV) function is closely related to functional capacity and survival in pulmonary hypertension (PH). Our prospective cardiac magnetic resonance imaging (CMR) study attempted to quantify the contribution of increased RV afterload to the decreased right ventricular ejection fraction (RVEF) in PH patients.
Material and methods: 72 patients with precapillary PH (aged 59±15 years; 35F) underwent right heart catheterization and CMR on a 1.5 T scanner (Siemens) with electrocardiographic gating (delay ±48h). The main diagnoses were chronic thromboembolic PH (n=41) and pulmonary arterial hypertension (n=18). RVEF was calculated by using contiguous 6 mm RV short axis cines (balanced SSFP).
Results: Patients had increased mean pulmonary artery pressure (45±12 mmHg) and pulmonary vascular resistance (9±4 wu) and decreased RVEF (30±15%). The RVEF was more strongly related to RV end-systolic volume (RVESV = 78±36 mL/m^2; r = -0.80) than to RV end-diastolic volume (RVEDV = 109±35 mL/m^2; r = -0.53) (each $P < 0.001$). The combined influences of mean pulmonary artery pressure and pulmonary artery pulse pressure explained 27% of RVEF variability. The combined influences of pulmonary vascular resistance and pulmonary arterial compliance explained 37% of RVEF variability.
Conclusion: The classical indices quantifying RV afterload explained less than 40% of RVEF variability in PH, thus suggesting that other factors (e.g., decreased contractility, tricuspid insufficiency, cardiac remodeling) played a major role in right heart dysfunction.

P1522

Chronic thromboembolic pulmonary hypertension and idiopathic pulmonary hypertension are the conditions predisposing to non-tuberculous mycobacterial lung disease (NTMLD)

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Non-tuberculous mycobacterial lung disease (NTMLD) is a rare disorder diagnosed in 0.6-1.8/100,000 people. Nevertheless NTMLD may be observed more frequently (2-6%) in the patients (pts) with predisposing conditions such as chronic lung diseases (COPD, cystic fibrosis), genetic disorders and many others. The aim of the present study was to assess the frequency and clinical features of NTMLD in CTEPH and IPAH pts. 250 patients (150 with CTEPH and 100 with IPAH) diagnosed and treated in the Department of Chest Medicine, National Institute of Tuberculosis and Lung Diseases in the period of 2002-2008 entered the study. NTMLD fulfilling the criteria of ATS 2007, was diagnosed in 9 pts (3.6%): 6 pts with CTEPH (4%) and 3 with IPAH (3%). Majority of patients presented with exacerbation of dyspnea. Chest CT scans revealed areas of infiltration with cavitation, surrounded by small nodules in 4/9 pts, cavities surrounded by small nodules in 4/9 and cavities only in 1 pt. NTMLD-related lung pathology developed in the areas with hypoperfusion, no parenchymal lung pathology was seen on CT scans taken before the disease development. The responsible pathogen was M. kansasii in all of the patients. NTMLD was observed more frequently in CTEPH and IPAH patients with BMI<25, with low cardiac output (CO) and low mixed venous blood saturation (satO2mv).
Conclusion: CTEPH and IPAH are probably the diseases predisposing to NTMLD. The disease develops in the hypoperfused regions of lung parenchyma, in the patients with low CO and satO2mv.

P1523

Circulating fibrocytes in pulmonary arterial hypertension

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Bone marrow-derived progenitor cells have been shown to participate in angiogenesis, vascular repair and tissue remodelling in pulmonary arterial hypertension (PAH). A unique subpopulation of peripheral blood mononuclear cells (PBMC), termed fibrocytes, has been demonstrated to develop myofibroblasic phenotype and contribute to the vascular remodelling process in hypoxic models of PAH. To extend these studies, we tested the hypothesis that circulating fibrocytes contribute

to human PAH and examined their possible role as predictors of activity and progression of disease.
18 patients with idiopathic and familial PAH and 20 healthy control subjects were studied. Circulating fibrocytes, identified as CD11b+CD34+vimentin+ cells and membrane CD11b expression were quantified by flow cytometry. The in vitro differentiation capacity of PBMC to fibrocytes was quantified.
We observed a slight decrease in the percentage but not in the number of circulating fibrocytes in the blood of PAH patients, compared with healthy control subjects (0.67±0.015% in control subjects vs 0.22±0.04% in PAH patients, $p<0.05$). Accordingly, a significant decrease in the percentage of differentiated CD45+vimentin+ cells in PBMC culture from PAH patients was observed (73±3% in control group vs 61±2% in PAH patients, $p<0.05$). Interestingly, the mean fluorescence intensity of CD11b was significantly increased on circulating fibrocytes in PAH patients (1112±116 in control group vs 2167±247 in PAH patients, $p<0.0001$), indicating their increased activation state.
Our data suggest that a more detailed analysis of circulating fibrocyte function and activation is needed in PAH patients.

P1524

Role of mast cells and chymase in pulmonary vascular remodeling

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Rationale: Mast cells (MCs) are implicated in chronic inflammation and tissue remodeling. However, a systematic investigation of pulmonary MCs/MC chymase is yet missing in idiopathic pulmonary arterial hypertension (IPAH) and chronic obstructive pulmonary disease (COPD).
Methods: Lung tissues obtained from donors, and IPAH and COPD patients undergoing lung transplantation were formalin-fixed and paraffin-embedded, followed by toluidine blue (TB) staining for MCs and immunostaining for MC chymase. Total and perivascular MCs were determined by counting MCs under light microscope equipped with computerized morphometric system. Perivascular MCs were categorized as granulated and degranulated and an index of granulation (IOG) [(number of granulated/degranulated MCs)] was determined.
Results: Pulmonary MCs were prevalent in IPAH and COPD patients; furthermore, perivascular MC count was significantly increased in the resistance vessels of patients ($p<0.05$ vs donors). Notably, the IOG was decreased by about 8 and 5 folds in IPAH and COPD patients, respectively (vs donors). Chymase-positive MCs were increased by 16 and 10 folds in IPAH and COPD patients, respectively (vs donors). The perivascular chymase-positive MCs were significantly increased in IPAH and COPD patients ($p<0.05$ vs donors). Interestingly, the chymase-positive MC subpopulations were about 42% and 48% of the MCs in IPAH and COPD patients, respectively; whereas it was 10% in donors.
Conclusion: The chymase released from activated perivascular MCs may potentially contribute to the pulmonary vascular remodeling in IPAH and COPD. Future studies are essential to substantiate the findings and to elucidate underlying pathomechanisms.

P1525

Frequency and impact on prognosis of signs of pulmonary veno-occlusive disease on high resolution computed tomography in patients with scleroderma associated pulmonary arterial hypertension

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Introduction: Pulmonary veno-occlusive disease (PVOD) is an uncommon form of pulmonary arterial hypertension (PAH) characterised by a progressive obstruction of small pulmonary veins. PVOD has been frequently reported in patients with scleroderma related PAH (SSc-PAH). High resolution chest computed tomography (HRCT) is a non-invasive diagnostic tool used to screen for PVOD. However, no data are available in SSc-PAH patients.
Aims: To evaluate the frequency and the impact on prognosis of signs of PVOD on HRCT in SSc-PAH.
Methods: We reviewed HRCT data in 34 consecutive SSc-PAH patients and 30 systemic sclerosis (SSc) patients.
Results: Lymph nodes enlargement (57.7% vs 3.6%), centrilobular ground-glass opacities (46.2% vs 10.7%) and septal lines (73.1% vs 7.1%) were significantly more frequent in SSc-PAH patients as compared to SSc patients (all $P<0.005$). Indeed, 61.5% of SSc-PAH had ≥2 radiological signs of PVOD on HRCT. 53.8% of SSc-PAH patients had evidence of pericardial effusion ($P<0.001$). Cardiomegaly and pulmonary artery enlargement were significantly more frequently observed in SSC-PAH patients ($P<0.001$). Pleural effusion was observed in one patient (3.8%) in the group SSc-PAH, whereas no SSc patient had a pleural effusion. Survival in SSc-PAH patients with ≥2 radiological signs of PVOD was significant lower compared to those ≤1 radiological sign of PVOD ($P<0.05$).
Conclusion: Signs of PVOD are frequent on HRCT in patients with SSc-PAH compared to SSc patients without PAH. These signs allow clinicians to detect PVOD in SSC-PAH patients. Survival in affected patients is poor.

P1526

How does variation of the bag volume in inert gas rebreathing cardiac output measurements influence the reproducibility in patients with pulmonary diseases?

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Background: Cardiac output (CO) is an important hemodynamic parameter, however its determination is difficult in daily clinical routine. Non-invasive inert gas rebreathing (IGR) showed promising results in recent investigations with the volume of the rebreathing bag (V_b) being an important factor. The aim of our study therefore was to evaluate the influence of different V_b on the reproducibility of IGR.
Methods: The collective consisted of 45 patients (age 26 to 88 years). The CO was determined in patients with obstruction (group A), restriction (group B) and pulmonary healthy controls (group C). For V_b of 2200 ml, 1700 ml and 1200 ml two repeated measurements were taken each. The determination of lung function was performed using bodyplethysmography.
Results: Pulmonary obstruction was diagnosed in 12 patients (FEV_1 52±21%) and restriction in 11 patients (VC 61±16%). The mean CO did neither differ between the groups ($p=0.1$) nor for the different V_b ($p=0.2$). The mean bias between the repeated measurements was 0.2±0.9l/min for V_b=2200ml, 0±0.7l/min for V_b=1700ml and 0.3±0.7l/min for V_b=1200ml. There was no statistically significant difference between the groups for the different V_b=2200ml ($p=0.7$), 1700ml ($p=0.4$) und 1200ml ($p=0.2$).
Conclusion: The reproducibility of IGR is not negatively affected by V_b, so that it can be varied between 1200 und 2200 ml. This is especially important when V_b has to be reduced due to incomplete inspiration. The trend to a worse reproducibility at extreme volumes at rest should be further investigated. For now, measurements should only be compared directly when identical V_b were used.

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Characteristics of patients with pulmonary arterial hypertension associated with congenital heart disease in the French PAH registry

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Background: Epidemiological data relative to pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD) are scarce. In the first French PAH registry conducted in 2002-2003, CHD accounted for 11.3%.
Objective: To analyze PAH associated with CHD in patients enrolled in the second prospective PAH registry initiated in 2006.
Methods: PAH-related clinical and outcome data were reviewed and analyzed from the registry.
Patients and results: 2585 patients with PAH were enrolled in 26 PAH centers. CHD-PAH (n = 255) accounted for 9.8%, including 95 isolated pre-tricuspid shunts (mainly ASD), 134 isolated post-tricuspid shunts (mainly VSD), 11 combined pre-and post-tricuspid shunts and 15 complex CHD. 60% of patients were females and mean age at diagnosis was 37 years. The diagnosis of PAH was done simultaneously with the diagnosis of CHD in 37% of the cases and in 60% PAH appeared during the follow-up of CHD. At study entry, 52% of patients were in NYHA functional class (FC) III or IV, 6MWD was 370±105 m and pulmonary hemodynamics were: mPAP= 59±20 mmHg, CI= 2,7±1.1 L/min/m^2 and PVR= 12.5±10.3 WU. 47% of NYHA II and 43% of NYHA III patients were not receiving PAH-specific therapies. In treated patients (n = 164), NYHA FC improved (59% in NYHA FC I-II at last follow-up). During the 3-year follow-up period, 20 patients died and 7 patients were transplanted.
Conclusions: PAH is a complication of a previously known CHD in 60% of cases. ASD is the main CHD that is diagnosed concomitantly or after PAH. Less than a half of NYHA III patients are offered PAH-specific therapies. Mortality was low during the short period of follow-up.

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Idiopathic pulmonary arterial hypertension in the elderly: Data from the French registry

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Rationale: Data from recent registries have shown that the mean age of patients with pulmonary arterial hypertension (PAH) is increasing.
Aim: To describe clinical and haemodynamic characteristics of incident (i.e. newly diagnosed) patients >70 yr included in the French Registry with a diagnosis of idiopathic (IPAH), heritable (HPAH) or anorexigen-related (ArPAH) PAH.
Methods: Data from all newly-diagnosed adult patients with IPAH, HPAH or ArPAH included in the French Registry between November 2006 and November 2009 were collected. Patients without right heart catheterization confirmation and those with a pulmonary capillary wedge pressure > 15 mmHg were excluded.
Results: Among the 350 patients who met the inclusion criteria, 100 (28.5%) were >70 yr. Clinical and haemodynamic characteristics at time of diagnosis are shown in the Table. Survival rates at 1, 2 and 3 years were 79%, 60% and 47% in those >70 yr, as compared to 92%, 84% and 76% in the 18-70 yr group, respectively ($p<0.01$).

	>70 yr	18–70 yr
Number of patients	100	250
IPAH : HPAH : ArPAH, %	88 : 0 : 12	81:7:12
Sex ratio F/M	1	1.29
Age, yr	76±3	51±14
NYHA FC, I-II : III : IV, %	16 : 63 : 21	24 : 62 : 14
6-min walk distance, m	242±108	349±119
Right atrial pressure, mmHg	8±5	9±6
Pulmonary artery mean pressure, mmHg	44±9	52±13
Pulmonary capillary wedge pressure, mmHg	9±4	9±4
Cardiac index, L/min/m^2	2.4±0.6	2.4±0.8
Pulmonary vascular resistance, d·s·cm^{-5}	716±276	904±498

Conclusions: These results show an increasing proportion of elderly male among patients with IPAH and ArPAH. Patients >70 yr have a worse outcome than younger patients despite a less severe haemodynamic impairment.

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Assessment of operability by means of CTPA and perfusion SPECT in patients with chronic thromboembolic pulmonary hypertension

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Objective: Chronic thromboembolic pulmonary hypertension (CTEPH) is curable with pulmonary endarterectomy (PEA). The criteria for identification of PEA-amenable patients need to be standardized.The aim of this study was to evaluate the value of rigidly registered CT pulmonary angiography (CTPA) and perfusion SPECT in differentiating between operable and non-operable patients.
Methods: 49 patients with CTEPH (21 men, 58±13 years) were evaluated for PEA by interdisciplinary board using available diagnostic information and served as the gold standard.SPECT was evaluated by a lobe based visually assessed perfusion score ranging from 0 [no perfusion] to 1 [normal perfusion]),after which the percentage of vascular obstruction (PVO) was calculated: PVO = [1 – Perfusion score] × 100. By CTPA, the vascular obstruction index (OI) of central, peripheral, and global PA bed and diameters of large vessels(pulmonary artery (PA), aorta (Ao) and PA/Ao) was determined. In angiography PA pressure (PAP), PA resistance (PVR) and wedge pressure (PCm) were determined. Receiver operating characteristics (ROC) analysis was performed.
Results: Mean PAP, PVR and PCm was 48±11 mmHg, 878±461dynes sec cm^{-5}, and 11±5 mmHg. 30 patients were chosen as candidates for PEA. Hemodynamic values were not able to differentiate between operable and non-operable patients. PVO and central OI separated PEA-amenable patients (both $p<0.001$) resulting in the area under the curve of 0.845 (sensitivity of 83% and specificity of 83%) and 0.805 (sensitivity and specificity of 86% and 84%).
Conclusion: An accurate interpretation of rigidly registered CTPA and perfusion SPECT may contribute to stratification of operability in patients with CTEPH.