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. All patients were asymptomatic for any pulmonary manifestations. 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 anticoagulants. However, plain 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 only patients.
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 aphtosis 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 telethorax 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 disease that makes its prognosis dull.

P2322

Prevalence of hepatopulmonary syndrome in candidates for liver transplantation in Santa Casa Hospital (Porto Alegre-Brazil)

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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) $\geq$15 mmHg on room air, and in larger 64 years PaO2 $<$70 mmHg or PA-aO2 $\geq$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, strengthen 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 102 (75%) 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 EchoCG 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%), 2d 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 49,3±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

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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".

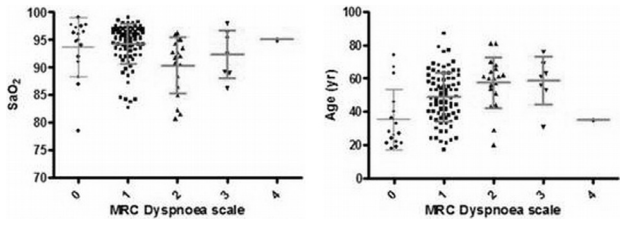

Figure 1. Error bars represent mean and standard deviation.

Results: 44 (27%) patients were dyspnoeic (24% excluding the 5 with severe coexistent respiratory disease). In univariate analyses, oxygen saturations (SpO_2), 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 SpO_2 to dyspnoea grade, although age and SpO_2 did not account for most of the variation (r^2=19%).

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

P2326

The correlation between hepatopulmonary syndrome and cirrhosis etiological diagnosis in candidates for liver transplantation at Santa Casa Hospital

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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 patients 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), from February 2002 to April 2009. The present 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

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Background: The 55-center, observational, US Registry to EValuate Early And Long-term pulmonary arterial hypertension (PAH) 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 demographics, 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 years and 9±6 years (P = 0.002), respectively. APAH-CHD had similar functional class (FC) at diagnosis compared with IPAH/FPAH (FC: I, 5%; II, 51%; III, 32%; and IV, 11% vs FC: I, 7%; II, 40%; III, 43%; and IV, 10%, respectively; P = 0.64). Hemodynamics were also similar at diagnosis. Similar proportions were treated with endothelin receptor antagonists (APAH-CHD, 40%; IPAH/FPAH, 45%; P = 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±14% 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.

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 6.7% in 95 patients with lymphangioleiomyomatosis (LAM) as defined by sPAP>35 mmHg at echocardiography.
Objective: To evaluate the hemodynamic characteristics and pulmonary function in patients with LAM and PH not explained otherwise.
Methods: A retrospective multicenter study was conducted in patients with LAM with precapillary PH (mean PAP ≥25 mmHg, pulmonary artery wedge pressure <15 mmHg, and normal or decreased cardiac index at RHC).
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 I/II in 10%, III in 50%, and IV in 40%. A single patient had right heart failure. Six minute walk distance was 340±84 m. Hemodynamic characteristics were: mean PAP 32±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±25%, FEV1/FVC 47±15%, and DLco 29±13%. PaO2 was 7.4±1.3 kPa on room 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: CNMR 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 i/v drug users with native valves infective endocarditis (IE) we have examined 30 pts (28 men, 2 women, 31,9±4,9 years, i/v drug usage-2 to 8 years). 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 morbid 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 polysegmental opacities in 16 pts, diffuse parenchymal lung injury in 13 pts including 3 pts with HIV. Tuberculosis was negative in all cases. PH was revealed in all pts (mean pulmonary artery systolic pressure (PASP) 47,6±11,7 mmHg). Positive correlations were found between fever duration and PASP, 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±7,1 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±8,6% vs 96,2±3,4% in other pts, p<0,01).
Conclusions: We have found out different patterns of lung injury in i/v drug users with predominant tricuspid valve IE. Degrees of PH depend on lung involvement and valve's vegetation sizes.

P2330

Endothelium dysfunction in systemic lupus erythematosus patients with pulmonary hypertension

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Endothelium dysfunction plays one of the most important parts in progress of cardio-vascular pathology in SLE.
Objective: To determine the state of endothelium dependent and endotheliun 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 asses the endothelium dependent vasodilatation (EDVD) and endothelium independent vasodilatation (EIDVD) 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 not 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 EDVD 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 EIDVD in SLE group without PAH and in control group did not differ, but in SLE-PAH patients the EIDVD was 1.6 times less compared with control.
Conclusion: Abnormality of EDVD is dominated in SLE patients. The most evident disorder of EDVD 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 impared EIDVD.

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 were defined according to the American College of Rheumatology criteria. PAH was defined as a mPAP> 25mmHg with a PCWP ≤ 15 mmHg in the absence of significant ILD. Survival was assessed from time of diagnosis of PAH.
Results: 133 with SSc-PAH were included; 112 (84%) had lcSSc. LcSSc were more likely to be women (92% vs. 71%, p<0.01). 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 hyponatremia (33% vs. 15%, p=0.05), and a shorter 6MWD (264±101 vs. 357±119m, p=0.02), but there were no significant differences in PFTs. Hemodynamics revealed less severe PAH in dcSSc with a lower mPAP (38±8 vs. 43±13mmHg, p=0.08) and PVR (6.8±3.6 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-PAH 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 hyponatremia, 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 dinas/seg/cm5, in association with severe liver disease or portal hypertension. PPH has important implications for the prognosis of chronic liver disease patients. The prevalence of PPH is conflicting in medical literature, ranging from 2 to 9% in larger 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 23% (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

Praevalence of pulmonary arterial hypertension in the HIV cohort of the University Bonn: Results of the PAHIBO study

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Background: PAH (pulmonary arterial hypertension) is a rare and life-threatening complication of HIV (human immunodeficiency virus) infection. An investigation of the presence of HIV infection is a standard diagnosis in patients with unexplained PAH. Previous studies suggest a prevalence of PAH in HIV patients close to about 0.5%, but are limited in the study design. Therefore the exact prevalence is unclear.
Methods: The PAHIBO study is investigating all HIV patients (n = 700) at the University Hospital of Bonn in a prospective cross-sectional study. If systolic pulmonary arterial pressure (sPAP) is > 35 mmHg in Doppler echocardiography, right heart catheterization is followed.
Results: To date, 340 patients were enrolled, 44/340 (13%) had an echocardiographic sPAP > 35 mm Hg. The right heart catheterization was carried out so far in 22/44 patients with echocardiographically proven PH. In 4 cases, a precapillary PAH (including a complete work-up and exclusion of other causes of PAH) was diagnosed, of which 1 case was already known, in 13 cases postcapillary PH and in the remaining 5 cases, the exclusion of a manifest PH. Thus, the PAHIBO study reveals a prevalence of HIV-associated PAH of at least 4/340 cases (1.2%).
Conclusion: The prevalence of HIV-associated PAH is probably higher than previously described. Severe cardiac and respiratory comorbidities are very frequent in the examined HIV cohort. If these results should be confirmed in the extended cohort of 700 patients, a regular echocardiographic screening in asymptomatic HIV patients is to discuss.

P2334

Characteristics of PAH associated with pretricuspid shunts in the French PAH registry

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The diagnosis of pulmonary arterial hypertension (PAH) associated with pretricuspid shunts (PTS) is often made in adulthood. The characteristics of this subgroup of CHD-PAH are not well established.
Objective: To review and analyze patients with PAH and PTS enrolled in the second prospective French PAH registry, initiated in 2006.
Results: CHD-PAH (n=255) accounted for 9.8% of PAH. 100 patients had PTS: atrial septal defects (ASD) ostium secundum type (n=93), ASD sinus venosus type (n=3), ASD ostium primum type (n=1), and partial abnormal pulmonary venous return without ASD (n=3). 72% of patients were female. At diagnosis, mean age was 38 years (range, 0-81 years). 17 patients were diagnosed before 18 years and 28 after 60 years. In 15 patients CHD was repaired before PAH diagnosis. In patients with open shunts, PAH diagnosis was made at time of diagnosis of CHD in 56% of cases, whereas the diagnosis of CHD followed the diagnosis of PAH in 16% of cases. PAH appeared during the follow-up of the CHD in 28% of cases. At inclusion, 5% of patients were in functional class (FC) IV, 61% were in FC III and 34% in FC I or II. 6MWD was 360±107m. Pulmonary hemodynamics were: mPAP=53±16mmHg, CO=5.02±1.7L/min and PVR=11±8.3WU. 50% of patients in FC II or III received PAH specific therapy. During the 3-year follow-up period, 9 patients died and 3 underwent heart-lung transplantation.
Conclusion: In this registry, the proportion of CHD-PAH due to PTS compared to other forms of CHD is higher than in previously reported series. The natural history of PTS remains poorly understood as PAH can be diagnosed throughout life. PAH specific therapies are not widely used in the PAH associated with PTS population.

P2335

Health-related quality of life in patients with pulmonary arterial hypertension

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Background: The aims of the present study were (1) to investigate global and disease specific health-related quality of life (HRQOL) in individuals with Pulmonary Arterial Hypertension (PAH), (2) to examine the correlation between HRQOL and exercise capacity (6-minute walk test, 6MWT) and (3) to compare patientsratings with the age-matched general German population.
Methods: 63 consecutive patients (DANA-Point 1) of our pulmonary hypertension program (age 57±17.1 years; 33 male [53%]; 21 [34%] combination therapy; BMI 24.4±4.6) were evaluated by the 6-minute walk test (6MWT), the standardized global SF-36 questionnaire and the specific "Cambridge Pulmonary Hypertension Outcome Review" (CAMPHOR).
Results: PAH patients were found to have a reduced HRQOL in physical SF-36 domains. In contrast, the mental health of our PH patients is excellent with SF-36 subscales reaching best possible scores. CAMPHOR "quality of life" score reached the upper tertile, whereas "symptoms" and "activity" scores were in the middle tertile, indicating a reduced physical performance. No significant correlations were found between the 6MWT (401±114 meters) and HRQOL scales. Compared to the age-matched norm population, HRQOL self-ratings were significant reduced in six of eight SF-36 subscales ($p \leq 0.01$) except for "pain" (p=0.300) and "mental health" (p=0.093).
Conclusion: The study results suggest especially a physical HRQOL limitation for PAH patients. The 6MWT might not be a sufficient surrogate marker for HRQOL domains. HRQOL adds important patient sensitive information beyond the 6MWT and should be considered for clinical study design.

P2336

Systemic sclerosis associated with pulmonary arterial hypertension is most severe in African American patients

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Background: African Americans (AA) with Systemic Sclerosis (SSc) have a lower age at disease onset, a higher frequency of diffuse (vs. limited) skin involvement, and an overall worse prognosis compared to other ethnic groups.
Objective: To estimate the differences in hemodynamics, clinical presentation and survival between AA and Caucasians with SSc associated pulmonary arterial hypertension (SSc-PAH).
Methods: We conducted a study of 75 consecutive SSc patients diagnosed with PAH by heart catheterization (mPAP≥25mmHg and PCWP≤ 15mmHg) at a single center. Kaplan Meier estimates were calculated and mortality risk factors were analyzed.
Results: The cohort included 9 AA and 66 Caucasians. Forty one (54.7%) patients were in WHO functional class III or IV. There was no difference in age between the 2 groups (59±13 vs. 61±11 years P=0.7), or disease duration (11±11 vs. 12.7±13 years, P=0.7). AA patients had a higher prevalence of systemic hypertension (66.7% vs. 30.3%; P=0.03), higher mPAP (51±12 vs. 41.8±11 mmHg; P=0.02) and PVR (14.8±9.5 vs. 8.04±4.6 Wood units; P=0.01), and lower SVI (24±10.5 vs. 32±10 mL/m^2, P=0.054), but similar PCWP (11.6±2.6 vs. 10.18±3.4 mmHg, P=0.25). Renal dysfunction (defined as eGFR <60) (33.3% vs. 45.5%; P=0.49), extent of disease (88.9% had limited disease vs. 89.2%; P=0.9) and treatment between AA patients and Caucasians were not different. Median survival did not differ between the two groups (3.2 vs. 4.1 years, P=0.4).
Conclusions: SSc-PAH is significantly more severe in AA compared to Caucasians and is characterized by right ventricular dysfunction.

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Clinical characteristics of the pulmonary hypertension (PH) in a cardiologist service

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Introduction: Although many epidemiological data of primary Pulmonary Hypertension (PH) have been recorded, the literature as regards the characteristics of the general population suffering from PH is nevertheless scarce.
Aim: To determine the clinical characteristics of the different types of PH in a cardiologist service of a general hospital patients.
Results: 204 out of the 321 patients evaluated from January 1994 to December 2009 met the inclusion criteria. The average age was 73,36 (±17.48) years old, 55% of whom were women. The average initial PSAP was 74±9.2 mmHg. The predominant type found was PH owing to left heart disease 88 cases (43,13%), the most common etiology being mitral valve pathology (53/87). Pulmonary arterial hypertension (PAH) represented 23% of the overall, there being ten cases of left-right shunts, four cases of primary PH, a case of scleroderma, another one of anorectics and two cases of portal hypertension. In eleven cases (13%) PH was associated with respiratory disease while the remaining 13% corresponded to PH caused by chronic thrombosis and/or pulmonary embolism. The average age of patients with PAH was lower (60 vs. 70). The predominant sex in all types of PH was female (2:1) except for those of systemic shunts, where no differences were found (1:1), and those owing to lung disease, with a male predominance (1:4.5).
Conclusions: 1. Half of the recorded cases in our series correspond to PH owing to heart disease, mainly related to rheumatic valve disease. 2. The age of our patients was high, being significantly lower in those cases of PAH. 3. Female predominance was to be found in all cases, except for the cor pulmonale with a majority of male patients.

P2338

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/m^2 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.

P2339

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, heterogenous 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.