133 Slow and fast lung compartments in cystic fibrosis measured by nitrogen multiple-breath washout (N_2 MBW)

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Objectives: To demonstrate that the volumes of under-ventilated lung regions and their specific ventilation can be measured from routine N_2 MBW tests in CF.
Methods: Triplets of N_2 MBW in 37 CF subjects aged 11–45 years and 74 age and sex matched controls. The fast and slow ventilating lung compartments and their specific ventilation were determined from semi-log plots of expired N_2 volume per breath vs breath number.
Results: Two-compartment lung emptying was not seen in healthy controls but in all CF subjects (FEV_1 mean (SD) 84.8 (19.9) %predicted; abnormal Lung Clearance Index (LCI) in 36/37, range 7.28–18.89). In CF, the slowly ventilated lung compartment ranged from 34–67% of full FRC, with specific ventilation (regional alveolar tidal volume/regional lung volume) 1.8–13.1%. LCI increased as specific ventilation of the slowly ventilated lung compartment decreased (r^2=0.70, $p < 0.001$). Over-ventilation of the fast lung compartment occurred in early but not late disease.
Conclusion: The magnitude and function of under- and over-ventilated lung volumes can be easily derived from routine N_2 MBW in CF. Reported values agree with previous modelling-derived estimates of impaired ventilation. The new compartment analysis extends our understanding of CF lung pathophysiology.

134 Nutritional status, lung function and LCI among children with CF

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Background: Nutritional status in patients with cystic fibrosis is associated disease outcome. Lung function and Lung Clearance Index (LCI) are both good predictors of disease severity.
Aim: To evaluate the association between nutritional status and lung function and LCI among children and adolescents with CF.
Methods: Nutritional status and pulmonary function of 77 patients with CF (53.2% males, mean age 10.73 years), were evaluated. Anthropometry was expressed as z-scores for age and sex using WHO Anthro software and classified following the WHO criteria, and Frisancho references for Mid-Upper Arm Circumference percentile (MUACp) and Triceps Skinfold Thickness percentile (TSTp). BMIpercentile (BMIp) was also calculated. Phase angle (PhA) and body cell mass (BCM) were assessed by BIA. Disease severity was assessed by FEV_1 (ATS/ERS guidelines), LCI (measured by multiple breath washout test, (EXHALYZER D, Ecomedics).
Results: The mean value for Weight/Height z score was –0.04 (±1.65), Height/age z was –0.37 (±1.19), Weight/age z was –0.22 (±1.39), BMI/age z was 0.07 (±1.32), MUACz was –0.12 (±1.29), TSFz was 0.06 (±1.03). The mean value of the BMIp was 50.16. The mean PhA value was 4.94 (±0.91). Mean values for the respiratory tests were FEV_1: 90 (±25)% predicted and LCI: 9.36±2.99.
Statistically significant correlation was found between FEV_1% and BMIp, WAz, BAz and PhA ($p = 0.001$, $p = 0.009$, $p = 0.001$, $p = 0.001$, respectively). LCI was significantly related only with BMIp, ($p = 0.004$).
Conclusions: All the nutritional status parameters seem to correlate significantly with FEV_1 in CF patients. However, only BMI has a significant correlation with LCI.

135 MEF25 as parameter to evaluate effects of mannitol inhalation

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Objectives: In our centre various parameters (absence of side effects, improvement of lung function, increased sputum mobilisation, subjective improvement) are used to evaluate the effect of mannitol inhalation 6 (4–8) weeks after start of therapy. If no positive effect is seen, mannitol inhalation is discontinued. However, the results for various lung function parameters are not always congruent. We investigated differences between FEV1 and MEF25 results.
Methods: After initiation dose assessment patients were treated with inhalative mannitol 400 mg twice daily. After 6 (4–8) weeks effects of therapy were evaluated by medical history and whole body plethysmography. Complete data could be collected from 18 patients. Results for lung function parameters before and after introduction of therapy were compared by Wilcoxon matched-pairs signed ranking test. $p < 0.05$ was chosen as level of significance.
Results: In contrast to previous clinical studies with larger groups of patients we did not see a significant improvement of FEV1 after introduction of therapy (before mean 60.5%, 95% CI 49.7–71.3%, median 56.4%; after mean 61.6%, 95% CI 49.9–73.2%, median 60.2%; $p = 0.79$, n.s.) in our patients. Only 5/18 showed an improvement of FEV1 >10%. However, MEF25 improved significantly (before mean 18.9%, 95% CI 12.3–25.5%, median 15.3%; after mean 26.1%, 95% CI 16.9–35.4%, median 22.8%; $p < 0.005$). 6/18 patients showed no improvement of both FEV1 and MEF25.
Conclusion: MEF25 may serve as an additional, more sensitive parameter to evaluate the effects of mannitol inhalation.

136 Nocturnal oximetry in children with cystic fibrosis (CF)

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Objectives: Intermittent nocturnal observations may miss oxygen desaturations occurring during REM sleep. In our unit, nocturnal oximetry is performed on the first night of admission for all CF children. Night-time monitoring is discontinued if oxygenation is normal and the patient is improving. We aimed to determine whether daytime physiology is predictive of nocturnal hypoxaemia.
Methods: A retrospective comparison of nocturnal oximetry, daytime physiology and $HbA1_c$ of all CF children admitted in the past year.
Results: 76 children with CF were admitted, 21/76 elective and 55/76 acute admissions. Table 1 summarises the clinical characteristics and nocturnal oximetry results. There was a weak positive correlation between Mean SpO_2 and admission FEV_1 ($p \leqslant 0.0001$, r^2=0.27) and admission FVC ($p = 0.0003$, r^2=0.18). Nocturnal oxygen was commenced in 4/76 children (5.2%) as their mean SpO_2 <93%. All of them had an FVC <60%. 3 of the children improved, 1 died.
Conclusion: Patients with an admission FVC of <60% warrant close monitoring for nocturnal hypoxaemia. In those who are cardiovascularly stable, performing an oximetry study on their first night of admission may be less disruptive than routine observations for the whole admission.

Table: Clinical characteristics and nocturnal oximetry results

Age (years)	median [range]	12 [0.1–17.5]
Admission FEV1 (% predicted)	mean [95% CI]	75 [70.67–79.4]
Admission FVC (% predicted)	mean [95% CI]	84 [80–88.4]
Best FEV1 in past year (% predicted)	mean [95% CI]	89.2 [85.4–93]
Best FVC in past year (% predicted)	mean [95% CI]	97.7 [94.3–101]
Mean SpO_2 (%)	median [range]	97 [91.4–99.7]
Desaturations ⩾4%/hour	median [range]	1.1 [0–10.05]
Percentage time O_2 saturations <90%	median [range]	0.02 [0–10.9]
HbA1c (%)	median [range]	5.7 [4.2–7.1]