Evaluation of airway resistance through the interrupter technique in cystic fibrosis

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Few studies have been published on airway resistance measurements using the interrupter technique (IT). We performed a cross-sectional study, evaluating 38 children and adolescents with Cystic Fibrosis (CF), followed at the outpatient CF clinic of Hospital São Lucas from Pontificia Universidade Católica do Rio Grande do Sul. Airway resistance (Rint) was measured by the IT, followed by spirometry in all patients. Measurements were repeated after inhalation of salbutamol in order to evaluate bronchodilator response. There was a strong correlation between inverse Rint and forced expiratory volume in one second (FEV1) (r=0.8, p<0.001) and fair correlations between the inverse Rint and mid expiratory flow (MEF) (r=0.74 p<0.001) and between inverse Rint and body mass index (BMI) (r=0.62 p<0.001). The accuracy of bronchodilator response by the IT was tested through the ROC (receiver operating curve), comparing results with spirometry bronchodilator response. An area of 0.75 under the curve was obtained, for the cutoff point of −28% of Rint, achieving a sensitivity of 66% and a specificity of 92%. The findings suggest that Rint shows good correlation with spirometry parameters, although the IT is not sufficiently accurate to replace spirometry in the evaluation of bronchodilator response.

Markers of oxidative stress in exhaled breath condensate: prediction of one-year development of pulmonary function and nutritional status in cystic fibrosis

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Aims: To evaluate relation of oxidative stress markers (MOS) in exhaled breath condensate (EBC) and development of pulmonary function (forced expiratory volume in 1 sec.; FEV1) and nutritional status (body mass index; BMI) parameters in CF patients.

Methods: 25 CF patients (14 males) with mean age 24.1 years were examined. FEV1, BMI and EBC MOS were evaluated at the beginning of the study. EBC was collected using ECOScreen condenser. Examined MOS included sum oxidized nitrates and nitrates (NOx) and 8-isoprostan (8-IP) and were assayed using liquid chromatography and ELISA method, respectively. FEV1 and BMI were examined also after one-year follow-up and their differences from initial values were calculated and correlated with EBC MOS using stepwise regressions.

Results: Mean EBC concentrations of NOx and 8-IP were 13.8 μmol/ml and 16.0 μg/ml, respectively. Mean FEV1 and BMI values changed from 64.0 to 57.9% pred. and from 21.9 to 22.0 kg/m², respectively. Change in FEV1 correlated with initial EBC 8-IP concentration (p<0.05) and change in BMI with initial EBC NOx concentration (p=0.039). No other correlations were observed.

Conclusions: We found weak correlation of EBC MOS and pulmonary function and nutritional status development in CF patients.

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Decline in pulmonary function after introduction of macrolide maintenance therapy

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Background: Chronic Pseudomonas aeruginosa (PA) infection in cystic fibrosis (CF) patients is associated with increased decline in pulmonary functions. In 1998 macrolide maintenance therapy (MMT) was introduced in our clinic as adjuvant to inhalation antibiotics in CF patients with chronic PA infection. In this study we investigated the effects of MMT on decline in pulmonary function.

Methods: Adult CF patients with chronic PA infection and MMT were compared to CF patients without chronic PA infection. Patients with infection with B cepacia complex, S. maltophilia, or A. xylosidans were excluded. Developments in pulmonal function, body mass index (BMI), and hospitalisation between 2000 and 2007 were investigated retrospectively.

Results: A total of 64 CF patients with chronic PA infection were included and compared to 23 CF patients without chronic PA infection. Median age in 2000 was 28.5 and 30.9 years, respectively. BMI was comparable throughout the years. Number of in-hospital days was higher in patients with chronic PA infection (19 versus 0 days, p<0.001). At baseline median forced expiratory volume in 1 second as % of predicted (FEV1) was lower in patients with chronic PA infection (62.3% versus 79.0%, p<0.001), but the rate of decline in FEV1 over the years, estimated by linear mixed models, was not higher (1.28% versus 1.31% decline per year, p=0.722).

Conclusion: The introduction of MMT in patients chronically infected with PA resulted in a lower yearly decline in pulmonary function compared to data in literature. The observed decline is comparable to patients without chronic PA infection.

Influence of early diagnosis in the development of bronchopulmonary disease in cystic fibrosis

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Objectives: To assess the impact of early diagnosis in the development of bronchopulmonary disease in school age patients (5–8 years) with Cystic Fibrosis.

Methodology: We selected those with sputum cultures including one culture per month. We compared 2 cohorts of patients: Group ED: Early detection (<2 months: prenatal diagnosis + neonatal screening + early clinical diagnosis), and DD: delayed diagnosis. We reviewed microbiological parameters, lung function, lung CT and hospitalizations for pulmonary exacerbation.

Results: We collected 29 patients, 18 in the DP group and 11 in the DD group. We analyzed 602 sputum, 40.7% were positive, the percentage was lower in the ED group (31.9% versus 53.7%) (p<0.0001). In both groups, Staphylococcus aureus was the most prevalent, being significantly more frequent in the DD group (p<0.005), whereas Pseudomonas aeruginosa was in the DD group (p=0.13). 63.6% of patients in the DD group were chronic carriers of Staphylococcus Aureus against 38% of the ED group, and 9% carriers of Pseudomonas aeruginosa and Burkholderia cepacia complex in the DD group vs 0% in group DE. No differences in lung CT between groups were found. Lung function was normal in 93.3% of the ED group compared with 75% of the DD group. DD group had more hospital admissions.

Conclusions: We observed differences in the number of positive cultures, iso- lation germ type, presence of chronic carriers, respiratory functional changes and pulmonary exacerbations. All this makes us support that early diagnosis, may improve the morbidity of cystic fibrosis by preventing or delaying the same bronchopulmonary disease.