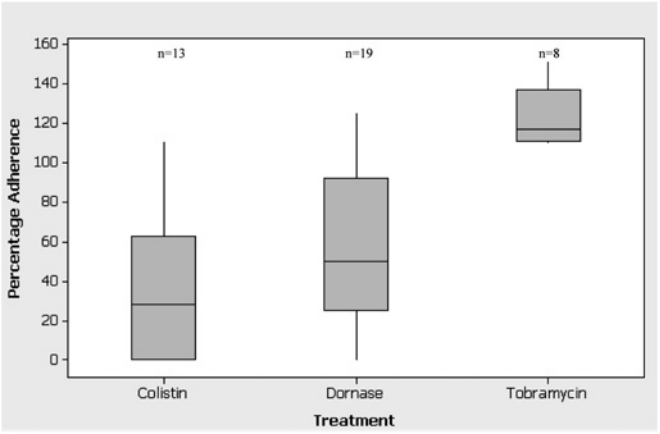

Abstract P238 Figure 1 Percentage adherence with nebulised treatments in cystic fibrosis.

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]

Abstract P240 Table 1 Pneumothorax management in cystic fibrosis patients

Treatment	Episodes of pneumothorax	Resolution rate	Second line treatment	Complications	Late recurrence
Observation	13	8 (62%)	4 (chest drain)	0	4 (40%)
Chest drain	22	13 (59%)	8 (surgery)	4 deaths	8 (62%)
Chemical (talc) pleurodesis	18	6 (33%)	2 (surgery)	1 death	4 (57%)
Surgery	10	5 (50%)	–	5 deaths	1 (20%)

Conclusion Thus, we have confirmed that spontaneous pneumothorax is a poor prognostic factor in CF, with 57% of patients experiencing a recurrent pneumothorax within the first year and poor one and 2-year survival rates. Surgical treatment was associated with a lower risk of recurrence, but at the expense of an increase in mortality. Randomised controlled trials of treatment options and the formulation of guidelines are required to determine the timing and optimal management to prevent recurrent pneumothorax in CF patients.

P241 ACCUCHEK (TM) MEASUREMENT OF BLOOD GLUCOSE CORRELATES WITH TRUE LAB GLUCOSE MEASUREMENT IN CHILDREN SCREENED FOR CYSTIC FIBROSIS RELATED DIABETES

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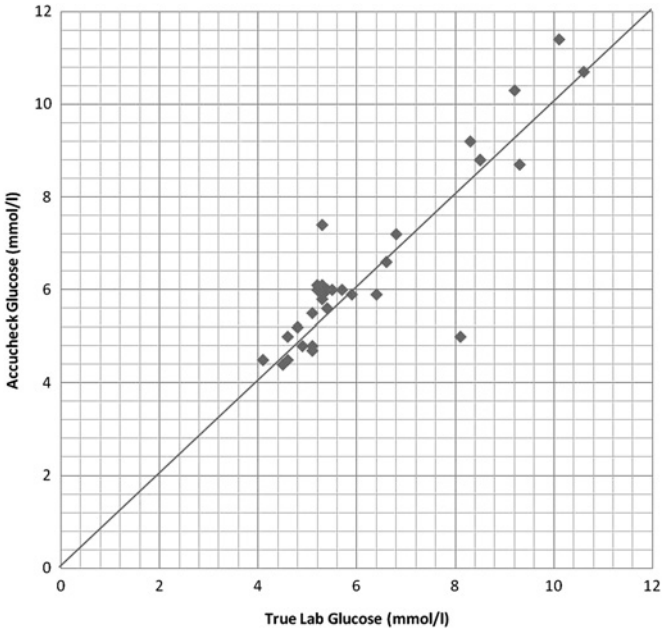
Background Screening for cystic fibrosis related diabetes (CFRD) is recommended as part of the cystic fibrosis annual review. For children over 10 an oral glucose tolerance test (OGTT) is recommended, but this is time consuming and adds to the burden of assessments and investigations of the annual review. As part of a proposed wider pragmatic study to evaluate the feasibility of home OGTT testing in this patient group, we assessed the level of agreement between blood glucose concentration measured by near-patient testing using the Accucheck glucometer and that measured in the laboratory using standard methods (True Lab Glucose—TLG).



Abstract P241 Figure 1

Methods 26 children with CF aged 1 yr—16 yrs undergoing annual review were studied. Blood taken at the time of venepuncture was tested for glucose concentration using an Accucheck glucometer, and results compared with TLG. For children under 10 a single, random blood glucose was tested using each method. For those aged 10 or over samples taken after an overnight fast, and 2 h after a standard OGTT oral glucose load were tested using each method. Bland and Altman statistics were used to assess limits of agreement between methods of glucose measurement.

Results 31 pairs of glucose measurements were obtained. TLG measurements ranged from 4.1 to 10.6 mmol/l. Abstract P241 Figure 1 shows the Accucheck glucose measurement plotted against the TLG for each sample, and the line of equality. The agreement between Accucheck and TLG was investigated using the method of Bland and Altman and showed that the Accucheck gave results a mean (SD) of 0.26 (0.84) mmol/l higher than values obtained on TLG.

Conclusions There is a clinically acceptable agreement between blood glucose measured by Accucheck compared with TLG. Just one child with a TLG result that might have prompted further investigation could have been missed, had Accucheck been the sole method used. This suggests that blood glucose measurement in the context of OGTT screening for CFRD could perhaps be performed using near-patient testing in an out of hospital setting (eg, by parents at home) without compromising test validity. Further research is needed to establish if this is the case.

P242 BONE DENSITY AND TESTOSTERONE LEVELS IN MALE CYSTIC FIBROSIS PATIENTS

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Background Cystic Fibrosis (CF) is the commonest inherited life threatening condition in the UK and is a multisystem disease. CF related low bone mineral density (BMD) is defined as a Z score 2 SDs below the age and gender matched mean reference value and has been reported in 13% of CF patients.[1] Its aetiology is multifactorial with hypogonadism being a potential factor. CF Trust guidelines[2] suggest BMD should be measured and addressed regularly and testosterone levels measured annually in adult males.

Aim To assess the prevalence of low BMD and testosterone in male CF patients attending the All Wales CF Centre.

Methods Annual review records (2008—2009) of BMD derived from DEXA scans and testosterone levels were reviewed retrospectively.

Results 65 male patients (age 18—51 years) were included of whom 60 had a recorded testosterone and BMD. 12 patients (20%) had CF related low BMD (at hip or lumbar spine) of whom three had a low testosterone (<8.0 nmol/l). 12 (20%) patients had low testosterone levels. Of these twelve, three had low leutinising hormone (LH) and nine normal LH and all had normal levels of follicle stimulating hormone.

Discussion CF related low BMD occurs commonly in men with CF with a quarter of those with low BMD also having low testosterone levels. Low testosterone levels per se also occur frequently in this population and recent CF Trust guidelines[2] have suggested an endocrinology opinion should be sought where testosterone is found to be low.

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