190 Ventilation inhomogeneity in patients with cystic fibrosis aged 10–18 years

U. Pradal[1], A. Borruso[1], L. Menin[1], E. Fedrigo[1], B.M. Assael[1]. [1]*Azienda Ospedaliera Universitaria Integrata Verona, Cystic Fibrosis Center, Verona, Italy*

Spirometry is used to detect and monitor airway disease in CF but its normality cannot rule out the presence of early lung function abnormalities due to the obstruction of peripheral airways. The Lung Clearance Index (LCI), a measure of ventilation inhomogeneity, is used to detect early functional damage in infants and children with CF. Over the years life expectancy and lung disease control improved significantly in CF, such that at present an increasing number of patients reach adulthood with a normal spirometry. Aim of the present study was to understand if ventilation inhomogeneity is a common finding in these patients.

60 CF patients with FEV_1 >40%pred (30 males) aged 10–18 years were studied. Spirometry and whole body plethysmography were performed according to standard procedures. LCI was measured by Helium rebreathing in a water sealed spirometer with continuous CO_2 removal and O_2 supply in all patients and a group of healthy controls.

LCI values were significantly correlated with the measured lung function parameters, i.e. FEV_1%pred ($p < 0.0001$), $FEF_{25–75}$%pred ($p < 0.0001$) and FRC/TLC ($p = 0.0008$).

High LCI values were found in 41/60 patients (68%). Airway obstruction was less common (FEV_1 was abnormal in 14 patients, 23%). It is remarcable that 27/46 patients (59%) with normal FEV_1, 23/38 (60%) with normal $FEF_{25–75}$ and 14/24 (58%) with normal FRC/TLC had elevated LCI values.

These results suggest that ventilation inhomogeneity is common in CF patients 10–18 years old. In most cases common methods to study lung function fail to identify CF patients with airway damage thus leading to a possible delay in treating the disease.

191 Resistance and reactance measured by impulse oscillometry: Structural functional correlation in adult cystic fibrosis

K. Hurt[1,2,3], A. Nair[4], S. Sheard[4], D. Hansell[3,4], M. Hodson[2,3], O. Usmani[2,3], D. Bilton[2,3]. [1]*Brighton and Sussex University Hospitals NHS Trust, Respiratory Medicine, Brighton, United Kingdom;* [2]*Royal Brompton Hospital, Respiratory Medicine, London, United Kingdom;* [3]*Imperial College, London, United Kingdom;* [4]*Royal Brompton Hospital, Radiology, London, United Kingdom*

Objectives: Cystic fibrosis is characterised by airway obstruction. Reactance at 5Hz (X_5) measured by impulse oscillometry (IOS) determines the capacitive properties of the peripheral lung. Resistance measurements are frequency independent in health but in peripheral airway obstruction, resistance at 5Hz (R_5) is raised. IOS is quick, easy to perform and correlates with spirometry. It has a linear relationship with residual volume/total lung capacity ratio (RV/TLC). The aim of this study was to assess the relationship of IOS parameters with gas trapping measured on high resolution CT thorax (HRCT).

Methods: IOS, spirometry and plethysmography were performed in 24 adult patients with CF (17–56 years, FEV_1 26–119% of predicted). Two radiologists independently scored percentage gas trapping to the nearest 5% on HRCT scans. These scores were correlated with the physiological measurements.

Results: There was a strong inverse linear relationship evident between X_5 and RV/TLC ($r = –0.82$, $p < 0.01$) and CT gas trapping ($r = –0.65$, $p < 0.01$). X_5 correlated with FEV_1 ($r = 0.90$, $p < 0.01$), but the relationship was not linear. R_5 correlated with FEV_1 ($r = –0.73$, $p < 0.01$) and CT gas trapping ($r = 0.57$, $p < 0.01$) but not RV/TLC ($r = 0.47$).

Conclusion: These results suggest X_5 could be used as a convenient method to measure gas trapping in CF adults and may give as much information as plethysmography. The linear relationship with gas trapping indices and X_5, which is not present at lower FEV_1 values, indicates X_5 should be considered a potentially useful marker in severe CF lung disease.

192 Volumetric capnography is correlated with spirometry in cystic fibrosis patients

P.M.N. Oliveira[1], M.A.G.O. Ribeiro[1], A.A. Almeida-Junior[1], J.D. Ribeiro[1]. [1]*State University of Campinas – Medical School, Pediatrics, Campinas, Brazil*

Objectives: To compare and to correlate volumetric capnography (Vcap) and spirometry to evaluate cystic fibrosis (CF) pulmonary function.

Methods: A cross sectional study including a cohort of 58 CFpatients (31 female) at a University Reference Center of CF in 2012. All of them were evaluated with Vcap (CO2MOplus, Novametrix) and spirometry (CPFS/D, MedGraphics). The patients were out of pulmonary exacerbation. The median age was 14 years (9–24) and BMI 17.3.

Of the 58 patients 16 (27.6%) had normal and 42 (72.4%) abnormal spirometry. When comparing the groups with normal and abnormal spirometry (Mann-Whitney test) there was a significant difference in the Vcap phase 3 slope (P3Slp) ($p = 0.001$) and pulse ($p = 0.048$). In addition, there was a strong negative correlation between FEV1 and P3Slp ($p < 0.01$, $r = –0.714$).

Conclusion: Vcap is a quick and simple tidal breathing test and it is feasible to evaluate lung pulmonary disease in CF patients. P3Slp was correlated with FEV1. The method has the potential to be introduced as a screening tool into clinical routine. Vcap requires further evaluation before this technique can be recommended for monitoring CF lung disease.

193 Influence of nebulized Obracin® versus Tobi® on ciliary beat frequency

M. Boon[1], M. Jorissen[2], M. Proesmans[1], F. Vermeulen[1], K. De Boeck[1]. [1]*University Hospital Leuwen Belgium, Pediatrics, Leuwen, Belgium;* [2]*University Hospital Leuwen Belgium, Otorhinolaryngology, Head and Neck Surgery, Leuwen, Belgium*

Introduction: Tobramycine for inhalation is an established treatment for patients with CF and *Pseudomonas aeruginosa* infection. However, it is expensive and in many countries reimbursement is limited. Therefore, tobramycin solution for iv administration, is often used as an alternative.

Study objective: To examine the influence of Obracin® (O) versus Tobi® (T) on ciliary beat frequency (CBF) in an *in vitro* respiratory epithelial cell model.

Methods: We have previously constructed and validated a model for evaluation of nebulized drugs on CBF. Nasal epithelial cells were derived from non-CF surgery specimens and cultured as a monolayer. Evolution of CBF was expressed as % of the baseline value. Changes in CBF induced by nebulization with O (160 mg/4 ml), T (300 mg/4 ml) and isotonic saline (S) as control condition were measured and compared with mixed model analysis. Three samples of 3 different patients were exposed to nebulization of either T, O or S, on 3 consecutive days.

Results: Nebulization with O resulted in a significant decrease of CBF compared to nebulization with T (p 0.0001), even in a lower concentration of tobramycin. This was also true on each separate day (p 0.046 for day 1, p 0.026 for day 2 and p 0.008 for day 3). One hour after nebulization with O, CBF decreased to 59% of baseline ($p < 0.0001$), compared to 92% after nebulization with T (p 0.116) and 97% after nebulization with S (p 0.536). This inhibitory effect of O on CBF is possibly caused by additives in the parenteral solution.

Conclusion: Nebulisation of Obracin® impairs ciliary activity *in vitro.* Therefore, it may be advisable to prefer Tobi® in patients with CF or non-CF bronchiectasis.