Doenças pulmonares intersticiais

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Foram apresentados cinco trabalhos sobre Doenças Pulmonares Intersticiais.

EVALUATION OF THE SHORT-FORM 36-ITEM QUESTIONNAIRE (SF-36) TO MEASURE HEALTH-RELATED QUALITY OF LIFE IN PATIENTS WITH IDIOPATHIC PULMONARY FIBROSIS (IFP). Martinez *et al.* – Os questionários de qualidade de vida são divididos em gerais e específicos. Neste estudo um questionário geral traduzido e validado no Brasil foi aplicado a um grupo de pacientes com fibrose pulmonar idiopática e demonstrou que a dispnéia medida pela escala de Mahler tem a melhor correlação com os diversos domínios. Questionários específicos de qualidade de vida devem ser desenvolvidos para doenças intersticiais e são importantes porque o tratamento também resulta em freqüentes efeitos colaterais.

OBSTRUCTIVE PATTERN IN ASYMPTOMATIC LONG TERM RHEUMATOID ARTHRITIS (RA) PATIENTS: A PULMONARY FUNCTION TEST (PFT) AND HIGH RESOLUTION COMPUTED TOMOGRAPHY (HRCT) EVALUATION. Carvalho *et al.* – Obstrução ao fluxo aéreo é comum na artrite reumatóide, mesmo em não-fumantes (16% numa série recente) e é mais freqüente se há síndrome de Sjögren associada. Este estudo demonstra a maior sensibilidade de tomografia de alta resolução (padrão em mosaico) em relação à espirometria na detecção do comprometimento das vias aéreas na artrite reumatóide.

INTRATHORACIC TRACHEAL WIDENING (ITW): A NEW RADIOLOGICAL FINDING IN IDIOPATHIC PULMONARY FIBROSIS. Rubin *et al.* – O alargamento da traquéia intratorácica é freqüente na fibrose pulmonar idiopática e os autores chamam a atenção para este sinal radiológico pouco salientado. De modo inesperado não houve correlação com pior sobrevida nesta grande série de pacientes.

CARDIAC INVOLVEMENT IN SARCOIDOSIS-ECHOCARDIOGRAPHY AND MYOCARDIAL SCINTIGRAPHY WITH THALLIUM 201. Reis Santos *et al.* – Arritmias complexas, distúrbios de condução ou insuficiência cardíaca podem ser a manifestação inicial da sarcoidose. Na doença diagnosticada o estabelecimento de comprometimento cardíaco dita tratamento agressivo com corticóides. No ecocardiograma vários achados foram encontrados neste estudo, mas são inespecíficos. A média de idade nesta série foi elevada para sarcoidose. O mapeamento com tálio demonstra áreas segmentares de captação reduzida, o que corresponde a áreas de substituição fibrogranulomatosa, Estes defeitos são inespecíficos e podem ocorrer com isquemia miocárdica e em outras doenças que resultam em infiltração miocárdica e miocardiopatia. No estresse, os defeitos de perfusão na sarcoidose são reduzidos, ao contrário da doença coronariana. Na presença de angiografia coronariana normal defeitos de perfusão no tálio sugerem fortemente sarcoidose miocárdica, como em 3 casos da presente série.

PULMONARY DYSFUNCTION IN JUVENILE RHEUMATOID ARTHRITIS. Mártire *et al.* – Este estudo demonstra que manifestações pulmonares são freqüentes na artrite reumatóide juvenil, sendo pleurite a mais comum. A utilização de difusão de CO e de tomografia de alta resolução poderia resultar em maior detenção de anormalidades. Metotrexato é comumente utilizado no tratamento de artrite reumatóide e pode resultar em toxicidade pulmonar. A toxicidade usualmente se apresenta como uma pneumonite intersticial, mas também pode resultar em alçaponamento de ar como sugerido na presente série, em que se encontrou correlação inversa entre o tempo de uso do MTX e o $FEF_{5-75\%}$.

EVALUATION OF THE SHORT-FORM 36-ITEM QUESTIONNAIRE (SF-36) TO MEASURE HEALTH-RELATED QUALITY OF LIFE IN PATIENTS WITH IDIOPATHIC PULMONARY FIBROSIS (IPF).

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Objective: health related quality of life (HRQL) is an important domain for measuring the impact of chronic diseases. IPF is a serious disorder frequently leading patients to disability. The purpose of this study was to examine the validity of the general HRQL instrument SF-36 in IPF patients, by assessing the relationship between dyspnea ratings plus lung function, and the scores for the components of the questionnaire. Methods: we have studied 33 IPF patients (open lung biopsy = 25), with no significant comorbidity, and a mean age of 59.6 ± 10.9 years. There were 21 men and 12 women, and the mean FVC was 70.1 ± 21.2%. Patients were submitted to spirometry and rest arterial blood gases. Dyspnea was assessed using the basal dyspnea index (BDI-Mahler). Results: Dyspnea was an important complaint for IPF patients (BDI = 5.4 ± 2.6). There was severe impairment in four domains of the SF-36 questionnaire physical functioning (PF), physical role (PR), general health perception (GHP), vitality (V) and social functioning (SF). The scores for emotional role (ER) and mental health index (MHI) were also slightly decreased. Strong correlations were found between BDI scores and SF-36 components (PF r = 0.82; V r = 0.82; GHI r = 0.57; MHI r = 0.60; SF5 = 0.52). Weak correlations were found between SF-36 components and lung functions parameters. Conclusions: HRQL is an important outcome in evaluating IPF patients. SF-36 is a useful

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instrument to measure HRQL also in IPF populations. Dyspnea has a major impact in the HRQL of such subjects.

OBSTRUCTIVE PATTERN IN ASYMPTOMATIC LONG TERM RHEUMATOID ARTHRITIS (RA) PATIENTS: A PULMONARY FUNCTION TEST (PFT) AND HIGH RESOLUTION COMPUTED TOMOGRAPHY (HRCT) EVALUATION.
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To evaluate the incidence of pulmonary involvement in long term RA we prospectively performed HRCT and PFT in 29 asymptomatic patients with more than 15 years of disease duration (24.8 ± 8.0 y.). Overall PFTs were normal, with a median predicted FVC of 97.0%; FEV_1, 103.5%; FEV_1/FVC, 105.4%; $FEF_{25-75\%}$, 75.0%; TLC, 110.1% and RV of 119.1%. On the HRCT, altered distribution of the ventilation (ADV), characterized by a mosaic pattern, was found in 12 patients (41.4%). The absolute value of FEV_1/FVC was significantly lower in patients with ADV (p = 0.044, 70.7 ± 15.0 x 79.3 ± 10.7). These patients were placed on corticosteroids more often during their disease course (p = 0.015, 10/12 ADV x 6/17 no ADV). The presence of bronchiectasis on HRCT was more common in the group with ADV (p = 0.03, 6/12 x 2/17) as well as bronchial wall thickening (p = 0.04, 11/12 x 9/17). The presence of ADV reduced the presentation of dependent densities (p = 0.004, 1/12 x 10/17). After excluding patients with secondary Sjögren syndrome, the presence of ADV correlated more significantly with PFT parameters of obstruction [FEV_1 (%), p = 0.006; FEV_1/FVC, p = 0.01; $FEF_{25-75\%}$ (%), p = 0.01]. We found that a mosaic pattern suggestive of ADV is a common finding in long term RA patients. Such finding is closely related to a more obstructive pattern on PFT and to more prescriptions of corticosteroids. We conclude that an obstructive pattern is a common pulmonary involvement in asymptomatic long term RA patients.

INTRATHORACIC TRACHEAL WIDENING (ITW): A NEW RADIOLOGICAL FINDING IN IDIOPATHIC PULMONARY FIBROSIS.
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In order to verify the occurrence of intrathoracic tracheal widening (ITW), we had study the X-ray of 132 patients with histologic confirmed idiopathic pulmonary fibrosis (IPF), between 1970-1996, coming from our hospital. The ITW was determinated when the difference between the cervical trachea and the thoracic trachea diameter was superior to 1mm, meaning the presence of widening. ITW is related with negative subpleural pressure determined by the inspiratory effort against the increase pulmonary elastic recoil in IPF. In our patients, ITW was observed in 49 (37.1%) cases. Although in patients with ITW we had worst prognostic factors like lung function tests, duration of symptoms, age, sex, dyspnea, index, histologic pattern and tomographic profusion, there was no statistical significance difference between patients with or without this radiological characteristic. There was no correlation, as well, with the survival rates of this group (42 months for all and 32.6 months for dead patients). It is interesting to observe that others intersticial lung disease like sarcoidosis or extrinsic allergic alveolitis do not have this radiological finding. We conclude that, although it is not a prognostic factor, the ITW is a particular finding of IPF, present in a numerous group of patients, easy to be verify and this observation associated with others findings like clubbing and interstitial lung infiltrate orient to the diagnosis of this disease.

CARDIAC INVOLVEMENT IN SARCOIDOSIS – ECHOCARDIOGRAPHY AND MYOCARDIAL SCINTIGRAPHY WITH THALLIUM 201.
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Introduction: sarcoidosis (Sarc) is a granulomatous systemic disease and also may involve the heart and in the majority of cases as asymptomatic way. We studied 30 patients (Pats) with Sarc through echocardiography (Echo) and myocardial (Myoc) Scintigraphy (Scin), to determine the heart involvement. Methods: the age was between 33-73 years old, mean 50.97 ± 10.23; 5 (16.67%) males and 18 (60%) blacks. All of them were submitted to Echo and 22 to Myoc imaging with Thallium 201 (Tl 201) at rest and after Dipyridamole (Dip) infusion; the elderly (over 65 years old) did not do it. Four that had abnormal Scin were studied through angiography (Ang). Results: at Echo, 10 (33,33%) had diastolic dysfunction (Diast Dysf), 1 restrictive type, 3 were elderly and 1 had left ventricular hypertrophy (LVH), 6 (20%) LVH; 3 (10%) degenerative valvular disease; 3 (10%) had mitral valve prolapse, 3 (10%) had dilated cardiomyopathy, one also had septal aneurism. Those Pats submitted to Myoc Scin, 12 (54.55%) had normals exams and from these, 5 (41.67%) had abnormal Echo, and from the 10 who had abnormal Scin, 7 had abnormal Echo too, there were not statistical differences (p = 0.369) in these findings. Diast Dysf was present in 2 normal (16.67%) Scin. And in 2 abnormal (20%). Reduced uptake of Tl 201 in septal (58.33%) and in anterior (45.95%) regions at stress were the most common findings at Scin. Three of these with abnormal Scin were submitted to Ang and were normals. Conclusion: even though we have a short number of Pats studied, we think that Echo and Scin with Tl 201 at rest and with pharmacologic stress test with Dip can help to determine the involvement of the heart in Sarc.

PULMONARY DYSFUNCTION IN JUVENILE RHEUMATOID ARTHRITIS.
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Fifty pediatric patients, aged between 6 and 18, with juvenile rheumatoid arthritis, systematic onset, were selected according to the American Rheumatism Association, classes I to IV of functional joint limitation index. Clinical data was analyzed as well as previous pulmonary disease, chest X-rays and spirometry. Predicted values and lower 95º percentile were calculated from the regression equation of Knudson for FVC, FEV_1, VEF_1/FVC and $FEF_{25-75\%}$. Thirty-one children (62%) had some kind of lung illness during rheumatoid disease. Arthritis preceded the pulmonary disease in 22 cases; it was simultaneously in 5 and succeeded in 4 patients. Pleurisy was the most common finding during the course of the disease (28%); interstitial infiltrates was finding in 22%. It was find out that spirometry was the most sensitive method to document pulmonary disease. Pulmonary impairment became evident in 48% patients, with predominance of restrictive ventilatory pattern (36%), lower FVC and normal FEV_1/FVC. Simultaneous use of spirometry and simple chest X-rays increased detection of abnormalities, indicating that they are correlated complementary methods. Decrease in lung volume did not correlate with functional articular limitation, typer and index; treatment (salicylates, gold, methotrexate); period of treatment and the duration of the disease. There was a negative correlation between $FEF_{25-75\%}$ and treatment time with methotrexate (0.69 – Pearson).