Real-World Evaluation of the CF-Able Score as a Clinical Prognostic Tool in Cystic Fibrosis

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RATIONALE: The CF-ABLE score is an easy-to-use prognostic scoring system, validated in a large cohort from the national CF Registry of Ireland. This weighted score ranges from 0-7 and predicts risk of poor outcome, defined as death or requirement for lung transplantation, over a significant time period using commonly available clinical parameters such as age, body mass index (BMI), forced expiratory volume in 1s (FEV₁) and frequency of exacerbations. In our original 2013 derivation study, people with CF (PWCF) with a score ≥5 were deemed to have a 26% risk of poor outcome within 4 years. This study aimed to evaluate real-world clinical performance of the CF-ABLE score in a large single-center CF cohort over a 4-year period.

METHODS: Ethical approval was granted by Beaumont Hospital Ethics Committee. PWCF (n=130) were recruited from the 2013 Beaumont Hospital CF outpatient clinic list. Participants were evaluated over a two-year lead-in period from 2013-2015. The cohort was then subdivided into PWCF who transitioned to a CF-ABLE score of ≥5 during this entry period, and PWCF who did not. Outcomes for each PWCF who recorded a score of ≥5 were assessed 4 years after their score was recorded. RESULTS: Of the 130 PWCF studied, 47 (36.2% of the total cohort) recorded a CF-ABLE score of ≥5 within the study entry period. Of these 47 patients, 18 died or were transplanted (38.3%) within 4 years of reaching a score of ≥5. A further 6 were on the active transplant list at 4 years, giving a total of 48.9% dead, transplanted or listed for transplant within 4 years of crossing the threshold of CF-ABLE ≥5. Of the 23 PWCF who were neither deceased nor listed in the 4 years after crossing the threshold score, 5 had been referred but not accepted onto the transplant list and a further 5 had declined referral despite meeting referral criteria. Of those with a score ≥5, only 13 (27.7%) did not merit referral at 4 years. Of the patients who did not have a CF-ABLE score of ≥5, one died within the following 4 years. The cause of death was unrelated to his disease. None were listed or transplanted. CONCLUSION: The CF-ABLE score is better at identifying PWCF at increased risk of poor outcome than was initially thought. In contrast, patients have an outstanding chance of not progressing to death/transplant/listing in the next 4 years if their score is <5.

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