223 Outcome of care for home management with intensive input in adult CF patients during pulmonary exacerbations (PExs) – a comparative prospective study with hospital care

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Background: Evidence of an inferior outcome of care for patients treated at home in comparison with those treated in hospital is a cause for concern. Reasons for the poorer outcome include reduced support at home from the multidisciplinary team (MDT) in comparison to hospital care. A new intensive home treatment service was organised mimicking hospital care, in which patients on IV treatment are visited at home twice weekly by a CF physiotherapist and nurse, who assess patients, spirometry and weight, reinforce compliance, the importance of rest and good diet.
Patients and Methods: We prospectively compared the outcome of PExs treated with intensive home input by the MDT with that of hospital care and of the standard home care without intensive assistance. Treatment site was decided as a consensus between the MDT and the patient. Outcome measures analysed were changes in spirometry, weight and symptom score from day 1 to day 14 of treatment and time until the next PEx needing IV treatment.
Results: 180 PExs in 58 patients were analysed. There were no differences in age, gender or baseline spirometry between the groups, but baseline BMI was higher in the new home service group ($P = 0.008$). FEV1 and BMI improvement were significantly greater in the hospital group in comparison to the home with input group ($P = 0.01$ and $P = 0.02$ respectively) and with the standard home group ($P = 0.02$ for FEV1 and $P = 0.03$ for BMI). The time until the next PEx was similar in all groups.
Conclusions: Despite extensive efforts to improve home treatment, outcome measures remained better for those treated in hospital. The impact of this study is important for the organisation of care of CF exacerbations.

224 Predictors of the time until the next pulmonary exacerbation in adult CF patients

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Introduction: Acute infective pulmonary exacerbation (PEx) is an important feature of CF. PExs are common and associated with impaired quality of life and accelerated decline in lung function. It is therefore goals of optimal CF management not only the treatment of such infections, but also the prevention of PExs and to increase the time between exacerbations. We aimed to assess factors associated with a shorter time until the following PEx.
Patients and Methods: We prospectively analysed 181 PExs in 58 adult CF patients (32 male, mean age 26 years) treated with intravenous (IV) antibiotics. PExs were methodically diagnosed according to a combination of subjective and objective parameters. Outcome measures on day 14 of treatment were documented, including FEV1, FVC, PEFR, FEF25–75, BMI, C reactive protein (CRP) and symptom score (SS). The days until the following PEx needing IV antibiotics were calculated.
Results: Lower spirometry values at the end of PExs were associated with shorter times until the following exacerbation, $P < 0.001$ for FEV1, FVC and PEFR. Higher symptom scores were also associated with shorter times until the following PEx ($P = 0.02$). Measurement of small airway disease (FEF25–75), CRP and BMI at the end of treatment of PEx did not predict the time until the following exacerbation.
Conclusions: Poorer lung function tests and greater symptoms were associated with a shorter time until the following PEx. BMI and CRP measurements did not predict the time until the following PEx. These results have an impact on the outlook of patients when planning their care in CF centres.

225 Symptomatic sinus disease in cystic fibrosis and its effect on lung function

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Introduction: Structural and functional abnormalities of sinuses are common in CF but not all patients are symptomatic. It is thought that sinus disease may be associated with increased lower respiratory tract pathology.
Methods: From our cohort of adult CF patients we identified patients labelled to have symptomatic sinus disease at annual review. We obtained following data from Port CF: baseline FEV1, decline in FEV1 over 1 year, sputum microbiology and presence or absence of ABPA. Full data was available for 106 patients.
Results: We identified 50 patients with symptomatic sinus disease and 56 patients without. The baseline characteristics are shown in the table. The baseline FEV1 was statistically greater in the symptomatic group compared with asymptomatic (2.34 vs. 1.92). There was a trend towards greater deterioration in FEV1 in symptomatic group.

Baseline characteristics and results

	Sinus disease		Total (%)
	Symptomatic (%)	Asymptomatic (%)	
n	50 (47.17)	56 (52.83)	106
Male	23 (46)	29 (51.78)	52
Female	27 (54)	27 (48.22)	54
Mean age	29.62	29.07	29.34
Mean FEV1	2.34	1.92, $p = 0.019$	2.13
Drop in FEV1	0.20 (8.78)	0.06 (3.3), $p = 0.064$	0.13 (6.31)
Pseudomonas	40 (80)	50 (89.2)	90 (84.9)
NTM	5 (10)	1 (1.78)	6 (5.66)
ABPA	4 (8)	10 (17.85)	14 (13.2)

Discussion: These results show that symptomatic sinus disease is associated with less severe lung disease. Other investigators have previously shown that patients with visible nasal polyps have better lung function than age matched peers. Larger studies are needed.

226 Obstructive sleep apnea and nocturnal hypoventilation in adults and children with cystic fibrosis

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Chronic airway obstruction may determine early hypoxaemia during exercise and sleep even in patients with normal diurnal blood gases. The aim of our study was to establish the occurrence of sleep-disordered breathing (SDB) in a cohort of patients with cystic fibrosis (CF), followed in our Cystic Fibrosis Unit. We therefore studied 55 patients (49% M; 36% children (mean age 4.3 ± 3.9, SaO_2 % awake 97.7 ± 1.5); 64% adults (mean age 20.9 ± 4.9, BMI 21.1 ± 3.6, SaO_2 % awake 97.1 ± 1.1). All patients, in stable clinical conditions, underwent a nocturnal standard polysomnography (sleep monitoring system Compumedics S-Series) and respiratory functional evaluation during the day. Scoring of respiratory events was performed according to standard criteria. Polysomnographic monitoring revealed the occurrence of SDB, defined as an apnea-hypopnea index (AHI) > 5, in 52% of adults and in 56% of children. In adults we found mean AHI 16.8 ± 7.8, mean nocturnal SaO_2 $93.9\% \pm 2.3$, mean SaO_2 min $88.2\% \pm 2.2$; in children we found mean AHI 12.5 ± 5.8, mean nocturnal SaO_2 $93.7\% \pm 1.2$, mean SaO_2 min $89.2\% \pm 3.2$. In both adults and children we found a similar prevalence of upper airways obstruction (sinusitis and rhinitis), a major cause of SDB, in groups with SDB and without SDB. In adults we found a lower mean FEV1% in the group with SDB as compared to the group without SDB (50% vs 64%, respectively).
These data show an high prevalence of sleep-disordered breathing in patients with cystic fibrosis. In these patients SDB occurs in an early phase of life and in adulthood SDB seems to be associated to a lower respiratory function.