

MONDAY, SEPTEMBER 26TH 2011

327. Novel clinical features of acute and chronic lung diseases

2983**Prevalance of pulmonary hypertension in chronic myeloproliferative diseases**

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To evaluate the incidence of pulmonary hypertension (PH) in essential thrombocythemia (ET) and polycythemia Vera (PV), which are chronic myeloproliferative disorders, 70 patients (55PV+15ET), who have no co-morbidity causing PH, were prospectively evaluated. All patients had echocardiography done by the same cardiologist, spirometry and DLCO. Two parameters were used for PH: mean pulmonary artery pressure (PAP) calculated by Mahan formula >25 mmHg and right ventricular systolic pressure (RVSP) >35 mmHg. For the first parameter, 3 (4,3%) patients (1ET, 2PV) and for the second parameter, 4 (5,7%) patients (2ET, 2PV) had PH. Patients' characteristics are reported in the table.

Conclusion: When compared with the previous few studies done by small populations, PH incidence in our study was found very low. This result may be related

Patient's characteristics

	PV (n=15)	ET (n=55)	PV+ET (n=70)
Sex (M/F)	7/8	36/19	43/27
Age	54.4±16.8	55.2±12.3	55.0±13.3
Duration of disease (year)	6.7±4.5	7.2±4.7	7.1±4.6
Jak2 mutation	86.7	50.9	58.8
Dyspnea (%)	20	23.6	22.9
Splenomegaly (%)	13.3	20	18.6
Non pulmonary thrombosis (%)	26.7	10.9	14.3
Bleeding (%)	0	12.7	10.0
Hct (%)	43.7±5.0	39.4±4.6	40.4±5.0
Basal thrombocyte (×1000000000/L)	592.7±327.5	1108.5±371.7	997.9±418.7
Last thrombocyte (×1000000000/L)	400.9±191.5	559.0±231.5	525.1±231.6
Mean thrombocyte (×1000000000/L)	535.4±189.0	711.9±241.8	673.6±241.5
RVSP (mmHg)	31.4±11.9	28.2±6.6	28.8±7.7
Mean PAP (mmHg)	20.8±4.8	19.9±3.8	20.1±4.1
FVC (%)	109.2±13.8	108.9±22.5	108.9±20.8
FEV1 (%)	106.3±16.0	109.4±19.7	108.7±18.9
DLCO(Hb) (%)	128.1±23.0	120.4±19.7	122.1±20.5
Warfarin (%)	6.7	3.6	4.3
Aspirin (%)	93.3	96.4	95.7
Hydroxyurea (%)	100	87.3	90

to effective treatment (cytoreductive treatment) of myeloproliferative diseases in the majority of the patients.

2984

Adipokines and bone loss at the terminal stage of chronic respiratory failure

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Aim of this study to evaluate associations between the adipokines (tumor necrosis factor-alpha (TNF-α) and its receptors, leptin, adiponectin), body composition and bone mineral density (BMD) in the patients with terminal stage of chronic respiratory failure.

Material and methods: 47 patients with end-stage of chronic respiratory failure (COPD, emphysema and cystic fibrosis) and 35 healthy subjects were estimated. Bone mineral density, body composition was measured by dual-energy X-ray absorptiometry (DEXA) at the lumbar spine (LS) and left femur neck (FN). We estimated respiratory function testing, serum levels TNF-α, TNFR-1, TNFR-2, leptin, adiponectin.

Results: We identified a decreased BMD characterized by T-score <-1.0 in 43/47 patients, as measured on the FN or the lumbar spine (LS). There was negative association between TNF-α and BMD (r=-0.43, p=0.04). Parameters of body compositions and serum concentrations of leptin and adiponectin were significantly associated with FN hip and LS. Serum leptin levels was significant lower (p=0.047) and adiponectin concentrations was higher (p=0.039) in the osteoporosis group. Serum leptin significantly positively correlated with parameters of body composition, serum adiponectin concentrations was negative association with TNFR-1, TNFR-2 (p=0.007). There was a significant inverse relationship between leptin and adiponectin.

Conclusion: These results shows possibly role of adipokines in the increasing of bone loss at the terminal stage of chronic respiratory failure.

2985

Bronchoalveolar interleukin-1 beta: A marker of bacterial burden in children with community-acquired pneumonia (CAP)

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Objective: To assess the relationship between concentrations of bronchoalveolar cytokines and bacterial burden in children with CAP.

Method: 58 children were divided into two subgroups: referral (n=28), and treated (n=30) CAP. Bronchoalveolar lavage was performed in the most abnormal area on chest radiograph by fiberoptic bronchoscope. Bronchoalveolar lavage fluid was processed for quantitative bacterial culture. The concentrations of bronchoalveolar lavage cytokines (tumor necrosis factor-alpha, interleukin-1 beta, interleukin-6, interleukin-8, and interleukin-10) also were measured.

Results: Thirty-two patients had a positive bacterial culture (bronchoalveolar lavage > or = 10 colony-forming units/mL), and made up 76% of pathogens recovered at high concentrations. The concentrations of bronchoalveolar lavage interleukin-1 beta were 181.1±16.3 and 45.1±10.6 pg/mL (mean ± se) in the children with positive and negative bacterial culture, respectively (p <.001). Bronchoalveolar lavage interleukin-1 beta was significantly higher in the children with a high bacterial burden (p <.001), with mixed bacterial infection (p <.001), and with CAP (p <.001), compared with values in patients without these features. The relationship between bacterial load and concentrations of bronchoalveolar lavage interleukin-1 beta was very strong in the children with referral CAP but was borderline in treated CAP.

Conclusions: Concentration of bronchoalveolar lavage interleukin-1 beta was cor-

related with bacterial burden in the alveoli, it may be a marker for progressive and ongoing inflammation in children who have not responded to CAP therapy.

2986

Features of lung damage in adult patients with severe influenza A (H1N1)

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Methods: 445 adult patients (36,5±13,5 years) were hospitalized with a suspected influenza A (H1N1) from October 2009 to January 2010. In 84% of cases diagnosis was confirmed by RT-PCR test. Clinical, laboratory, and X-ray examinations, complex study of respiratory function were carried out.

Given the threat of influenza pandemic in 2011 the aim was the study of detection frequency, risk factors for lung damage, and features of convalescence period in patients with severe influenza A (H1N1).

Results: The average score on Charlson index was 1,55±0,90. In 181 patients pneumonia was diagnosed, double pneumonia - in 23% of cases. Acute respiratory distress syndrome developed in 12 patients and they needed in ventilation support. Four patients died within 20 days (IQR of 10-25). The relative risk of death was associated with higher estimates for Charlson index (RR 1,38; 95% CI 1,04 to 1,82, p=0,02) and APACHE II scale (RR 1,242; 95% CI 1,284 to 1,602, p=0,005). In convalescents the increased lung pattern on radiographs (39%), decreased lung diffusion capacity (45%), restrictive changes (15%) were the most frequent and long-lasting changes. 28% of patients having these changes applied for medical care within 6 months after being discharged.

Conclusion: The high score on Charlson index and APACHE II scale are risk factors for development of severe pneumonia in presence of influenza A (H1N1). Changes of ventilation functional tests associated with persistent disorders of pulmonary circulation remain for long time in 45% of patients undergoing pneumonia. This group of patients is of particular interest and requires further observation.

2987

Specialist palliative care is more than drugs – A retrospective study of ILD patients

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Background: Little is known about the palliative care needs of patients with Progressive Idiopathic Fibrotic Interstitial Lung Disease (PIF-ILD). As part of a study to develop a complex palliative intervention at the end of life, we retrospectively studied patients dying in 2 London Hospitals.

Aims: To assess the palliative care needs and management of PIF-ILD patients in 2 London ILD centres.

Methods: Patients' records from Royal Brompton Hospital (RBH) and King's College Hospital (KCH) were extracted to assess palliative care needs, use of palliative treatments and whether end of life preferences were documented and achieved.

Results: 45 PIF-ILD patients were identified (26 RBH,19 KCH). Patients at RBH were younger (37-81y, median 61y) and predominately white British (23/26) compared to KCH's older, more racially diverse population (70-99y, median 82y, 6/19 non-white).

17/45 patients had specialist palliative care team involvement. Nearly all patients experienced breathlessness in their last year of life (42/45) and almost a third of patients experienced chest pain (13/45). Additional symptoms included cough, fatigue and depression/anxiety.

All patients given opioids (22/45) or benzodiazepines (8/45) had documented benefit. Non-pharmacological treatments were rarely used.

Few patients had preferred place of care (8/45) or preferred place of death (6/45) documented and the majority of patients died in hospital (34/45).

Conclusion: Despite demographic variation, the patients experienced similar symptoms. There was use of standard pharmacological treatments with symptom benefit. Non-pharmacological interventions were seldom used and documentation of preferred place of care and death was poor.

2988

Community-acquired pneumonia in pediatric patients with connective tissue disorders: Manifestations and clinical course

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Objective: High rates of connective tissue disorders (CTD) among children modify

MONDAY, SEPTEMBER 26TH 2011

clinical course of community-acquired pneumonia (CAP) in pediatric patients that leads to diagnosis and therapeutic mistakes.

Aim: To study role of CTD in CAP clinical course in children.

Methods: 171 children aged 0-18 years with CTD and CAP were observed. CAP was clinically and radiographically diagnosed with detection of serum antibodies (IgG and IgM) against intracellular pathogens measured by enzyme-linked immunosorbent assay (ELISA) and PCR.

Results: Recurrent course of CAP (with 3 and more episodes in 5-year period of study) was in 106 (61,9%) patients and in all cases followed a viral respiratory infection. 156 (91,2%) children had abnormal perinatal history. Asthma (A) was diagnosed in 59 (34,5%) children. CAP caused by *Mycoplasma pneumoniae* (Mp) was in 88 (51,5%) patients; by *Cytomegalovirus* (Cmv) and Mp – in 42 (24,5%); by *Chlamydia pneumoniae* (Cp) – in 18 (10,5%); by Cp and Mp – in 33 (13,5%) patients. 78% of patients with A, CTD and CAP and 53,6% of patients with CTD and CAP had pulmonary hypertension (PH). 76,4% of patients with recurrent CAP demonstrated radiographic and clinical evidence of pulmonary fibrosis (PH), 30,2% of them had A. 12,5% of children with A, CTD, recurrent CAP and PF had CT evidence of development of pneumatocele (PC).

Conclusions: 1. Manifestations of CTD were in all children with CAP. 2. All patients with CTD and recurrent CAP had abnormal perinatal history. 3. High frequency of PH and PF was observed in patients with CTD and recurrent CAP. 4. 12,5% of children with A, recurrent CAP, CTD and PF had evidence of development of PC.

2989

Clinical features of a new hypersensitivity pneumonitis: Salami brusher's disease

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We observed 5 consecutive cases of hypersensitivity Pneumonitis (HP) in subjects working in a salami factory. The workers had to clean the mould growing on salami's surface by using a wire brush. The working population was of 30 female subjects, 5 of them developed a HP (17%). Two were smokers (40%) and, other two were asthmatic. All patients presented with an acute clinical manifestation with cough, high fever, dyspnea and hypoxia occurring after a short period after exposure (240±60 minutes). Three of them presented at the emergency department and a chest x-ray showed and alveolar interstitial pneumonitis and were treated as a community acquired pneumonia. Skin prick test were positive for *Penicillium spp* in 3 cases and for *Cladosporium spp* and *Aspergillus spp* in other 2. The results of serum immunoglobulin (Ig) G and IgA antibodies against *Penicillium spp* were positive in 3 patients, 2 patients were positive to *Aspergillus Fumigatus*. Pulmonary function test demonstrated a reduction in diffusing capacity in all 5 patients (60±15% of predicted). A bronchial hyperresponsiveness to methacoline was present in all the patients, the mean dose of methacoline causing a 20% fall in FEV1 was 3.63±4.70 mg/mL. BAL data showed a lymphocytosis 44.4±8.2%, CD4+ were 26.5±6.3%, CD8 + 53.5±8.2. Four patients had a complete recovery after changing work, and one was treated with oral steroids for increasing dyspnea and severe interstitial involvement. Salami's brusher disease is a new type of HP, the prevalence is high in exposed subjects, and is common even in smokers. The disease has an acute clinical onset. The probable antigen is *Penicillium*, but *Cladosporium* and *Aspergillus* may play a key role.