

Cystic Fibrosis in Ireland: A Comparative Case Series Demonstrating Longitudinal Impact of Systemic and Therapeutic Advances

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Abstract

Background: Over the past two decades, cystic fibrosis (CF) management in Ireland has undergone accelerated transformation, marked by the introduction of newborn screening, systematic eradication of *Pseudomonas aeruginosa*, improved nutritional protocols, and the advent of CFTR modulator therapies. We present a comparative case series of three individuals born in 1978, 1988, and post-2015, each reflecting distinct phases of this national paradigm shift, to examine the cumulative impact on disease expression, trajectory, and prognosis.

Cases: Patient 1 (b.1978) exemplifies the pre-modulator cohort with severe early respiratory decline necessitating bilateral lung transplantation at 22. His post-transplant course has been complicated by chronic rejection, renal failure (eGFR 26), CF-related diabetes, and coronary artery disease, highlighting irreversible multisystem morbidity typical of care prior to specialist center consolidation. Patient 2 (b.1988), diagnosed at age 15, experienced a decline in FEV₁ to 61% by age 32, followed by a 50% improvement within 6 weeks of initiating elexacaftor/tezacaftor/ivacaftor (Kaftrio). Sustained lung function (ppFEV₁ 115%) and decreased exacerbation burden reflect the modulator's reversal of progressive decline. Patient 3 (b. post-2015) is under proactive management from diagnosis, in alignment with national newborn screening and NCPCF standards. National cohort data show improved FEV₁ and BMI in this age group, with chronic *Pseudomonas* prevalence falling to 0% in some centers, suggesting sustained pulmonary preservation and deferred systemic complications.

Conclusion: This series parallels national trends demonstrating that CFTR modulator access, early eradication protocols, and structured multidisciplinary care are associated with significant improvements in lung function, nutritional status, and reduction in chronic infection. The prognostic divergence across these patients captures the accelerated pace and clinical efficacy of Ireland's evolving CF care infrastructure.