different mix of patients in the CF registry than the US insurance based health system despite the US CF foundation's attempts to reduce barriers to care. This study compared registry and routine deaths in UK and USA.

Methods UK and US routine deaths for ICD-10 E84 are provided in 5 year age bands and the age of death was taken as the midpoint age in the band. Medians were calculated with STATA.

Results US routine data are not yet published for 2007 onwards. The Abstract P236 table 1 shows total deaths and median age at death derived from routine and registry data for USA and UK.

Abstract P236 Table 1

Year	UK Total deaths routine	UK median age at death registry/ routine	UK registry Total deaths (n) (%) missing	US total deaths routine	US median age at death registry/ routine	US registry total deaths (n) (%) missing
2009	147	27/27	141 (6) (4%)	NA	26.1/NA	440 (NA) (NA)
2008	122	27/27	100 (22) (18%)	NA	26.3/NA	420 (NA) (NA)
2007	133	24/27	106 (27) (20%)	NA	25.9/NA	394 (NA) (NA)
2006	112	NA/27	NA	438	25.2/27	362 (76) (21%)
2005	108	NA/27	NA	414	NA/27	NA
2004	137	26/27	123 (14) (10%)	461	24.2/27	358 (103) (29%)
2003	117	24/24	136 (33) (24%)	447	25.4/27	370 (77) (21%)
2002	118	NA/24	NA	485	25.2/27	426 (59) (14%)

CF deaths in UK and US 2002 to 2009.

Discussion Both the US and UK have around 20% of CF deaths occurring outside centres though in 2009 in the UK this fell to 4%. In 3 out of the 5 years that allow comparison the median age at death was higher in UK registry data compared to the USA. Contrary to expectation the median age at death derived from routine data are higher than the age of death from registry data in all years where comparison is possible. Interestingly, in the US routine data the distribution were different to the UK with many more very old (over 70) patients. The increased age at death in routine data may in part reflect the imprecision related to the routine data only providing age at death in 5 year bands. The apparent higher proportion of very elderly in the US is intriguing and invites further study.

P237 DOES DESATURATION ON EXERCISE PREDICT NOCTURNAL HYPOXIA IN CHILDREN WITH CYSTIC FIBROSIS?

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Introduction Exercise induced arterial hypoxia (EIAH) is well documented in cystic fibrosis (CF); though its relationship with nocturnal hypoxia remains unclear. In adult CF, hypoxia occurs more frequently during sleep than exercise,[1 2] but this has not been investigated in children.

Aim To identify whether EIAH is a reliable predictor of nocturnal hypoxia in CF children.

Methods Retrospective case note review of 34 children with CF. All performed the modified shuttle test,[3] overnight pulse oximetry and spirometry in the second week of an elective admission for intravenous antibiotics. EIAH was defined as drop in SpO_2 ≥4% from baseline.[4] There is no agreed definition of nocturnal hypoxia, we assessed numerous overnight variables, mean and lowest SpO_2 and % time with SpO_2 <92% in sleep. Data were analysed using SPSS software (SPSS).

Results 34 subjects were assessed, median age 12 (range 7–16) years, and FEV_1 72 (25–103) % predicted. Nine demonstrated EIAH and five had nocturnal desaturation (SpO_2 <92% >10% sleep time). All children with SpO_2 <92% for >10% of sleep also had EIAH, this was significant (p=0.0086). The relationship between nocturnal hypoxia and lowest exercise SpO_2 (p=0.0076) was also significant. However, 6 of those with EIAH were not hypoxic overnight. The sensitivity of EIAH detecting sleep hypoxia was 75% and specificity 80%. Positive predictive value (PPV) was 33% and negative predictive value 96%.

Conclusion The PPV of EIAH for detecting nocturnal hypoxia is poor, but the definition of EIAH must be questioned. SpO_2 varying from 90% to 87% does not meet EIAH criteria, yet a fall from 97% to 93% does. We must also note that although the relationship between nocturnal hypoxia and EIAH appears significant the number of subjects in this group is extremely small. We conclude EIAH is not a reliable predictor of nocturnal desaturation and advocate a sleep study to confidently detect sleep hypoxia.

Abstract P237 Table 1

Sleep Exercise	SpO_2 < 92% for <10% of sleep n=32	SpO_2 <92% for >10% of sleep n=3	p Value	Drop in SpO_2 <4% n=11	Drop in SpO_2 ≥4% n=23	p Value
Baseline SpO_2	97 (87–99)	94 (89–97)	0.1449	97 (94–99)	97 (87–99)	0.5397
% Change in SpO_2	1 (−5–7)	10 (4–13)	0.0128	1 (−6–3)	2 (−11–18)	0.294
Number with EIAH	6 (16%)	3 (100%)	0.0086	1 (9%)	6 (26%)	0.3844
Lowest SpO_2	95 (88–100)	84 (79–90)	0.0076	95 (93–99)	95 (79–100)	0.204

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P238 PRESCRIPTION ISSUE DATA AS A MEASURE OF ADHERENCE WITH NEBULISED THERAPY IN AN ADULT CYSTIC FIBROSIS CENTRE

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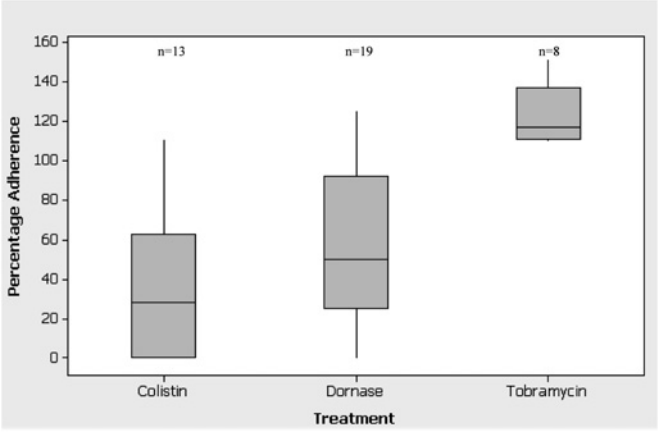
Introduction and Objectives Patients with cystic fibrosis are often prescribed complex time-consuming medication regimens. We know from studies in chronic diseases that most patients will be at best only partially adherent with their medication. We studied adherence with nebulised therapy in our adult CF clinic, by measuring the actual quantity of medication prescribed as recorded in General Practitioner and hospital computer prescribing records. The results were compared with the CF centre staff's predictions of adherence on an individual patient basis.

Methods We requested prescription issue data for 26 patients on nebulised dornase α, colistin and tobramycin both from the patient's general practitioners and from the hospital pharmacy. We compared the actual quantity of medication prescribed to each patient to the expected number of doses that they should have received during a 12-month period and calculated a percentage adherence based on these figures. Based on previous work in our difficult asthma clinic we assigned obtaining =80% of doses as being adherent; =50% but <80% as partially adherent and <50% as non-adherent with the medication.



Abstract P238 Figure 1 Percentage adherence with nebulised treatments in cystic fibrosis.

Results Full prescription data were received for 22 of the 26 patients included in the study. Of these 15% (2/13) were adherent; 23% (3/13) were partially adherent and 62% (8/13) were non-adherent with nebulised colistin. 26% (5/19) were adherent; 26% (5/19) were partially adherent and 48% (9/19) were non-adherent with dornase α 100% (8/8) patients were adherent with nebulised tobramycin. The cystic fibrosis team correctly predicted only 40% (111/280) of the respective adherence rates found.

Conclusions The use of prescription data provides useful information regarding adult CF patients' adherence with high-cost nebulised therapy. In our clinic adherence rates were low, and CF clinicians were poor at predicting adherence rates in individual patients. Used in isolation prescription data are a useful indicator of non-adherence, but does not accurately measure positive adherence without consideration of other methods of measurement. We are currently extending this study to compare pharmacy issue data with other methods of measuring adherence in adult CF patients.

P239 RADIATION EXPOSURE IN ADULTS WITH CYSTIC FIBROSIS

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Introduction Patients with CF can expect to undergo many investigations using ionising radiation for the management of their disease: now that the majority of patients will survive well into their 5th decade, life-time exposure to such radiation, a known risk factor for the development of malignancy, may be important. To investigate this further, we looked at the amount of ionising radiation given in our adult CF clinic over a 1-year period.

Methods All imaging studies associated with ionising radiation in 253 adult CF patients were reviewed and assessed for their impact on management. Radiation was calculated using standard reference doses[1–3] and expressed as (milliSievert [mSv]), also referred to as the effective dose.

Results A summary of the results can be found in the Abstract P239 table 1. There was an average annual radiation dose of 1.66 mSv with 54% of studies leading to a change in management. Overall, although only 11% of chest x-rays altered patient management, patients with more severe CF disease (DIOS, CFRD and infection with transmissible Pseudomonas strains) had a greater cumulative radiation dose and this was more likely to alter management.

Conclusions Although the CF population receives a significant dose of radiation from medical investigations each year, many impact on patient management. CFRD, DIOS and infection with transmissible Pseudomonas are associated with greater levels of radiation than the average CF population, in keeping with more significant disease burden in these individuals. Care should be taken when ordering investigations associated with ionising radiation, to reduce the long term effects, as life expectancy is increasing.

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P240 PNEUMOTHORAX MANAGEMENT IN CYSTIC FIBROSIS PATIENTS

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Introduction Spontaneous pneumothorax is a well-recognised complication in cystic fibrosis and is associated with increased morbidity and mortality. Although chemical pleurodesis and surgery are successful in preventing recurrence in non-CF patients, there are few data regarding their efficacy in CF.

Method To look at this further we reviewed the management and outcome of all 28 patients (mean age at first pneumothorax 26 years [range 18–54], mean FEV_1 32% predicted [17–68], 18 female) in our centre who had developed a pneumothorax (58 episodes [1–7]) from 1993 to 2010.

Results Nine sustained bilateral pneumothoraces (either at initial presentation or as a subsequent event) and 16 recurrence (mean interval 4 months [1–12]). The outcomes of the initial and second line treatments are shown in the Abstract P240 table 1. 14 patients (50%) died within 12 months and 20 (71%) within 2 years of their first pneumothorax, and five deaths occurred after surgery (mean 10 days [3–21]). The highest risk of recurrence occurred in the chest drain group (62%) followed by chemical pleurodesis (with 4 g talc) (57%), observation (40%) and the surgical group (20%).

Abstract P239 Table 1 Radiation exposure in cystic fibrosis

	Mean % predicted FEV_1 [SD]	Mean CXR [% impacting care]	Mean CT thorax [% impacting care]	Mean HRCT [% impacting care]	Mean Radiation Dose (mSv) [% impacting care]
All patients (n=253)	75.7 [23]	2 [10.7]	0.04 [40]	0.03 [37.5]	1.66 [54]
CFRD (n=102)	66.2 [22.6]	2.58 [8.7]	0.05 [60]	0.04 [50]	2.62 [55]
DIOS (n=5)	88.3 [28.5]	4.2 [9.5]	0.2 [0]	0	6.83 [31]
Transmissible Pseudomonas (n=109)	70.7 [22.4]	2.4 [9.8]	0.05 [40]	0.04 [50]	2.39 [52]
Other Pseudomonas (n=75)	77.4 [25.8]	1.8 [10.4]	0.03 [100]	0.03 [50]	1.12 [50]
B Cepacia (n=10)	69.7 [17]	1.5 [13.3]	0	0	1.01 [7]