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 status (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.

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 precise 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® EcoMedicals).

Methods: we studied 41 CF infants (23 males), aged 28±10.5 weeks and 37 HS (21 males), aged 25±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 expiratory flow in relation to total expiratory time). FRC was obtained with sulfur-hexafluoride (SF6) wash-in/wash-out 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.

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 inhales 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 +219ml/year or from −13 to +135% 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.

Supported by a grant of Solvay Pharma.