with a high transpulmonary gradient pressure, systolic pulmonary arterial pressure 85 mmHg, medium pulmonary arterial pressure 53 mmHg, pulmonary arterial wedge pressure 12 mmHg and pulmonary vascular resistance 9,56WU. Vasoreactivity test with adenosine was negative. Pulmonary arteriography detected diffuse flow amputation of peripheral pulmonary arteries without thrombus. An extended workout excluded other causes of pulmonary hypertension. Oral anticoagulant, diuretic and specific pulmonary vasodilator drugs were recommended. The patient improved the functional class and remains stable. **Conclusion:** Actinic pulmonary endarteritis is a possible cause of pulmonary hypertension in patients submitted to thoracic radiotherapy in past.

PREVALENCE OF PULMONARY CIRCULATION ABNORMALITIES IN LIVER TRANSPLANTATION CANDIDATES

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Background: The early identification of abnormalities in the pulmonary circulation is essential in patients with end-stage liver disease. The most important findings in this setting are the hepatopulmonary syndrome (HPS) and the portopulmonary hypertension (PPH). **Purpose:** To determine the prevalence of HPS and PPH in hepatic transplantation candidates. **Methods:** In a retrospective fashion, we evaluated all patients with advanced liver disease (liver transplantation candidates) that have been evaluated in pulmonary circulation outpatient clinic, from January 2005 to December 2007. Inclusion criteria were performed blood gas analysis and transthoracic Doppler ecochardiography. We used European Respiratory Society Task Force on Pulmonary-Hepatic Vascular Disorders diagnostic criteria to HPS and PPH. **Results:** A total of 140 cirrhotic patients were included in our study; 88 males (62.9%), mean age of 54.8 years (SD $\pm$ 11.5, range 21 to 74 years). Main arterial blood analysis data were PaO_2 94.5 mm Hg (SD$\pm$ 15.3, range 55 to 128.7 mm Hg) and Alveolar-arterial gradient 12.9 mm Hg (SD $\pm$ 11.7, range 0 to 50.7 mm Hg). Doppler echocardiography detected intrapulmonary shunt in 104 patients (75.7%). However, hepatopulmonary syndrome was diagnosed in only 47 patients (33.6%). Right ventricle systolic pressure was estimated in only 80 patients (mean 33.5 $\pm$ 9.5 mm Hg, range 20 to 79 mm Hg). Seven patients (5%) had signs of pulmonary hypertension on Doppler echocardiography. Right heart catheterization was performed in 10 patients: no PH in 6 patients, high flow circulatory status in 3 patients, and PPH in 1 patient. True prevalence of PPH in this study was 0.7%. **Conclusions:** Hepatopulmonary syndrome is a common pulmonary circulation abnormality in liver transplant candidates. Further, pulmonary hypertension is uncommon condition, and hyperdynamic circulatory state is more frequent hemodynamic pattern than portopulmonary hypertension.

COMPARISON BETWEEN TRANSTHORACIC DOPPLER ECHOCARDIOGRAPHY AND RIGHT HEART CATHETERIZATION FOR PULMONARY HYPERTENSION EVALUATION

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Background: Pulmonary hypertension (PH) is either an idiopathic condition or can be associated with several pathologies. Transthoracic Doppler echocardiography (D-echo) is a screening test for PH and has a good accuracy in this setting. However, this is an operator-depending method having as a drawback potential disagreement with the right heart catheterization (RHC) findings. **Purpose:** To compare Doppler echocardiography and right heart catheterization data in a group of patients with a clinical suspicion of PH. **Methods:** In a retrospective fashion, we evaluated all patients that underwent to transthoracic D-echo and RHC to investigate PH from January 2005 to December 2007. Pulmonary hypertension was diagnosed by RHC (gold standard method) if mean pulmonary arterial pressure (mPAP) was more than 25 mm Hg. To calculate accuracy of D-echo, we use tricuspid regurgitation velocity (TRV) $\geq$ 2,8 m/s to define PH case. **Results:** Eighty one patients were included, 45 women (55.5%), mean age of 51.3 years (SD $\pm$ 15.9; range 16 to 78 years). Main etiologies of PH were thromboembolic disease (n=18, 22.2%) and lung disease (n=12, 14.8%). Pulmonary hypertension prevalence in this group was 76.5%. No major complications were reported in RHC. Doppler echocardiography sensitivity, specificity, positive and negative predictive values were, respectively, 84.1%, 50.0%, 85.5% and 47.4%. All false-negative D-echo were mild PH cases by RHC (range mPAP 26 to 38 mm Hg). **Conclusions:** Right heart catheterization is a major step during the evaluation process of patients with pulmonary hypertension previously evaluated with transthoracic Doppler echocardiography. It is a safe method, with no serious complication in this series. Further, RHC avoids many incorrect diagnosis of pulmonary hypertension, and describes the hemodynamic profile and severity of PH.

PULMONARY THROMBOENDARTERECTOMY IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: INITIAL EXPERIENCE OF HOSPITAL DE CLINICAS DE PORTO ALEGRE

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Background: Pulmonary thromboendarterectomy (PTE) is the main therapy for Chronic Thromboembolic Pulmonary Hypertension (CTPH). It\'s a major surgery perfomed in a few medical centers. **Purpose:** To describe the initial experience of Pulmonary Circulation Group of Hospital de Clinicas de Porto Alegre in perform PTE in CTPH. **Methods** This a prospective case series of patients that underwent to pulmonary thromboendarterectomy between February 2002 to July 2006. We review all medical records and test results. **Results:** Nine patients were submitted to pulmonary thromboendarterectomy; mean age of 36,8 years-old (SD ±14,3 years-old; rande 22 to 70 years-old); 5 males (55%). Preoperative estimated pulmonary systolic arterial pressure (PSAP) by ecochardiogram was 76,6 mmHg (SD± 23,7 mmHg; rande 48 to 116 mmHg). Surgical classification of CTPH was type 1 in 5 patients (56%), type 2 in 3 patients (33%) and type 3 in 1 patient (11%). All procedures did with extracorporeal circulation and total circulatory arrest. Mean length of stay in hospital was 39,1 days (SD ± 17,4 days; range 18 to 75 days). Complications reported were reperfusion lung edema, pneumonia and ischemic stroke. One death occurs in patient with type 3 CTPH in return of extracorporeal circulation due to right ventricle cardiogenic shock (mortality 11%). Functional class and symptoms improved ($p< 0,05$). After 3 months of surgery, new echocardiogram detected significant hemodynamic improve (PSAP 34,6 ± 6,7 mmHg; rande 25 to 44 mmHg) ($p< 0,05$). **Conclusion:** Pulmonary thromboendarterectomy is an effective therapy to patients with Chronic Thromboembolic Pulmonary Hypertension, resulting in better functional class and hemodynamic condition, with acceptable mortality in initial experience of Pulmonary Circulation Group of Hospital de Clínicas de Porto Alegre.

REAL TIME BIOTELEMETRY APPLIED TO THE SIX-MINUTE WALKING TEST IN PATIENTS WITH PULMONARY HYPERTENSION

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Background: The six-minute walk test (6MWT) is a submaximal exercise test that can be performed by a patient incapable to do a maximal exercise tests. The test is very simple and reproductible. It was demonstrated that the distance walked in 6 minutes was independently related to mortality in patients with primary pulmonary hypertension, and that patients walking less than 332 m had a significantly lower survival rate. Real time biotelemetry applied to exercise test was not studied in patients with pulmonary arterial hypertension (PAH). **Purpose:** The aim of this study was to describe this new technology to do 6MWT in these patients. **Methods:** We conducted a review of the medical records of 41 patients with PAH who underwent 6MWT in our institution. Patients with PAH associated to COPD were excluded. PAH was defined as an estimated systolic pulmonary arterial pressure by echococardiogram greater than 35 mmHg. The test was carried out in a 27-meters corridor and was coaching by a clinical nurse or a physician. Heart rate (HR) and pulse oximetry (SpO_2) were constantly monitored by means of real time biotelemetry. **Results:** Between October 2004 and December 2006, real time biotelemetry 6MWT was performed in 937 patients.. Fifty-two tests were performed in 41 patients with PAH (27 men; mean age, 52 years). Main diagnosis was CTPH in 15 patients, idiopathic pulmonary fibrosis in 7, diffuse connective disease in 3, pneumoconiosis in 3 and 13 in others. The mean distance walked was 366 m (SE± 140 m). Thirty six patients (69,2%) had a significant desaturation (³ 4%). The mean systolic pulmonary artery pressure measured by echocardiography was 59,3 ± 23,2 mmHg. Thirty per cent of the patients had PaO_2 at rest < 70 mmHg. No significant complications were detected during the real time biotelemetry 6MWT. **Conclusions:** The use of real time biotelemetry permits that heart rate and pulse oximetry were precisely and constantly monitored throughout the 6-minute walking test

PREVALENCE OF PULMONARY ARTERIAL HYPERTENSION USING DOPPPLER ECHOCARDIOGRAPHIC METHOD IN SICKLE CELL DISEASE

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Background: Pulmonary arterial hypertension (PAH) in sickle cell disease is considered to be directly related to hemolysis events. The current knowledge about its pathophysiology focus on the decreased bioavailability of the nitric oxide through the plasmatic arginase route. PAH in these patients has been related to a poor prognosis and an overall early mortality. Therefore, routine screening for PAH is recommended. **Purpose**: To determine the prevalence of PAH by doppler echocardiogram in a group of patients with sickle cell anemia. **Methods:** In a retrospective fashion, we evaluated the doppler transthoracic echocardiogram findings of 16 patients with sickle cell disease from our outpatient pulmonary circulation clinic over 18 months (January/2005 to July/2006). PAH was defined as an estimated of a systolic pulmonary arterial pressure ≥ 35mmHg. **Results:** The study group was composed of six men (37.5%); age 30.2 ± 14 years (mean ± SD).