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Journal club

Improved survival in cystic fibrosis patients with severely impaired lung function

A forced expiratory volume in one second (FEV $_1$) less than 30% predicted has been accepted as the threshold at which 50% of patients with cystic fibrosis (CF) survive 2 years or less. However, this estimate, made in the early 1990s, does not take into account recent developments in treatment. This cohort study aimed to re-evaluate the survival of CF patients with severely impaired lung function.

Two hundred and seventy-six CF patients whose FEV_1 was first observed to be less than 30% predicted between 1990 and 2003 were included in the cohort. The patients were followed up in 2-year subcohorts until 2007 and their survival was assessed. The authors showed an important improvement in the average survival of CF patients with severely impaired lung function. Median survival for patients who entered the cohort most recently (2002–2003) was 5.3 years, more than four times that for those who entered the study in the early 1990s, when median survival was 1.2 years. The authors observed a clear stepwise improvement in survival from 1994 to 1997. This was concurrent with the introduction of nebulised recombinant human DNase. A steady improvement in nutritional status also occurred in the time period studied, and an increased risk of death was associated with a body mass index below 19 kg/m².

The survival of patients with CF and an FEV_1 less than 30% predicted has improved markedly over the last two decades, with a median predicted survival of 5.3 years.

► George PM, Banya W, Pareek N, et al. Improved survival at low lung function in cystic fibrosis: cohort study from 1990 to 2007. BMJ 2011;342:1008.

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