151* Use of the Northern X-Ray scoring system over time in adult CF patients

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CF is a progressively destructive disease, and the longitudinal assessment of clinical status is pivotal in managing individuals with the condition. Clinical scoring systems can aid this process, and one commonly used in the UK (the Northern Score) was introduced in 1994 and until recently this formed part of the annual data recorded for every patient on the UK CF Trust database. However, although it has been shown to have a good correlation with clinical parameters in cross sectional studies, there are no data indicating its value over time in individual adult CF patients. To assess this further, we compared the Northern score with spirometry (FEV1%predicted), nutritional state (BMI) and IV antibiotic use (days/year) in a group of adult CF patients attending our unit over a 7 year period between 1999 and 2005. Continuous data was available for 100 patients for at least 5 years over this time. Relationships were explored both cross-sectionally and longitudinally using the SPSS v.12 statistical package.

As expected, for cross sectional data, there was a significant correlation between Northern score and IV antibiotic use ($r=-0.397$), BMI ($r=-0.331$), and FEV1%predicted ($r=-0.662$) (all $p<0.01$). For longitudinal data over a 5 year period, there was a similar correlation between the change in Northern score and deterioration in clinical parameters (BMI $r=-0.209$, $p<0.05$; FEV1%predicted $r=-0.389$, $p<0.01$; IV antibiotic use $r=0.313$, $P<0.01$).

Thus, we have shown that the Northern score can be used to support the clinical assessment of changes in physical state in adult CF patients over time. Although it is now no longer required for the annual returns in the new UK CF Trust database, clinicians may wish to continue its use as a valuable clinical tool.

152 Relationship between HRCT score, lung function and clubbing in patients with cystic fibrosis (CF)

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Introduction: Patients with CF develop progressive lung disease as a result of persistent infection and airway inflammation. CT abnormalities are variable and include bronchiectasis, peribronchial wall thickening, mucous impaction and small airway disease. The aim of this study was to evaluate whether routine clinical parameters could predict the extent and pattern of lung disease seen on HRCT scan.

Method: Over the past 3 years patients attending the Leeds Adult CF unit have undergone routine chest CT scan to assess disease severity. Scans were scored for a bronchiectasis (BS) and small airways disease (SAS). Data for FEV1, FVC, FEF 25–75, body mass index (BMI), severity of clubbing, intravenous (IV) antibiotic courses, 6 minute walking test and pancreatic status were retrospectively collected. Statistical analysis was performed using Spearman correlation and Mann-Whitney tests (SPSS 12.0). $P<0.05$ was regarded as statistically significant.

Results: 69 consecutive HRCT scans were scored, median age (range) 22.7 years (16.8–49.9). BS significantly correlated with BMI, FEV1, FVC, FEF25–75 and severity of clubbing. SAS had a significant, but weaker correlation with FEV1, FVC and FEF25–75, and demonstrated no relationship to BMI or clubbing. The number of days of intravenous antibiotics following HRCT and 6 minute walk distance only significantly correlated with BS. CFRD and pancreatic insufficiency were associated with higher BS and SAS ($P<0.05$).

Conclusion: The best clinical predictors of bronchiectasis on HRCT would seem to be clubbing, poor BMI, reduced 6 minute walking distance and a greater requirement for IV antibiotic therapy. Spirometry measures do not seem to be able to predict the presence of small airways disease.

153 Comparison of lung function tests (LFT) in infants with CF and healthy subjects (HS)

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The mechanism underlying the onset of lung disease and early respiratory morbidity in CF infants is poorly understood. The events in early life exert an important influence on the outcome. Infant LFT are a non-invasive and reproducible technique that can give precious data about the presence of bronchial obstruction.

Aim: evaluation of tidal breathing flow-volume loops and FRC in asymptomatic infants with CF in comparison to HS, performed with a respiratory function testing device (Exhalizer® EcoMedics).

Methods: we studied 41 CF infants (23 males), aged 28.1±15.3 weeks and 37 HS (21 males), aged 25.5±10.2 weeks; p = n.s. Tidal breathing parameters were measured: RR (respiratory rate), VT/kg (tidal volume pro kg), Tptef/Te (ratio of time to reach peak espiratory flow in relation to total expiratory time). FRC was obtained with sulfur-hexafluoride (SF6) washin-washout technique using an ultrasonic flow-meter. LFT were performed during induced sleep (Chloral Hydrate 10% 50–75 mg/kg os). Results are shown in the table.

We observed that mean values of Tptef/Te were significantly lower in CF infants than in HS ($p=0.025$) and mean values of FRC/kg were highest in CF patients ($p<0.0001$).

Conclusions: air trapping and early airways obstruction are present in CF subjects but not in healthy infants. Our findings suggest the presence of early functional abnormalities in airways function in asymptomatic CF infants.

	CF (Mean±SD)	HS (Mean±SD)	P
RR (br./min)	53.8±23.9	52.8±12	ns
VT (ml/kg)	7.53±1.45	6.9±1.2	ns
Tptef/Te (%)	21.9±12.4	28.6±13.5	<0.025
FRC/kg (ml/kg)	28.5±9.2	19.2±3.2	<0.0001

154 Home monitoring using portable spirometry and SMS reminder service in CF

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Home monitoring using portable spirometers has gained increasing interest. The aim of this pilot study was to evaluate Spirotel® in monitoring CF patients.

At home 4 CF patients (age 12–19 yrs) recorded flow volume curves and symptoms daily during 6 months. All were chronically colonized with *P. aeruginosa* and inhaled TOBI (28 days on, 28 days off). FEV1 and symptoms such as cough, sputum production, dyspnea, sleep disturbance and wheeze, scored as low/medium/high, were evaluated. Cumulated scores of symptoms were calculated for each day. Slope of regression curve of FEV1 data versus time over 6 months was calculated. Slope of regression curve of FEV1 over sliding periods of the past 14 days were daily determined ('daily slope of FEV1'). Data were transferred each week by internet using home telephone and internal modem and using a cable during a medical visit. Compliance to TOBI inhalations was evaluated by SMS medication reminder service. Patients were alerted by SMS on day 1 (start tobi) and day 28 (stop tobi) during cycle ON TOBI. On day 28 patients send by sms number of vials not taken. FEV1 change over 6 months ranged between −255 to +219 ml/year or from −13 to +13% predicted/year. Daily cumulated number of symptoms was 3–16. No correlation was found between daily cumulated number of symptoms and 'cumulative daily slope of FEV1'. Between 0 and 7 vials each cycle of TOBI were forgotten.

Conclusion: Spirotel® allows home monitoring of lung function on a daily basis (Telemedicine). No correlation was found between cumulative 14 days FEV1 and reported symptoms. SMS reminder service is an elegant system to remind patient of taking TOBI.

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