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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).

Diameter of the right ventricle, left atrial size and ejection fraction had means of 2.14cm, 3.66cm and 67.18% respectively. Six out 16 patients presented abnormalities in the left ventricle: restriction to relaxation (2 pts.) and increase of the left ventricle mass (5 pts.). One patient had a mitral prolapse and other a mild aortic insufficiency. The prevalence of PAH was 19%. **Conclusions:** The prevalence of PAH in patients with sickle cell disease was 19%. This result is lower than reported on previous studies and may be related to small sample and new age group. This is an expanding field.

IN-HOSPITAL MORTALITY OF ACUTE PULMONARY THROMBOEMBOLISM: COMPARISON BETWEEN PATIENTS WITH OBJECTIVE DIAGNOSIS AND NON-CONFIRMED SUSPICION

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Background: Pulmonary thromboembolism (PE) is frequent in hospital setting. Follow-up of patients with non-confirmed PE is unknown. **Purpose:** To compare mortality between patients with confirmed PE and non-confirmed suspicion. **Methods:** Historical cohort. Included adult patients (³18 years) with suspicion of PE identified by perfusion lung scan, CT-angiography, pulmonary angiography or PE ICD-9/ICD-10 on admission or discharge charts. We excluded patients with incomplete or lost medical records. **Results:** Of 741 patients, 687 were included. Mean age was 61.53 ± 16.75 years, men 292 (42,5%). Primary PE (beginning before admission) in 330 cases (48%) and secondary PE (beginning after admission) in 357 (52%). In 120 patients (17.5%), PE was objectively confirmed, in 193 (28.1%) was objectively excluded, but in 374 cases (54.4%) diagnostic approach was incomplete. In-hospital mortality was 19.1% (n=143). In multivariate analysis, hypotension (beta 2,49, IC95% 1,35-4,63), PE objectively confirmed (beta 2,199, IC95% 1,15-4,21), non-conclusive approach (beta 1,70, IC95% 1 – 2,87), cancer (beta 2,868, IC95% 1,80-4,45), secondary PE (beta 1,57, IC95% 1,02-2,41), inclusion in 1996-1997 (beta 1,71, IC95% 1,15-2,67) and thoracic or abdominal infection (beta 1,71, IC95% 1,08-2,71) were associated with highest in-hospital mortality. **Conclusion:** Patients with acute pulmonary thromboembolism objectively diagnosed have higher mortality than patients with objectively excluded PE. Further, non-conclusive approach of PE was an independent risk factor for in-hospital death

DETECÇÃO FENOTÍPICA DE PSEUDOMONAS AERUGINOSA HIPERMUTÁVEIS EM PACIENTES COM FIBROSE CÍSTICA EM HOSPITAL UNIVERSITÁRIO TERCIÁRIO.

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Introdução. *Pseudomonas aeruginosa* é o patógeno predominante no trato respiratório de pacientes com fibrose cística e a falha no tratamento desses pacientes colonizados cronicamente tem sido associada com a presença de subpopulações hipermutáveis. **Objetivos.** O objetivo deste estudo foi avaliar a prevalência de *Pseudomonas aeruginosa* hipermutáveis em amostras clínicas do trato respiratório inferior provenientes de pacientes fibrocísticos e verificar a relação entre fenótipo hipermutante e resistência aos antimicrobianos. **Materiais e Métodos.** Foram isoladas 200 *P. aeruginosa* de amostras representativas do trato respiratório inferior de 83 pacientes com fibrose cística. Para verificar a presença (ou ausência) de colônias de subpopulações resistentes dentro das zonas de inibição, bem como a sua quantificação, foi realizado teste de suscetibilidade modificado utilizando discos de ceftazidima, imipenem, meropenem, ciprofloxacina e tobramicina. As subpopulações resistentes foram isoladas e novo teste de suscetibilidade foi realizado. Os isolados cujos diâmetros de zona de inibição foram reduzidos em ≥5 mm em três ou mais antibióticos em relação à zona de inibição original foram considerados hipermutáveis. **Resultados e Conclusões.** Foi possível identificar subpopulação hipermutável em 56 (28,0%) isolados de 43 (51,8%) pacientes. A subpopulação hipermutável, quando testada individualmente, apresentou maior resistência aos antimicrobianos que a população predominante. A detecção de subpopulações hipermutáveis por laboratórios clínicos tem grande valor, pois estas subpopulações podem refletir mais precisamente os índices de resistência de *P. aeruginosa* em pacientes com fibrose cística.

TÉCNICA DE USO DOS DISPOSITIVOS INALATÓRIOS EM PACIENTES ASMÁTICOS

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Introdução: O corticóide inalatório (CI) é a principal medicação para o tratamento de manutenção da asma e a técnica inalatória adequada é fundamental para o controle da doença. Objetivos: avaliar a técnica de uso dos dispositivos inalatórios no tratamento de manutenção da asma. Métodos: estudo transversal, prospectivo, em pacientes com diagnóstico de asma e em acompanhamento ambulatorial. A coleta dos dados clínicos foi realizada por questionário padronizado aplicado após consulta ambulatorial. Os pacientes foram testados quanto às etapas de utilização de seus dispositivos inalatórios pelos membros da pesquisa. Resultados: Foram estudados 253 pacientes, sendo que 128 (50,6%) realizaram a técnica inalatória correta em todas as suas etapas. O uso correto da técnica inalatória se associou com o estado civil (uso correto mais fre-