FEV₁ decline in cystic fibrosis

I read the paper by Taylor-Robinson et al.1 with interest with regard to modelling the decline in forced expiratory volume in 1 second (FEV₁) in patients with cystic fibrosis (CF). The authors have made the assumption that expressing spirometry results as a per cent of predicted (PP) is a valid way to use lung function over a wide age range. Young adult patients with cystic fibrosis die with absolute FEV₁ values just as low as those found in older patients with chronic obstructive pulmonary disease (COPD), so PP is not likely to be a valid methodology. When young CF patients were transplanted on the basis of an adult-derived PP cut-off value, it was found that those operated on had a worse survival than those who were not,² and this was due to the fact that in young subjects, a given PP value is at a higher absolute level than that in older subjects. We have shown,³ ⁴ as have others,⁵ that survival is better reflected by relating FEV₁ to a power of height rather than to a predicted value. The authors should consider reworking their data with other ways of standardising their data to see if CF patients’ lung function can be better assessed. When comparing these alternative methods with the method of relating results to a predicted value, they should also consider using a single continuous prediction equation, such as the Lambda-Mu-Sigma equation⁶ that overcomes any hiatus in moving from paediatric to adult equations.

Martin Miller
Correspondence to Professor Martin Miller, Institute of Occupational and Environmental Medicine, University of Birmingham, Birmingham B15 2TT, West Midlands, UK; m.r.miller@bham.ac.uk

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