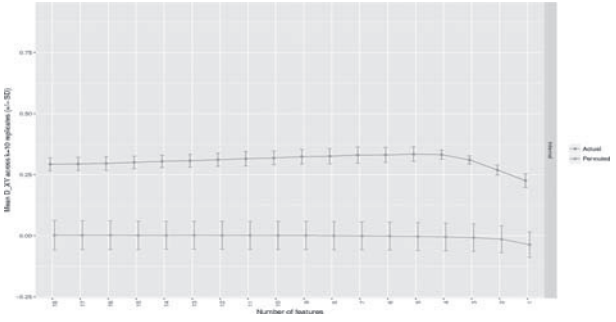
by Least Absolute Shrinkage and Selection Operator (Lasso) regression for Overall Survival (OS), OS<6 months and OS<12 months. OS prediction was quantified using Somers D_{XY} statistic, which varies from 0 to 1, with increasing concordance between observed and predicted outcomes. 6- and 12 month survival were described by area under the curve (AUC) scores.

Results Median OS was 270 (IQR 140–450) days. The primary OS model assigned high weights to 4 predictors: age, performance status, white cell count and serum albumin, and after cross-validation performed significantly better than would be expected by chance (mean D_{XY} 0.332 (+/-0.019) figure 1). However, validation set D_{XY} was only 0.221 (0.0935–0.346), equating to a 22% improvement in survival prediction than would be expected by chance. 6- and 12 month OS signatures included the same 4 predictors, in addition to epithelioid histology plus platelets and epithelioid histology plus C-reactive protein (mean AUC 0.758 (+/-0.022) and 0.737 (+/-0.012), respectively). The <6 month OS model demonstrated 74% sensitivity and 68% specificity. The <12 month OS model demonstrated 63% sensitivity and 79% specificity. Model content and performance were generally comparable with previous studies.

Discussion The prognostic value of the basic clinical information contained in these, and previously published models, is fundamentally of limited value in accurately predicting MPM prognosis. The methods described are suitable for expansion using emerging predictors, including tumour genomics and volumetric staging.



Abstract P243 Figure 1 Model performance as a function of the number of features for actual (red line) and permuted (blue line) data. Performance for Overall Survival (OS) is measured by D_{XY} for OS. Standard deviations from replicates of cross-validation are shown for each point as bars.

Clinical implications of cystic fibrosis

P244 EXTENDED CFTR SCREENING FOR PATIENTS WITH A CLINICAL DIAGNOSIS OF CF BUT ONLY ONE GENE ON INITIAL SCREENING

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Introduction Standard CF genotyping only identifies 94% of CF genes, resulting in the emergence of a "cystic fibrosis screen positive, inconclusive diagnosis" (CFSPID) designation. This, and the advent of genotype-specific CFTR directed therapies has highlighted the need for more comprehensive genotyping, particularly to identify rarer genes when only a single gene is found on initial screening. However, extended CFTR screening is an expensive investigation and we wished to assess its use and yield.

Methods Between 2014 and 2016 we identified 40 people with CF attending our large regional adult unit without two known pathogenic CFTR genes and offered them extended CFTR screening. We looked at the yield in terms of additional genes identified and their clinical significance in 37 of these (3 refused/did not attend).

Results A new molecular diagnosis (i.e., two pathogenic genes) was made in 18 (48.5%) people with CF. Genes associated with CFTR related disorders were found in a further 2 (5.5%), genes of uncertain pathogenicity were found in 3 (8.1%), and one or no genes were found in 14 (37.8%). Of the 18 people with CF with additional identified genes, 8 had those associated with responsiveness to the CFTR potentiator ivacaftor (4 × 3272–26 A≥G, 1 × 711+3 A≥G, 1 × R347H, 1 × 2789+5G≥A, 1 × S945L) and 2 of these (R347H and S945L) have recently been approved for ivacaftor use by the U.S. Food and Drug Administration.

Conclusions In people with a clinical diagnosis of CF but only one pathogenic gene on initial screen, extended CFTR screening identified a second gene in nearly half of cases. Furthermore, a significant proportion of the identified genes have been reported to respond to ivacaftor. It is therefore important that all people with CF without two known mutations undergo extended mutation screening in order to establish who may benefit should the current license for ivacaftor be expanded in the UK.

P245 INFLUENZA B OUTBREAK AT A LARGE ADULT CF CENTRE: CLINICAL CONSEQUENCES AND POTENTIAL CONTRIBUTING FACTORS

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Introduction In May 2016 an outbreak of influenza was declared on the adult cystic fibrosis (CF) ward at the Manchester Adult Cystic Fibrosis Centre (MACFC), where all patients are treated in individual rooms and strongly encouraged not to mix. The aim of this study was to investigate the outbreak, its clinical consequences, and potential contributing factors.

Method Notes of all patients admitted to the CF ward in May 2016 were retrospectively reviewed and data recorded included: patient location, respiratory viral PCR results, spirometry data (baseline measurements in the 6 month prior to onset, at onset of influenza, and at 3 months post infection), influenza vaccination status (all staff and patients), and measurements of ventilation in patient rooms.

Results Ten patients tested positive for influenza B; all were shown to have been infected with the same strain of the virus: B/Brisbane/60/2008. An outbreak timeline identified the likely index case (only patient admitted within the incubation period of influenza, first to develop symptoms and test positive for influenza B). Subsequently, 8 patients whose rooms