The bronchiectasis severity index derived from these models was composed of prior hospitalisation (5 points), MRC dyspnoea score (0–3 points), FEV_1 (0–3 points), bacterial colonisation (0- 3 points) Age (0–6 points) BMI <18.5 (2 points) Exacerbation frequency (0–2 points) and radiological extent (1 point). The AUC for mortality was 0.80 (0.74–0.86) and the AUC for hospitalisation was 0.88 (0.84–0.91). There was a clear difference in exacerbation frequency and quality of life using the St. Georges Respiratory Questionnaire between patients classified as low, intermediate and high risk by the score ($p < 0.0001$ for all comparisons).

In the validation cohorts, the AUC for mortality ranged from 0.81–0.84 and for hospitalisation was AUC 0.80–0.88.

Conclusions The bronchiectasis severity index identifies patients at risk of future mortality, hospital admissions and exacerbations.

S125 A RETROSPECTIVE STUDY CHARACTERISING CILIARY ULTRASTRUCTURE, LIGHT MICROSCOPY AND SPUTUM MICROBIOLOGY ASSOCIATIONS WITH LUNG FUNCTION DECLINE IN A LARGE ADULT PRIMARY CILIARY DYSKINESIA COHORT

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Background Primary ciliary dyskinesia (PCD) is an inherited disease related to ciliary dysfunction, with heterogeneity in clinical presentation, prognosis and ciliary ultrastructure. Our study aimed to comprehensively characterise a large cohort with respect to ciliary ultrastructure, beat frequency, sputum microbiology, mortality and lung function decline.

Method A cohort of 100 adult PCD patients was identified at a tertiary respiratory centre. A retrospective analysis of clinical age at presentation and diagnosis alongside ciliary ultrastructure, nasal nitric oxide, beat frequency, sputum microbiology, lung function at diagnosis and follow-up and mortality were recorded. Non-parametric multi-parameter analysis of variance and Spearman rank correlation statistical analysis was performed to identify significant associations with decline in lung function (FEV1%/year). Median duration of follow-up was 7.5years (range 2–30years).

Results Overall mortality was 4% (median age of death 55years). 12% of patients had a central pair/transposition defect, 37% missing outer dynein arms, 15% missing inner dynein arms, 28% no arms, and 3% had normal ultrastructure. There was no significant correlation between ciliary ultrastructure, beat frequency (range 0–13.9Hz) and nasal nitric oxide with clinical age at presentation (range 1–26 years) and diagnosis (range 1–72 years) or lung function at presentation and decline with follow-up. There was additionally no significant association between sputum isolation including Pseudomonas aeruginosa with lung function decline. 44% of patients had Pseudomonas aeruginosa chronic infection. The incidence of NTM colonisation was low (4%). Aspergillus species colonisation was additionally low (5%). The average lung function decline in the cohort was 1.45% FEV1/year.

Conclusions Comprehensive characterisation of an adult PCD cohort with ciliary ultrastructure, light microscopy, clinical presentation and follow-up data shows a relatively favourable outcome with optimum care. Ciliary ultrastructure, beat frequency and nasal nitric oxide does not predict prognosis. Contrary to parallel diagnoses such as cystic fibrosis and adult idiopathic bronchiectasis, microbiological isolation of Pseudomonas aeruginosa is not associated with a more rapid decline in lung function with optimal prophylaxis and care. Contrary to recent suggestion of low ciliary beat frequency and low nasal nitric oxide association with NTM susceptibility, we did not find a high incidence of NTM or Aspergillus species within this cohort.

S126 MOLECULAR EPIDEMIOLOGICAL ANALYSIS SUGGESTS CROSS INFECTION WITH PSEUDOMONAS AERUGINOSA IS RARE IN NON-CYSTIC FIBROSIS BRONCHIECTASIS

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Background Non Cystic Fibrosis Bronchiectasis (NCFBr) is a cause of significant morbidity and mortality. Pseudomonas aeruginosa, a key pathogen in NCFBr, is associated with premature mortality. Globally, common clones of P. aeruginosa have been recognised from clinical and environmental sources and in Cystic Fibrosis (CF) cross infection is known to occur. There are no robust data on cross infection in NCFBr. This evidence gap impacts on managing patients but was omitted from the BTS 2010 guidelines due to the paucity of data.

Aims To seek evidence of cross infection amongst NCFBr patients.

Methods Single centre cross sectional study: We studied 50 P. aeruginosa isolates from 40 NCFBr patients using two genotyping techniques (both blinded); an Array Tube (AT) method and Variable Number Tandem Repeat (VNTR) analysis. We included known CF clonal strains as internal controls. We then compared the data using genotype databases.

Results This is the largest cross infection study to our knowledge. We demonstrated that shared P. aeruginosa NCFBr genotypes were infrequent. Twelve patient isolates did not match any other isolate within the NCFBr collection or the databases. The most common clone, clone C (10%), is also known to be abundant in the environment. In ten patients where longitudinal isolates were examined, paired isolates gave matching genotyping data suggesting persistent infection. There was incomplete concordance between the Array-Tube and VNTR methods (88% agreement).

Conclusion There were no dominant Pseudomonas aeruginosa clones in NCFBr suggesting that the most prevalent mode of infection is sporadic and cross infection is rare. This may reflect the local infection control measures however. Multicentre studies are suggested to further assess the risks.

Mechanisms in carcinogenesis

S127 CHEMOTHERAPY SENSITISES MALIGNANT PLEURAL MESOTHELIOMA CELLS TO UNDERGO MSC-TRAIL INDUCED APOPTOSIS

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