**Introdução:** Linfangioleiomiomatose é uma doença multissistêmica, predominatemente dee mulheres, resultante da proliferação anormal de músculo liso expressando antígenos do melanoma levando a obstrução vias aéreas com degeneração cístcia pulmonar, infiltração linfática e de abdominal. **Objetivo:** Descrever associação de linfangioleiomiomatose e artrite reumatóide. **Materiais e métodos:** relato de caso e revisão da literatura pelo MEDLINE. **Resultados:**Mulher 54 anos, branca, não tabagista, com artrite reumatóide, uso crônico de imunossupressores e anti-TNFα, menopausada, tosse seca há 8 meses. Fáscies curshingóide, eupnéica, Sat 96% aa, presença de deformidades articulares. Ausculta pulmonar limpa. Radiografia tórax normal. TCAR imagens císticas arredondadas difusas em ambos pulmões, Avaliação funcional pulmonar normal. Fator reumatóide, FAN negativos. LBA com predomínio linficítico, pesquisa de pneumocistis negativa Anatomopatológico e imunohistoquimíca: lifangioleiomiomatose. **Discussão:** Linfangileiomiomatose é uma doença rara, predomínio feminino, idade média ao diagnóstico de 35 anos. Até 34% dos indivíduos apresentaram resultados espirométricos normais, com um declínio maior do que média da população normal. Dispnéia é o principal sintoma, desenvolvendo-se em metade dos pacientes. Tratamento com progestágenos, parece não diminuir ou até aumentar o declínio da DLCO. Estudos com 1 ano de tratamento com sirolimus mostraram aumentos de VEF1 e CVF, tendo como promissora a inibição da via mTOR no tratamento da LAM. Após extensa revisão por meio do medline não conseguimos encontrar descrição de associação entre linfangioleiomiomatose e artrite reumatóide e/ou com uso de anticorpos monoclonais anti-TNF α, porém há relatos de pelo menos 24 casos de doença intersticial pulmonar secundária ao uso dos mesmos.

## SARCOIDOSE PULMONAR E DE OUTROS ÓRGÃOS: SÉRIE DE PACIENTES EM ACOMPANHAMENTO NO SERVIÇO DE PNEUMOLOGIA DO HOSPITAL DE CLÍNICAS DE PORTO ALEGRE.

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Introdução: A sarcoidose é uma doença granulomatosa crônica, que pode afetar diversos órgãos, especialmente os pulmões. Freqüentemente apresenta-se com adenomegalias hilares, infiltrado pulmonar e acometimento cutâneo e ocular. Objetivo: Descrever os achados clínicos e funcionais de pacientes com sarcoidose em acompanhamento no Serviço de Pneumologia do Hospital de Clínicas de Porto Alegre. Método: Revisão do prontuário eletrônico dos pacientes com diagnóstico clínico-histológico de sarcoidose de qualquer órgão, consecutivamente atendidos no Ambulatório de Pneumopatias Difusas do Serviço de Pneumologia do HCPA no período de Outubro de 2006 a Março de 2007. Resultados: Foram estudados 21 pacientes, 15 eram do sexo feminino (71,4%). A média de idade foi 45,9 ± 7,8 anos. Todos os pacientes apresentavam acometimento pulmonar. Dois pacientes tinham sarcoidose cutânea, 2 sarcoidose ocular e 1 apresentava envolvimento multissistêmico. Os principais sintomas ao diagnóstico foram dispnéia em 8 (38%) e tosse em 3 (14,3%). Um paciente tinha hipercalcemia e 5 hipercalciúria. O diagnóstico de sarcoidose foi clínico em 9 pacientes, por biópsia transbrônquica em 4, biópsia cirúrgica em 3, mediastinoscopia em 3 e biópsia de linfonodo cervical em 2 pacientes. Nove pacientes tinham sarcoidose estádio I, 7 estádio II e 5 estádio III. Em 10 casos a espirometria foi normal, em 7 havia um DVO e em 3 um DV misto. A difusão foi normal em 5 casos, com redução leve em 7, moderada em 4 e grave em 4. Apenas 1 paciente tinha hipertensão pulmonar. Três pacientes apresentaram dessaturação significativa no teste da caminhada de 6 minutos. Doze pacientes receberam tratamento. Nove usaram prednisona, 1 usou prednisona e azatioprina e 1 prednisona e metotrexate. Conclusão: Os pacientes com sarcoidose apresentam comumente envolvimento pulmonar, e o tratamento realizado na maioria dos casos é a corticoterapia sistêmica ou somente o acompanhamento clínico.

## PULMONARY HYPERTENSION IN ONE PATIENT SUBMITTED TO THORACIC RADIOTHERAPY IN CHILDHOOD. IS ACTINIC PULMONARY ENDARTERITIS POSSIBLY THE LINK?

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**Background:** Radiotherapy has been related to abnormalities in lung parenchyma and vasculature. Lung fibrosis and pneumonitis are the most common described lesions. There are few cases of pulmonary endarteritis secondary to chest radiotherapy published in medical literature.**Purpose:** To report a case of an adult patient with severe pulmonary hypertension that has been submitted during childhood to thoracic radiotherapy to treat lung metastasis of Wilms tumor. **Case Report:** Patient is a twenty-nine years-old male with Wilms tumor at the age of one, submitted to surgery and abdominal radiotherapy. Four years later, thoracic radiotherapy was done to treat pulmonary metastasis. A complaint of exertional dyspnea was noticed about 7 years ago. At that time, pulmonary hypertension was diagnosed by transthoracic echocardiogram and patient was put on diltiazem with symptomatic control. Patient was admitted at the hospital with functional class III and signs of right cardiac failure. Further complementary evaluation with chest high resolution computed tomography and pulmonary function test were compatible with interstitial lung disease. Haemodynamic measures confirmed severe pulmonary hypertension

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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