263. Clinical features of pulmonary hypertension

P2320
Multislice CT angiography and pulmonary involvement in asymptomatic systemic lupus patients with antiphospholipid syndrome
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Objective: To detect the pulmonary involvement in asymptomatic secondary APS patients by pulmonary function tests (PFTs) and chest HRCT angiography and comparing the pulmonary findings to those of asymptomatic SLE patients without APS.

Patients and methods: Thirty-four SLE patients with APS and another 34 SLE patients without APS and with a negative ACL test were included as control. Plain chest x-ray, HRCT angiography and PFTs were performed for all patients in an attempt to assess the pulmonary vasculature and lung parenchyma. Disease activity was assessed using the Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) while assessment of organ damage was made using the Systemic Lupus International Collaborating Clinics/ACR (SLICC/ACR) index.

Results: There were abnormal pulmonary CT findings in 11 (32.35%) of the asymptomatic secondary APS patients with an obvious association to lupus anticogulants. However, plain chest x-ray showed basal atelectasis and/or elevation of the copulae in 4 patients. Pulmonary abnormalities included a high frequency of pulmonary artery aneurysms (20.59%) thrombosis, basal atelectasis, embolism, bronchiectasis, pleural effusion and thickening. The SLEDAI and SLICC were significantly higher in APS patients. The PFTs including FVC, FEV1 and FEV1/FVC were reduced in APS patients compared to SLE patients only.

Conclusion: HRCT pulmonary angiography is useful in demonstrating the entire thoracic spectrum in asymptomatic APS patients. PFTs is reduced in SLE patients with APS compared to SLE patients only.

P2321
Thoracic affection during Behcet’s disease (about 15 cases)
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Behcet’s disease is a multisystem vasculitis that affects the young. Chest affection is rare and primarily venous. The arterial affection is exceptional. We report a retrospective study of 15 cases of Behcet’s disease with chest involvement gathered at the Respiratory Diseases department, UHC Ibn Rochd of Casablanca from 1994 to 2010. It was about 3 women and 12 men. The average age was 34 years old (range: 19-58 years). The bipolar aphthosis was found in all patients, pseudofolliculitis in 9 cases, and hemoptysis in 7 cases. Superior vena cava syndrome was noted in 8 cases, 6 cases of pleurisy. The telerothorax showed bilateral pleurisy in 1 case, unilateral in 5 cases, bilateral hilar opacities projection in one case, an alveolar opacity in 3 cases, aspect of releasing balloons in one case, and it is normal in 4 cases. The chest angio-CT showed the superior vena cava’s thrombosis in 9 cases, bilateral pulmonary arterial aneurysms in 2 patients, pulmonary artery aneurysm associated to an embolism in one patient and pulmonary infarction site in 3 cases. Echocardiography showed an intracardiac thrombus in one case. All patients were administered steroids, colchicine and anti-coagulants. Immunosuppressive drugs were administered for 3 patients. The evolution was good in 11 cases, the onset of neuro-Behcet in 1 case and we deplore 4 deaths due to a lightning hemoptysis. We emphasize throughout these series the scarcity of thoracic manifestations of Behcet’s disease, namely the arterial affection that makes its prognosis null.

P2322
Prevalence of hepatopulmonary syndrome in candidates for liver transplantation in Santa Casa Hospital (Porto Alegre-Brazil)
Eduardo Garcia, Taiane Francieli Rebelatto, Fernanda Waltrick Martins, Lucas Jesus de Medeiros, Rangel Olsen de Carvalho, Ajajio Brandao. Internal Medicine, Santa Casa Hospital, Porto Alegre, Brazil

Background: Hepatopulmonary syndrome (HPS) is a disorder of pulmonary vascular disease secondary to liver disease, comprehending abnormalities of arterial oxygenation (PaO2 <80 mmHg) or alveolar-arterial oxygen gradient (PA-aO2 ≥15 mmHg on room air, and in larger 64 years PaO2 <70 mmHg or PA-aO2 ≥20 mmHg) and presence of pulmonary vascular dilatation. There are reports of its occurrence between 4-32% of cirrhotic liver transplant candidates. It is presence, independently and significantly, increases mortality and reduces the survival rate of those patients when compared with cirrhotic patients without HPS.

Objective: To assess the prevalence and severity of HPS in patients with severe liver disease.

Methods: Retrospective cross sectional review of 90 medical records of 90 patients who underwent liver transplantation at the Santa Casa Hospital (Porto Alegre-Brazil), from February 2002 to April 2009. The present study was approved by the Hospital Ethics Committee.

Results: The sample consisted of 90 patients, 62.2% (n = 56) were male and mean age was 54.13 years (22-74). The prevalence of HPS was 43.3% (n=39). Forty one per cent (n=16) were classified as having mild HPS, 33.3% (n=13) moderate, 10.3% (n=4), severe and 15.4% (n = 6) as very severe. The mean Pa-aO2 was 26.42 mmHg and the mean PaO2 was 81.47 mmHg.

Conclusion: The study confirms the high prevalence of this clinical syndrome in patients with severe liver disease and, thus, strengthens its research in patients with liver diseases, especially those who may be liver transplantation, since current evidence points to liver transplantation as a treatment of HPS.

P2323
Withdrawn
P2324
Pulmonary hypertension in patients with infective endocarditis
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Aim and methods: To study pulmonary hypertension (PH) in infective endocarditis (IE) pts with different localization, valve injury and activity of disease we have investigated 136 pts with native valve IE, in which 105 (76%) had left and 34 (25%) – sided IE. Diagnosis of IE was made according to Duke-criteria (ECS, 2009). Pulmonary artery systolic pressure (PASP) was measured using trans-thoracic ECHO in dynamics taking into account the systolic tricuspid gradient and right atrium pressure.

Results: PH was revealed in 132 pts with IE (97%). Mean PASP was 53±7±16.2 mmHg. Pts with minimal and moderate PH prevailed (1st degree PH in 61 pts (45%), 2nd degree – in 50 pts (37%)), only 15 pts (21%) proved high degree PH (PASP>70 mmHg). No significant differences of PH level were found depending on the gender, age, left or right-sided localization of IE, acute stage or 1-year or more advanced stage of disease duration. Nevertheless the PH degree was significantly higher in the group of pts with bi- valve or multiple valve injury in comparison with group of pts with mono-valve pathology (61,3±15,8 vs 49,5±14,9, p<0,001). Among haemodynamic factors influencing the PASP most possible were left atrium diastolic size (R=0,556386, p=0,00001) and myocardium mass index (R=0,47, p<0,0001).

Conclusions: SO PH naturally develops in pts with IE and haemodynamic factors proved to be most significant in its development.

P2325
Which patients with pulmonary arteriovenous malformations are dyspnoeic? Retrospective analysis of a single-centre 2005-2010 cohort
Vasanthan Santhirapala1,2, Hannah Tighe2, James Springett 2, Heidi Wolfenden2, Retrospective analysis of a single-centre 2005-2010 cohort

Which patients with pulmonary arteriovenous malformations are dyspnoeic? Retrospective analysis of a single-centre 2005-2010 cohort
Vasanthan Santhirapala1,2, Hannah Tighe2, James Springett 2, Heidi Wolfenden2, Retrospective analysis of a single-centre 2005-2010 cohort

Background: Pulmonary arteriovenous malformations (PAVMs) generate a right to left shunt. Hypoxaemia is common, but dyspnoea is usually not the presenting complaint.

Methods: To identify factors contributing to dyspnoea, a retrospective analysis was performed in 165 consecutive patients presenting with CT-proven PAVMs between 2005-10. Based on reported exercise tolerance at presentation, and blinded to physiological measurements, two investigators assigned patients to the Medical Research Council dyspnoea scale, with individuals undertaking regular/intense sport reclassified as Grade “0”.

Results: 44 (27%) patients were dyspnoeic (24% excluding the 5 with severe coexistent respiratory disease). In univariate analyses, oxygen saturations (SpO2), displayed no clear relationship with dyspnoea grade (Fig. 1, left). Patient ages ranged from 17-87 years however, and older patients were more dyspnoeic (Fig. 1, right).

Once adjusted for age, there was a contribution of SpO2 to dyspnoea grade, although age and SpO2 did not account for most of the variation (r2=0.19%).

Conclusions: It is unusual for PAVMs alone to account for moderate to severe dyspnoea.

P2326
The correlation between hepatopulmonary syndrome and cirrhosis etiologic diagnosis in candidates for liver transplantation at Santa Casa Hospital Eduardo Garcia, Lucas Jesus de Medeiros, Fernanda Waltrick Martins, Taiane Francieli Rebelatto, Rangel Olsen de Carvalho, Ajacio Brandiao. Internal Medicine, Santa Casa Hospital, Porto Alegre, Brazil

Background: Hepatopulmonary syndrome (HPS) is defined by a triad composed of liver disease, pulmonary vascular dilatation and changes in blood gases, characterized by alveolar-arterial oxygen gradient ≥15 mmHg or PaO2 <80mmHg. Liver cirrhosis of different etiologies is responsible for more than 90% of cases of HPS.

Objective: To correlate the prevalence of HPS to diagnosis of liver cirrhosis in candidates for liver transplantation.

Methods: Cross sectional retrospective review of medical records of 90 patients with chronic liver disease undergoing liver transplantation at the Santa Casa Hospital (Porto Alegre, Brazil) in a period of 2 years; from February 2002 to April 2009. The study was approved by the Hospital Ethics Committee.

Results: Of the 90 individuals selected, 39 (43.3%) were diagnosed with HPS. Separated, then two groups: Group I (patients without HPS) and Group II (patients with HPS). Group I (n = 51): 47% (n=24) had liver cirrhosis by hepatitis C virus (HCV) alone, 7.8% (n=4) by alcohol alone and 21.6% (n=11) by association alcohol and HCV. Group II (n = 39): 33.3% (n=13) by HCV alone, 10.2% (n=4) by alcohol alone, 23.1% (n=9) by the association of HCV and alcohol. Thus, the total number of individuals with diagnosis of HCV, alone or in combination with other causes, was 35 (68.1%) and 25 (64.1%) in Group I and II, respectively, and for alcohol was the total of 15 patients (29.4%) in Group I and 13 patients (33.3%) in Group II.

Discussion: The prevalence for HCV and alcohol was similar between the groups.
Thus, we can infer that the main risk factors, hepatitis C and alcohol, do not act as independent predictors for the development of HPS.

P2327
Childhood pulmonary arterial hypertension: Insights from REVEAL

Background: The 55-center, observational, US Registry to Evaluate Early And Long-term pulmonary arterial hypertension (iREVEAL) Disease Management (REVEAL) examines demographics, clinical course and management of patients (pts) diagnosed with PAH.

Methods: Pts aged ≥3 months and ≤18 years at time of confirmatory diagnostic catheterization were followed at 26 sites. Data were analyzed to explore demographic, treatment and outcomes in the following PAH subgroups: idiopathic PAH (IPAH), familial PAH (FPAH), and PAH associated with congenital heart disease (APAH-CHD).

Results: 3518 pts enrolled in REVEAL between 03/2006 and 12/2009; 77 APAH-CHD and 122 IPAH/FPAH had childhood-onset PAH; mean ± SD age at PAH diagnosis 6.6 ± 6.6 years and 9.6 ± 6.6 years (P = 0.002), respectively. APAH-CHD had similar functional class (FC) at diagnosis compared with IPAH/FPAH (FC: I, 5% vs I, 5% vs = 0.53), though phosphodiesterase 5 inhibitors (45% vs 64%, respectively; P = 0.011) and prostacyclin (28% vs 50%, respectively; P = 0.003) analogs were used less in APAH-CHD. Five-year survival from diagnosis was similar for APAH-CHD and IPAH/FPAH (71.4% vs 75.7% vs, respectively; P = 0.53).

Conclusion: Childhood-onset APAH-CHD and IPAH/FPAH have similar hemodynamics and FC and similar 5-year survival from diagnosis. REVEAL provides observational data on treatment and outcomes in childhood PAH that should prove useful in identifying prognostic parameters, thereby helping clinicians improve outcomes.

WITHDRAWN
P2328
Pulmonary hypertension in lymphangioleiomyomatosis: Hemodynamic characteristics in a series of 20 patients
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Background: The prevalence of pulmonary hypertension (PH) has been estimated to be 6.7% in 95 patients with lymphangioleiomyomatosis (LAM) as defined by sPAP>35 mmHg at echocardiography.
Methods: To evaluate the hemodynamic characteristics and pulmonary function in patients with LAM and PH not explained otherwise.
Results: Twenty patients were studied, with a mean age of 49±12 years. The median delay between the diagnosis of LAM and PH was 6.2 years. Dyspnea was NYHA class III in 10%, III in 50%, and IV in 46%. A single patient had right heart failure. Six minute walk distance was 340±84 m. Hemodynamic characteristics were: mean PA P2328 32.6±6 mmHg, cardiac index 3.5±1.1 L/min·m⁻², pulmonary vascular resistance (PVR) 376±184 dyn.s·cm⁻⁵, and pulmonary capillary wedge pressure 10±3 mmHg. The mean PAP was >35 mmHg in 3 cases. FVC was 76±28% of predicted, FEV1 42±15%, FEV1/FVC 74±15%, and DLco 29±13% vs 3.6±7%. 3 Pts on oral air. All patients were on long-term oxygen therapy. In five patients receiving bosentan, RHC after a median of 3.2 years demonstrated a median decrease of 42% in PVR. After a median follow-up of 2.9 years, one patient had died of cardiac arrest, and 5 patients had undergone lung transplantation.
Conclusion: Precapillary PH of moderate hemodynamic severity may occur in patients with LAM and severe pulmonary function impairment. Bosentan therapy might improve hemodynamic characteristics.
Support: CNMFR and FP7 of the European Commission.

P2329
Lung injury in intravenous drug users with infective endocarditis
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Aim and methods: To study lung injury and pulmonary hypertension (PH) in iv drug users with native infective endocarditis (IE) we have examined 30 pts (28 men, 2 women, 31.9±4.9 years, iv drug usage-2 to 8 years) and diagnosis of IE was according to Duke-criteria (ECS, 2009). Tricuspid valve localization of IE was revealed in 83% pts, tricuspid and aortic localization-14% and in 1 (3%)-isolated aortic IE. Co morbidity pathology included chronic HCV hepatitis in all pts and HIV infection in 5 pts.
Results: Lung injury was detected in 97%. X-ray revealed bi-lateral pleomorphic opacities in 16 pts, diffuse parenchymal lung injury in 13 pts including 3 pts with right atrium, right ventricle sizes (r=0.53, p<0.05; r=0.85, p<0.001; r=0.59, p<0.01, respectively). In pts with diffuse parenchymal lung disorders was found more prominent PH than in pts with infiltrative lung injury (59.9±1.1 vs 43.0±7.6, p<0.001). PASP was more (57.2±8.1) in pts with tricuspid vegetations larger than 1 cm, than in pts with small vegetations (less than 1 cm-45.5±10.7, p<0.05). The significantly decrease oxygen saturation was revealed in 14 pts with staphylococcal IE (90.4±6.8% vs 96.2±3.4% in other pts, p<0.01).
Conclusions: We have found out different patterns of lung injury in iv drug users with predominant tricuspid valve IE. Degrees of PH depend on lung involvement and valve’s vegetation sizes.

P2330
Endothelin dysfunction in systemic lupus erythematosus patients with pulmonary hypertension
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Endothelin dysfunction plays one of the most important parts in progress of cardio-vascular pathology in SLE.
Objective: To determine the state of endothelin dependent and endothelin independent regulation of vascular tone in patients with SLE-associated pulmonary arterial hypertension (SLE-PAH).
Methods: 80 patients with valid SLE diagnosis were examined with ultrasonographic scan of brachial artery to assess the endothelin dependent vasodilatation (EDWD) and endothelin independent vasodilatation (EIVD) according to reactive hyperemia test and test with nitroglycerine. PAH was manifested on the Doppler-echocardiogram in 35 SLE patients. The average duration of SLE in these patients was 14±1.6 years. A control group consisted of 20 healthy persons.
Results: There was no significant difference between the initial diameters of brachial artery in SLE (3.35±0.06 mm) and SLE-PAH groups (3.43±0.09 mm) compared with control group (3.46±0.07 mm). The significant reduction of EDWD on 34% in SLE patients compared to healthy persons was determined in the test with reactive hyperemia. Abnormalities of endothelium dependent vascular tone regulation in patients with PAH were more expressed (reduced on 48% compared with healthy persons) (p<0.05). At the same time EIVD in SLE group without PAH and in control group did not differ, but in SLE-PAH patients the EIVD was 1.6 times less compared with control.
Conclusion: Abnormality of EDWD is dominated in SLE patients. The most evident disorder of EDWD occurs in SLE patients during the PAH progress. In patients with PAH also noted reduction of vasoregulation function of smooth-muscular cells, because they had impaired EIVD.

P2331
Differences between limited and diffuse systemic sclerosis-related pulmonary arterial hypertension
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Background: Pulmonary arterial hypertension (PAH) is a leading cause of death in systemic sclerosis (SSc). Frequently associated with t limited SSc (lcSSc), PAH can also complicate diffuse SSc (dcSSc). Little is known about the characteristics of patients with dcSSc-PAH compared to lcssc-PAH.
Methods: Consecutive patients with SSc-PAH were followed prospectively. Lcssc and dcSSc patients were defined according to the American College of Rheumatology criteria. PAH was defined as a mPAP>25 mmHg with a PCWP <15 mmHg in the absence of significant ILD. Survival was assessed from time of diagnosis of PAH.
Results: 133 pts with SSc-PH were included, 112 (84%) had lcssc. Lcssc were more likely to be women (92% vs. 71%, p<0.001). dcSSc patients had a higher serum creatinine (1.0±0.4 mg/dL vs. 1.3±0.7, p=0.01) a higher proportion of hypertension (33% vs. 15%, p=0.05), and a shorter 6MWD (264±101 vs. 357±119m, p=0.02). Still, there were no significant differences in PFTs. Hemodynamics revealed less severe dcSSc in a lower mPAP (38±8 vs. 43±11 mmHg, p=0.08) and PCWP (6.8±3 vs. 9.4±6.2 Wood units, p=0.07). However, there was no difference in survival between the groups.
Conclusions: In this cohort of SSc-PH patients, 15% of subjects had dcSSc. While many characteristics were similar between the lcSSc and dcSSc groups, dcSSc patients were more likely to be men, have hypertension, renal insufficiency, and shorter 6MWD. Still, even though hemodynamics suggested less severe disease in dcSSc, survival did not differ between lcSSc and dcSSc. Whether this reflects intrinsic differences in the response to PAH between lcSSc and dcSSc remains to be determined and should be the focus of future investigation.

P2332
Prevalence of pulmonary arterial hypertension in hepatic liver transplant candidates in Santa Casa Hospital
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Background: Portopulmonary hypertension (PPH) is defined by the presence of pulmonary arterial hypertension, comprehending mean pulmonary artery pressure (mPAP) >25 mmHg at rest and pulmonary vascular resistance >120 dynes/cm²·m², at breath holding. \( PPH \) has been estimated to affect 1.5% of patients with liver disease, a prevalence of \( 2 \) in 95% in this studies.
Objectives: To estimate the prevalence and severity of pulmonary hypertension in patients with severe liver disease or portal hypertension.
Methods: Retrospective cross sectional review of 390 medical records regarding 375 patients with chronic liver disease undergoing liver transplantation at Santa Casa Hospital (Porto Alegre- Brazil), from February 2002 to April 2009. Fifteen patient records whose patients underwent retransplantation were excluded, as well as 31 patient records which lack information. The final sample had 344 patients. The present study was approved by the Hospital Ethics Committee. During the liver transplantation procedure, it was performed the pulmonary artery catheterization, and it has been made the measurement of the mPAP.
Results: The patients mean age was 52.9 years old (14-74) and male gender comprised 62.8% (n = 216) of this sample. The prevalence of PPH was 10.2% (n = 35). Seventy seven per cent (n = 27) were classified as having mild PPH (mPAP 25 - 35mmHg) and 25% (n = 8) as moderate (36-50mmHg). The mean mPAP was 20.7 mmHg.
Conclusion: The studied sample showed a slight increase in the prevalence of pulmonary hypertension when compared to former data in literature.
P2333
Pravastatin in patients with PAH: Results of a single-center study
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Background: The aim of the present study was to investigate the effect of pravastatin on cardiac structure and function in patients with PAH.

Methods: Twenty-four patients with PAH (age 34.6 ± 13.8 years, 10 females) were randomly assigned to receive either pravastatin (n = 12) or placebo (n = 12) for a period of 12 weeks. Cardiac structure and function were assessed using echocardiography at baseline and after 12 weeks of treatment.

Results: Pravastatin treatment was well tolerated and resulted in a significant decrease in left ventricular mass (−10.8 ± 4.1 g vs. +5.1 ± 3.2 g, p < 0.05) and in left atrial volume (−2.1 ± 1.3 mL vs. +1.1 ± 0.9 mL, p < 0.05). Additionally, pravastatin treatment led to a significant improvement in exercise capacity (6-minute walk test, 6MWT) (+51 ± 27 m vs. −12 ± 30 m, p < 0.05) and in quality of life as assessed by the SF-36 questionnaire (+5.7 ± 2.3 points vs. −0.8 ± 1.9 points, p < 0.05).

Conclusion: Pravastatin treatment is safe and effective in patients with PAH and has the potential to improve cardiac structure and function as well as exercise capacity and quality of life.
Survival and prognostic factors in patients with incident systemic sclerosis-associated pulmonary arterial hypertension from the French registry

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Background: Pulmonary arterial hypertension (PAH) is one of the leading causes of death in systemic sclerosis (SSc). Recently reported survival is better than in historical cohorts. However, no data on incident patients are available since 2006.

Aims and objectives: This study describes the characteristics and outcome of SSc patients enrolled in the multicentre French PAH registry since 2006.

Methods: SSc patients enrolled in the registry between January 2006 and November 2009 were prospectively included if they had PAH diagnosed within 1 yr prior to enrollment (incident patients). Patients with interstitial lung disease (ILD) were included if forced vital capacity (FVC)>70%.

Results: 91/145 SSc patients were included; 81% were in NYHA III or IV. Mean pulmonary arterial pressure (mPAP) was 40±10mm Hg, mean cardiac index was 2.6±0.8 L/min/m² and mean pulmonary vascular resistance (PVR) was 670±351 dyn sec cm⁻5. Overall survival was 90%, 76% and 54% at 1, 2 and 3 years, respectively. Male gender (HR: 2.44), age (HR: 1.044), desaturation after 6-minute walk test (HR: 0.93), PaO₂ (HR: 0.96) and cardiac index (HR: 0.52) were factors prognostic of survival. Other parameters did not reach statistical significance, including NYHA, PVR, 6MWD, BNP, DLCO/VA and mPAP.

Conclusions: These results confirm the poor prognosis for incident SSc-PAH patients even in the modern era. As with idiopathic PAH, gender and age appear to be important prognostic factors. Cardiac index, but not mPAP, had a significant impact on survival. For the first time, PaO₂ at rest and desaturation during exercise were shown to be significant prognostic factors in SSc-PAH.

Right axis deviation is a strong indicator of pulmonary hypertension in a risk population

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Pulmonary hypertension (PH) is diagnosed by right heart catheterization (RHC), but non-invasive methods play an important role in screening and follow-up. ECG is routinely performed in the clinical work-up but its clinical value has not been defined. We hypothesized that a simple parameter, such as the presence of right axis deviation (RAD) in standard ECG might be useful in the diagnosis and follow-up of PH.

We retrospectively analysed all patients who received a RHC and an ECG in our department between 2005 and 2010. The indication for RHC was heterogeneous, the pretest probability of PH was about 50%. We determined the ratio of the S and R waves in lead I in each patient, a value ≥1 (>90°) was considered as RAD. The investigator was blinded to the RHC results. The sensitivity and specificity of RAD to predict PH (mean PAP≥25mmHg) was determined.

N=317 patients were included. RAD was present in n=71 patients. Within these, PH was detected in n=65 patients, and was missing in n=6 subjects (p<0.001). In n=246 patients without RAD, PH was present in n=87 patients. The specificity of RAD for PH was 96%, whereas the sensitivity was merely 43%. Accordingly, the positive predictive value of RAD to predict PH was 92%, while its negative predictive value to rule out PH was just 35%.

Our retrospective analysis on a large, heterogeneous cohort of subjects including patients with and without PH suggests that the presence of RAD in the ECG has a high specificity but a low sensitivity for PH. This simple method may help to recognize PH patients within a population at risk for PH. In order to determine its appropriate role in a diagnostic algorithm, prospective, population based studies are needed.