Ten patients (none with cenocepacia) are using inhaled antibiotics (3 colistin, 1 Colobreath, 2 TOBI, 2 Cayston, 1 ceftazidime, 1 alternating Cayston and Promixin).

Conclusion This study shows that a significant proportion of our Burkholderia spp infected patients have organisms that are sensitive to currently available inhaled antibiotics. Given our positive experience, and with the expected availability of new inhaled antibiotics in the near future, perhaps the time has come to formally look at the use of inhaled anti-microbial therapy in this small but important cohort of CF patients.

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INVESTIGATING THE ROLE OF CHEST PHYSIOTHERAPY IN THE COLLECTION OF SPUTUM SAMPLES FROM INDIVIDUALS WITH CYSTIC FIBROSIS (CF)

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Introduction Chronic respiratory infection is responsible for the majority of the morbidity and mortality of CF patients. In order to guide treatment regimes and improve understanding of the pathophysiology of CF, airway secretions are sampled and analysed. Sputum is usually the selected method of sampling.

Often, large quantities of sputum are required to facilitate comprehensive laboratory testing. Hence, when designing studies, it is important to consider the quantity of sputum likely to be produced by patients and to ensure that the composition of the sputum samples is not altered by the procedure by which they are obtained.

This study aimed to investigate the effect of chest physiotherapy on the quantity and composition of sputum samples collected from individuals with CF. It was hypothesised that physiotherapy would increase the quantity of sputum produced, reduce the salivary content and alter the microbiological content. Methods Clinically stable adults with CF were recruited at outpatient clinics and randomised into group A (physiotherapy group, n = 21) or group B (no physiotherapy group, n = 25). Laboratory processing of the samples involved determining sample weights and counting human cells (alive respiratory cells, dead respiratory cells and squamous cells). The dissolved sputum was also transferred onto plates of cetrimide agar for culturing *Pseudomonas aeruginosa*. Colony-forming units (CFUs) were counted on the plates after 24 h.

Results Samples from the physiotherapy group had significantly greater weights than the no physiotherapy group (p < 0.001). When considering the total number of cells per gram of sputum, there was no statistical difference between the two groups (p = 0.396). However, the numbers of squamous cells per gram, and dead respiratory cells per gram were both significantly greater in the no physiotherapy group (p = 0.039 and p = 0.001 respectively). There were no significant differences between numbers of alive respiratory cells per gram (p = 0.487) or CFUs (p = 0.459).

Conclusion Whilst physiotherapy was found to increase the quantity of sputum collected, there were significant differences in sputum composition, suggesting that the two groups represent samples from different niches. Hence, when planning a study involving sputum analysis, the procedure by which the sample is obtained has to be considered when interpreting the results.

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A PROSPECTIVE COHORT STUDY OF INTEGRATED PALLIATIVE CARE OF CYSTIC FIBROSIS (CF)

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There are 140 deaths in the UK each year from CF, often on a transplant waiting list and often without specialist palliative care. A palliative physician and nurse joined our team in 2011, providing palliative care in parallel with standard CF care. We undertook a prospective study documenting symptoms and outcomes, the views of the CF team and the experience of the palliative specialists.

Over 3 years, 28 (10%) of 282 patients at our Centre had palliative input; their mean age was 31 (range 18-47) years and mean FEV1 was 0.86 L (24%); 17 (61%) died - 6 were on a transplant waiting list, 10 were unsuitable, and one died post transplantation; 4 have had transplantation and no longer need palliative input, 7 are in on-going care; 15 (88%) of deaths were on the CF ward and 2 at home. All patients who died had had palliative care. The main symptoms were breathlessness, cough, pain, vomiting, fatigue and low mood. The mean palliative assessment score was high at 2.9, indicating that life was dominated by symptoms. Palliative interventions included opioid, benzodiazepine, anti-emetic and anti-depressant medications and non-pharmacological interventions included relaxation techniques, massage, acupuncture and cognitive therapy. A survey was completed by 16 members of the CF team: all felt that palliative specialists should be part of the team and rated the model of care highly with a mean score of 4.1 (scale 1-5); 11 thought that patients had found input very helpful and 5 helpful; one patient declined a palliative consultation. The palliative specialists had increased their knowledge of CF, found it useful to meet patients earlier and had no difficulty in providing palliation in parallel with standard CF care. Their workload was high and they identified additional needs of bereavement counselling and managing the effects of deaths on other CF patients.

This integrated model was successful in overcoming barriers to specialist palliative care. Palliative specialists have improved their knowledge of CF and the CF team have learnt palliative skills

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TOO SWEET FOR TOO LONG?

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Background Cystic fibrosis related diabetes (CFRD) is associated with deterioration in clinical status. Lung function and nutritional status deteriorate up to 2–4 years *before* a diagnosis of CFRD based on the oral glucose tolerance test (OGTT). Timely detection and treatment is crucial.

Aims To evaluate:

- · adherence to CFRD screening guidelines and
- whether identifying stages of progressive Cystic Fibrosis Insulin Deficiency (CFID) using the extended OGTT altered management
- trends in weight, BMI and FEV1 in CFRD as compared to CF controls.

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Methods Retrospective analysis using patients' records. 7 patients with CFRD were compared to matched CF controls using mean z-scores for weight, BMI and FEV1.

Results Records of 59 children (23 males) were analysed, 21 children between 5–10 years and 38 >10 years. In the younger group, 80% (n = 17) had both HbA1c and random glucose tested as per our guidelines. Of 38 patients aged >10 years, 78% (n = 30) were screened by OGTT of whom 16% (n = 5) had the standard test. Table 1 summarises the results and shows the degree of glucose impairment on OGTT and the related grade of cystic fibrosis insulin deficiency (CFID).

Abstract P232 Table 1 The degree of cystic fibrosis insulin deficiency (CFID) in patients (>10 years) undergoing the extended OGTT

N (%)	Result	Glucose in mmol/L	
		Peak	2 hour
6 (20)	Normal	120	2
11 (36)	CFID1	≥8.2	<11.1
6 (20)	CFID2	≥11.1	<11.1
7 (23)	CFID3	<7	≥11.1
0 (0)	CFID4	≥7 with fasting hyperglycaemia	6

The mean weight and BMI z scores for those with CFRD compared to controls were -0.64 vs -0.02(p = 0.005) and -1.26 vs -0.03(P = 0.0001). There was a lower trend in FEV1 in CFRD, 1.87l (73.06%) vs 2.35l (89.03%). 3 patients with CFID3 and 1 with CFID1 later commenced insulin based on clinical grounds.

Conclusions Adherence to screening guidelines needs to be improved. Patients with CFRD have a significant declining trend in weight, BMI and FEV1 compared to controls. Some patients with CFID were commenced insulin on clinical grounds rather than results of extended OGTT. Whether treatment at earlier stages of CFID will slow down the rate of decline needs to be explored, but we have reverted back to the standard OGTT for the present.

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COUGH SWABS SHOULD NOT BE USED TO EXCLUDE NON-TUBERCULOUS MYCOBACTERIAL (NTM) INFECTION IN ADULTS WITH CYSTIC FIBROSIS

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Introduction and objectives People with cystic fibrosis (CF) are prone to airway infection with non-tuberculous mycobacteria (NTM) including *M. abscessus*, *M. avium* and *M. intracellulare*. Routine NTM screening is recommended for all patients at least once a year. In patients not able to produce sputum, cough swabs are often sent for NTM analysis. Anecdotally, we observed that several patients cared for in our centre had cultured NTM in sputum having previously been culture-negative from cough swabs. The objective of this observational study was to examine the diagnostic yield of cough swabs and sputum samples from CF adults with known NTM infection.

Methods We identified all CF patients being cared for in our large regional adult CF centre that had cultured any NTM species from cough swabs or sputum samples and are currently attending our NTM clinic. Demographics, clinical parameters and microbiology results were recorded and analysed.

Results 26 patients (19 male) were included: median age 24 years, 92% chronically infected with *P. aeruginosa*, 100% pancreatic insufficient, 62% CF-related diabetes, 27% ABPA. 381 sputum samples and 55 cough swabs were analysed. 251 (66%) sputum samples and 4 (7%) cough swabs cultured NTM (see Table 1). In the 4 cough swabs that cultured NTM, sputum samples also cultured the same species.

Conclusions Cough swabs have a very low diagnostic yield and their use did not contribute to identification of NTM infection in our adult CF patient population. We have therefore stopped sending cough swabs for NTM culture in our centre and our data suggests that cough swabs should not be used to screen for these organisms.

REFERENCE

1 Mycobacterium abscessus Suggestions for infection prevention and control. CF Trust, 2013

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PREVALENCE OF NON-PULMONARY COMPLICATIONS FOLLOWING LUNG TRANSPLANTATION IN ADULT PATIENTS WITH CYSTIC FIBROSIS (CF)

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Background Lung transplantation is currently the most effective means of improving quality of life and survival in patients with end stage CF. Improvements in surgical technique, lung preservation, immune suppression and infection management have improved short and long term mortality and morbidity.

The number of significant medical complications encounter following lung transplantation can have significant impact on long term management of CF.

Aim To identify the frequency of medical complications that occur in a large cohort of post-transplant CF patients.

Method Retrospective review of electronic records to assess complications in all patients with CF who underwent Lung transplantation between September 1992 and June 2015.

Results 54 patients underwent lung transplantation (heart/lung -3, lung/liver -1) at different transplant centres, (female-35, male-19 current median age 36 years (range,22–66), current 10, 15 and 20 year survival rates are 43%, 22% and 7% respectively (median 87 months). Complications are shown in Table 1.

9 (17%) post-transplant patients died (median survival 77 months). Of these, 3 (33%) died secondary to malignancy.

Conclusion In addition to organ rejection and infective causes common systemic complications included hypertensive disease (50%), gastro-oesophageal reflux disease (30%), chronic kidney disease (26%), and osteoporosis (19%). It is notable that one third of patient mortality was due to malignancy. As patient survival improves we may need to consider increased screening of these high risk patients. Frequent monitoring and excellent collaboration between transplant and CF centres may lead to earlier detection and treatment of these complications.

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