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Background: Pulmonary thromboendarterectomy (PTE) is the main therapy for Chronic Thromboembolic Pulmonary Hypertension (CTPH). It's a major surgery performed in a few medical centers. **Purpose:** To describe the initial experience of Pulmonary Circulation Group of Hospital de Clínicas 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 \pm 14,3 years-old; range 22 to 70 years-old); 5 males (55%). Preoperative estimated pulmonary systolic arterial pressure (PSAP) by echocardiogram was 76,6 mmHg (SD \pm 23,7 mmHg; range 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 \pm 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 $34,6 \pm 6,7$ mmHg; range 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 reproducible. 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 echocardiogram 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 (SpO₂) 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 \pm 140 m). Thirty six patients (69,2%) had a significant desaturation ($\geq 4\%$). The mean systolic pulmonary artery pressure measured by echocardiography was $59,3 \pm 23,2$ mmHg. Thirty per cent of the patients had PaO₂ 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 DOPPLER 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 ≥ 35 mmHg. **Results:** The study group was composed of six men (37.5%); age 30.2 ± 14 years (mean \pm SD).