Development and external validation of 1- and 2-year mortality prediction models in cystic fibrosis

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ABSTRACT

Introduction: We aimed to develop a clinical tool for predicting 1- and 2-year risk of death for patients with cystic fibrosis (CF). The model considers patients' overall health status as well as risk of intermittent shock events in calculating the risk of death.

Methods: Canadian CF Registry data from 1982 to 2015 were used to develop a predictive risk model using threshold regression. A 2-year risk of death estimated conditional probability of surviving the second year given survival for the first year. UK CF Registry data from 2007 to 2013 were used to externally validate the model.

Results: The combined effect of CF chronic health status and CF intermittent shock risk provided a simple clinical scoring tool for assessing 1-year and 2-year risk of death for an individual CF patient. At a threshold risk of death of ⩾20%, the 1-year model had a sensitivity of 74% and specificity of 96%. The area under the receiver operating curve (AUC) for the 2-year mortality model was significantly greater than the AUC for a model that predicted survival based on forced expiratory volume in 1 s <30% predicted (AUC 0.95 *versus* 0.68 respectively, p<0.001). The Canadian-derived model validated well with the UK data and correctly identified 79% of deaths and 95% of survivors in a single year in the UK.

Conclusions: The prediction models provide an accurate risk of death over a 1- and 2-year time horizon. The models performed equally well when validated in an independent UK CF population.