Abstract

Objective: A number of disease-modifying therapies have been introduced for people with cystic fibrosis (CF) over the past two decades. The cumulative effects of this changing landscape on Cystic Fibrosis Related Diabetes (CFRD) are unclear. We aimed to examine trends in CFRD epidemiology over time using data from the US Cystic Fibrosis Foundation Patient Registry (CFFPR).

Research Design and Methods: CFFPR data from 2003-2018 were queried to determine annual screening, incidence, and prevalence rates of CFRD. Incident CFRD cases were compared to non-CFRD cases in select years. Survival analyses were performed to estimate the cumulative hazard of CFRD given predictors of interest over the 15 years of study. Data were also grouped into three time periods (2003-2008, 2009- 2013, and 2014-2018) to investigate if the hazard of developing CFRD varied over time.

Results: CFRD screening rates increased from 2003-2018, particularly in 10-18 year olds. Although screening rates increased in adults, overall rates remain low. In 10-18 year olds, the incidence of CFRD was stable over time, while incident cases in adults steadily decreased, approaching incident rates in adolescents. Despite this, the prevalence of CFRD has gradually increased in adults, likely reflecting increased longevity. Age, female sex, Black race, severe mutation class, liver disease, poorer lung function, pancreatic insufficiency, enteric feeds, low and high BMI were all risk factors associated with CFRD.

Conclusions: Findings support the need for development of tailored CFRD screening algorithms and increased subspecialists to care for a growing population of adults with CF and CF-associated comorbidities.