## P320 | BEDSIDE
### Prognostic value of C-reactive protein in patients with Eisenmenger syndrome

G. Scognamiglio, A. Kempny, R. Alonso-Gonzalez, R. Agra-Bermejo, A. Uebing, P. Marino, L. Swan, M.A. Gatzoulis, K. Dimopoulos, S.J. Wort. *Royal Brompton Hospital and Imperial College London, National Heart and Lung Institute (NHLI), London, United Kingdom*

**Background:** Eisenmenger syndrome (ES) represents the extreme manifestation of pulmonary arterial hypertension in patients with congenital heart disease and is associated with substantial morbidity and mortality. Although C-reactive protein (CRP) is known to predict outcome in iPAH, little is known on its prognostic value in ES.
**Methods and results:** We included 1936 CRP measurements performed in a total of 225 adult ES patients (age 40.7±12.4 years, 33% male, 35% with Down syndrome) between years 2000 and 2012. High CRP values related to infection or blood transfusions were excluded from the analysis. During a median follow-up of 4.8 years (1149 patients-years) 50 patients died. The median CRP level was 6.0mg/L [IQR 2.0-10.0] and was higher on the last assessment in deceased patients compared to survivors (11.5mg/L [6.0-22.8] vs. 4.0mg/L [1.5-8.0], P<0.0001). On Cox regression analysis, CRP emerged as a strong predictor of mortality (HR=1.25, 95% CI 1.16-1.34, P<0.0001) and remained significant after adjustment for age, presence of Down syndrome and advanced therapies for PAH. Survival-ROC analysis identified an optimal cut-off value of 10mg/L. Patients with CRP>10mg/L had more than 3-fold higher risk of mortality (HR=3.66, 95% CI 1.98-6.80, P<0.0001, Figure).

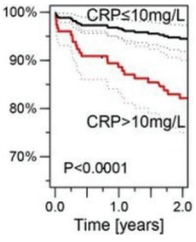

Figure 1

**Conclusions:** Serum CRP is a simple, but powerful marker of mortality in ES patients and should be incorporated in the risk stratification and routine assessment of these patients.

## P321 | BEDSIDE
### Absence of acute vasoreactivity response in patients with pulmonary arterial hypertension associated with toxic oil syndrome

E. Barrios[1], M.A. Gomez-Sanchez[1], C. Hernandez[2], M.T. Velazquez[3], N. Ochoa[1], J.M. Montero[4], J. Delgado[1], M.J. Ruiz-Cano[1], P. Escribano[1], J. Tascon[3]. *[1]University Hospital 12 de Octubre, Department of Heart Transplant and Pulmonary Hypertension, Madrid, Spain; [2]University of Las Palmas de Gran Canaria, Las Palmas de Gran Canaria, Spain; [3]University Hospital 12 de Octubre, Department of Hemodynamics, Madrid, Spain; [4]University Hospital 12 de Octubre, Department of Cardiology, Madrid, Spain*

**Purpose:** Acute vasodilator testing (AVT) is important in patients with pulmonary arterial hypertension (PAH). Positive test predicts a better survival and response to calcium channel blockers therapy. The last guidelines recommend performing this test in patients with PAH, but its usefulness in different forms than idiopathic PAH is unclear. The aim of this study is to analyze the results in AVT in patients with PAH associated with toxic oil syndrome (TOS).
**Results:** From 1993 to 2011 have been diagnosed in our hospital 32 late cases patients with PAH associated with TOS and have done an acute vasodilator test in 24 of them, in all cases the result was negative. The causes were analyzed as hemodynamic severity of disease at diagnosis, mutations in the BMPR2 gene or different histological changes. When comparing the results of right heart catheterization at the time of diagnosis among our population and a group of 52 patients with idiopathic PAH diagnosed in the same period, found no statistically significant differences but a tendency to greater severity in terms of mean RA pressure (9.8±5.9 vs 8.5±5.7 mmHg, p = 0.356), CO (3.8±1.2 vs 4.2±1, 4 lpm, P = 0.178) and PVR (17.2±9.4 vs 13.5±5.6 WU, p = 0.249). BMPR2 mutations in this population were not found vs 8 (15.4%) in iPAH, p= 0,048. A micromorphometric study of pulmonary vessels, in 4 TOS patients, showed that there was a marked venular component with intimal fibrosis, presence of double elastic membrane and lumen obliteration, in addition to plexiform arteriopathy (Fig 1).
**Conclusions:** A negative AVT in patients with PAH associated with TOS may be due to more severe hemodynamic disease with additional venules involvement. Additional studies are needed to correlate the absence of BMPR2 mutation and lack of vasoreactivity.

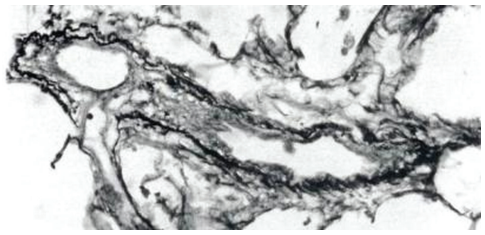
Figure 1. Study of pulmonary venules

## P322 | BENCH
### Pulmonary artery size and intrinsic wall properties in patients with pulmonary hypertension

C.-D. Botezatu[1], R. Enache[2], O. Nastase[1], B.A. Popescu[2], M. Rosca[2], C. Beladan[2], A. Calin[2], M.M. Gurzun[1], C. Ginghina[2]. *[1]Institute of Emergency for Cardiovascular Diseases "Prof. Dr. C.C.Iliescu", Bucharest, Romania; [2]University of Medicine Carol Davila, Institute for Cardiovascular Diseases Prof C.C. Iliescu, Bucharest, Romania*

**Background:** Pulmonary artery (PA) dilation is a common finding in pulmonary hypertension (PH). The relation between the degree of PA dilation, PH severity and PA stiffness (PAS) was not fully investigated so far.
**Purpose:** To assess the impact of PAS parameters and right ventricular (RV) pressure and volume overload on PA dilation in patients with PH.
**Methods:** We enrolled 43 consecutive patients (13 had congenital heart disease, 21 idiopathic PH, 9 left heart disease) with PH (mean PA pressure >25 mmHg) and 19 age- and gender-matched controls (normal PA pressure, no right chambers dilation or dysfunction). All subjects underwent transthoracic echocardiography. Longitudinal and transversal end-systolic (ES) and end-diastolic (ED) PA trunk diameters (D) were measured from the parasternal short axis view. According to PAEDD, PH patients were divided into 3 groups: 13 had normal or mildly (EDD <26 mm), 15 moderately (EDD 26-29 mm) and 15 severely dilated PA (EDD >29 mm). Systolic, end-diastolic and mean PA pressures were estimated using the tricuspid and pulmonary regurgitant jet velocities on CW Doppler envelopes. Parameters of PAS were assessed: compliance, distensibility, elastic modulus, stiffness index β. RV function was also evaluated.
**Results:** PH patients (49±18 years, 17 men) had significantly higher transversal and longitudinal PAEDD than controls (42±16 years, 10 men): 28.6±8.4 vs 19.1±1.8 mm and 57.3±11.1 vs 45.2±5,6 mm, respectively, both p<0.001. Both transversal and longitudinal PAEDD progressively increased across the 3 groups (p<0.001 and p=0.004, respectively) and so did PAS (PA strain, p<0.001 and PA dynamic compliance, p=0.005). Transversal PAEDD correlated significantly with PAS parameters (PA strain: r=-0.61, p<0.001; PA dynamic compliance: r-0.50, p=0.001; stiffness index β: r=0.52, p<0.001; elastic modulus: r=0.62, p<0.001; distensibility: r=-0.42, p=0.005), independent of PH etiology. We found no correlation between systolic, mean or end-diastolic pulmonary pressures and PA dilation in patients with PH. PAS did not correlate with TAPSE or S wave velocity at the lateral tricuspid annulus. Instead, a global RV function parameter (I/H) was significantly related to PAS (stiffness index β: r=0.33, p=0.036; elastic modulus: r=0.46, p=0.003).
**Conclusion:** Patients with PH have dilated PA and increased PAS, which is related to load-independent structural changes in the vascular wall in this setting. Moreover, PA dilates not only in its transverse axis but also in the longitudinal one. The clinical impact and the prognostic role of PA elongation in PH patients need further studies.

## P323 | BEDSIDE
### Comparison of hemodynamic parameters in treatment-naive and pretreated patients with pulmonary arterial hypertension (PAH) in the Phase III PATENT-1 study

Z.-C. Jing[1], N. Galie[2], H.-A. Ghofrani[3], M. Humbert[4], D. Langleben[5], L.J. Rubin[6], M.M. Hoeper[7], A. Fritsch[8], N. Davie[8], A.M. Keogh[9]. *[1]Shanghai Pulmonary Hospital, Tongji University School of Medicine, Shanghai, China, People's Republic of; [2]Institute of Cardiology, University of Bologna, Bologna, Italy; [3]University of Giessen and Marburg Lung Center (UGMLC), member of the German Center of Lung Research, Giessen, Germany; [4]Univ. Paris-Sud; Inserm U999; AP-HP, Hôpital Bicêtre, Service de Pneumologie, Le Kremlin-Bicêtre, France; [5]Lady Davis Institute for Medical Research, Jewish General Hospital, McGill University, Montreal, Canada; [6]University of California, San Diego, La Jolla, United States of America; [7]Clinic for Respiratory Medicine, Hannover Medical School, Hannover, Germany; [8]Global Clinical Development, Bayer HealthCare AG, Wuppertal, Germany; [9]St Vincent's Hospital, Sydney, Australia*

**Purpose:** In PATENT-1, treatment with riociguat, a novel soluble guanylate cyclase stimulator, significantly improved 6-min walking distance (6MWD) and hemodynamic parameters in patients with PAH compared with placebo. Here we present the detailed hemodynamic findings from PATENT-1 in treatment-naïve and pretreated patients.
**Methods:** In this Phase III, double-blind, placebo-controlled study, patients were randomized 2:4:1 to treatment with placebo, individually titrated riociguat (up to 2.5 mg tid), or a capped titration of riociguat (up to 1.5 mg tid; exploratory, ana-