Clinical characteristics and outcomes in the adult cystic fibrosis population in Europe from 2014 to 2024: analysis of the European Cystic Fibrosis Society Patient Registry

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Summary

Background Since 2018, important advancements in the medical care of people with cystic fibrosis, particularly the introduction and widespread use of highly effective cystic fibrosis transmembrane conductance regulator modulators, have contributed to the adult cystic fibrosis population growing substantially and has led to an increased need for tailored health-care approaches. Our study aimed to analyse the extent to which the clinical characteristics and treatment outcomes of adults with cystic fibrosis have evolved from 2014 to 2024.

Methods The European Cystic Fibrosis Society Patient Registry (ECFSPR), collects annual data for more than 55 000 people with cystic fibrosis. Longitudinal data from 20 countries in Europe with high patient coverage (>85%) from 2014 to 2024 were analysed, representing 80% of the whole ECFSPR cohort. Differences in annual cross-sectional estimates were assessed using regression models.

Findings Between 2014 and 2024, the number of adults with cystic fibrosis increased by 45·0%, from 50·9% to 60·5% of the total cystic fibrosis population. The number of adults older than 30 years nearly doubled. Among adults with cystic fibrosis who had not received a transplant, mean percent predicted FEV_1 improved from 66·1% to 78·8% (p<0·0001), with most of the gain occurring after 2020. Chronic *Pseudomonas aeruginosa* infection declined significantly (p<0·0001), whereas mean BMI increased significantly (p<0·0001), halving the proportion of individuals who are underweight. Age-related complications, such as malignancy, increased, whereas cystic fibrosis-specific complications and insulin-treated diabetes declined. The largest improvements were observed in individuals with at least one variant responsive to elexacaftor–tezacaftor–ivacaftor (ETI). The uptake of ETI increased from 2% in 2019 to 71% in 2024, associated with improvements in health indicators.

Interpretation From 2014 to 2024, the adult cystic fibrosis population in Europe expanded substantially due to marked improvements in treatment, particularly following the availability of ETI triple therapy from 2018–19 onwards. The growing number of people with CF surviving to adulthood is consistent with substantial effects from improved care and cystic fibrosis transmembrane conductance regulator modulators, such as ETI, and highlight evolving care needs of people with cystic fibrosis.

Funding None.

Lancet Respir Med 2026

Published Online
June 3, 2026
https://doi.org/10.1016/S2213-2600(26)00149-9

See Online/Comment
https://doi.org/10.1016/S2213-2600(26)00189-X

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Introduction

Cystic fibrosis is an autosomal recessive genetic disease caused by variants in the cystic fibrosis transmembrane conductance regulator (*CFTR*) gene. Characterised by progressive multisystem dysfunction—primarily affecting the lungs, pancreas, gastrointestinal tract, and reproductive system—cystic fibrosis has historically been associated with substantial morbidity and early mortality.[1,2] However, over the past three decades, cystic fibrosis has undergone a clear transformation in its clinical trajectory, primarily due to earlier diagnosis through newborn screening, improvements in antibiotic regimens and airway clearance therapies, and increasingly specialised, multidisciplinary care models.[1,3]

The most recent and arguably most profound advancement has been the development of CFTR modulator therapies from 2018–19 onwards across Europe, which target the underlying molecular defect in cystic fibrosis rather than its downstream consequences. These therapies, particularly the triple combination of elexacaftor–tezacaftor–ivacaftor (ETI), have resulted in substantial improvements in lung function, nutritional status, and quality of life for individuals with responsive genotypes.[4,5] As a result, cystic fibrosis is transitioning from a fatal childhood disease to a chronic condition increasingly managed into adulthood and beyond.[1]

This therapeutic progress has contributed to a substantial demographic shift; for the first time in history, the majority of people with cystic fibrosis in many

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See Online for appendix

Research in context

Evidence before this study

We searched PubMed, MEDLINE, and Embase from database inception to Dec 20, 2025, for studies reporting clinical outcomes in adults with cystic fibrosis, using combinations of the terms "cystic fibrosis", "adult", "registry", "survival", "lung function", "CFTR modulators", and "elexacaftor-tezacaftor-ivacaftor". No language restrictions were applied. We also screened reference lists of relevant articles. We prioritised large registry-based, longitudinal studies and assessed quality on the basis of study design, population size, duration of follow-up, and completeness of data capture. Previous studies have documented improvements in survival and lung function in cystic fibrosis populations, particularly following the introduction of cystic fibrosis transmembrane conductance regulator (CFTR) modulators, but few have comprehensively evaluated long-term trends specifically in adult populations across several countries.

Added value of this study

This study provides a large-scale, multinational analysis of more than 25 000 adults with cystic fibrosis from 20 European countries with high registry coverage over 10 years. Our study captures the transition from the premodulator era to the CFTR modulator era 2018–19 onwards, including widespread adoption of elexacaftor-tezacaftor-ivacaftor. The study offers detailed insights into changes in lung function, nutritional status, infection burden, complications, and survival in the adult population.

Implications of all the available evidence

The findings support a major shift in cystic fibrosis from a disease with short life expectancy towards a chronic adult disease, with substantial improvements in health outcomes associated with modern therapies. However, our results also highlight emerging challenges related to ageing, comorbidities, and the demands of the health-care system. These results underscore the need to evolve care models and ensure equitable access to CFTR modulators.

high-income countries are adults.[6] The European Cystic Fibrosis Society Patient Registry (ECFSPR) reported that, by 2022, more than 58% of people with cystic fibrosis in Europe were aged at least 18 years—a proportion that continues to increase. Adults with cystic fibrosis face a unique clinical profile compared with children and adolescents, marked by a greater burden of accumulated lung damage; more frequent comorbidities such as cystic fibrosis-related diabetes, osteoporosis, and liver disease; and increasingly, an increased risk for age-associated complications such as malignancy and cardiovascular diseases.[1,7] These changing patterns underscore the necessity for age-adapted care models, long-term health planning, and a broader research focus on ageing in cystic fibrosis.

Despite growing interest in the adult cystic fibrosis population, relatively few studies have comprehensively assessed how this group has changed over time, especially at the multinational level. Understanding long-term trends in lung function, nutritional status, infection status, and survival is essential for planning future clinical services, evaluating therapeutic effects, and identifying emerging needs among older adults with cystic fibrosis. Moreover, tracking the uptake of CFTR modulators—particularly ETI—can help inform policy decisions and clinical guidelines.

For more on the **ECFSPR** see https://pr.ecfs.eu/

To address this gap, we performed a longitudinal analysis of ECFSPR data from 20 European countries, with comprehensive national registry coverage, from 2014 to 2024. This period captures both the eras before and after the introduction of CFTR modulator therapies (from 2014 to 2019 and from 2020 to 2024), offering a unique opportunity to assess their real-world effects. Our objectives were to characterise longitudinal trends in the adult cystic fibrosis population size,demographics, clinical characteristics, and outcomes, to evaluate the effect of modulator therapies on health trajectories, and to identify future challenges in cystic fibrosis care as the adult population continues to grow and age.

Methods

ECFSPR

The ECFSPR is the largest cystic fibrosis registry worldwide, collecting data on people with cystic fibrosis from cystic fibrosis centres and national registries in the European region, as defined by WHO, and covers the period from 2008 to 2024 with increased coverage each year. In 2024, more than 55 000 people with cystic fibrosis from 47 countries were included in the ECFSPR. Data are provided to ECFSPR on an annual basis, in accordance with existing ethical approvals and data governance structures, and in accordance with specific inclusion criteria. The data collected include CFTR genotype, demographic and clinical variables, pulmonary and other cystic fibrosis-related complications, infections with cystic fibrosis-related pathogens, and chronic treatments. All variables are collected following standardised definitions. Further information can be found on the ECFSPR official website.

Study population

The ECFSPR increased considerably between 2008 and 2024, both in terms of coverage and the number

of countries included, which contributed to the number of people with cystic fibrosis included more than doubling. This aspect makes comparisons over time more challenging, given that some differences in demography and clinical outcomes could be attributed to the increasing coverage (ie, the expansion across countries and the inclusion of new countries) from 2008 to 2024. Thus, in this study, we decided to select a more homogeneous period and set of countries, which achieved high coverage (≥85%) during the whole study period, making the comparison over time more robust.

We considered data from 2014 to 2024, specifically from 20 countries with well established data collection and inclusion in ECFSPR, which achieved high coverage (≥85%) during the study period. Coverage was defined as the proportion of individuals with cystic fibrosis captured in national registries relative to independent epidemiological estimates; country-specific coverage percentages are provided in the ECFSPR annual reports. These countries are Austria, Czechia, Denmark, France, Germany, Greece, Hungary, Ireland, Israel, Italy, Latvia, the Netherlands, North Macedonia, Portugal, Serbia, Slovakia, Slovenia, Sweden, Switzerland, and the UK. This selection corresponds to the vast majority (around 80%) of people with cystic fibrosis included in ECFSPR in the study period.

Variable definitions

The genotype of people with cystic fibrosis is classified into three groups according to the presence of variants responsive to ETI:[8] at least one variant responsive to ETI (Phe508del, one of the 506 variants considered responsive to ETI and approved by the European Medicines Agency, or one of the 21 ETI variants considered responsive or probably responsive by the French compassionate programme); two non-responsive variants (among 955 class I variants and two class II variants); and other variants with unknown response to ETI and unknown variants.

The ECFSPR defines people with cystic fibrosis as having a chronic infection with *Pseudomonas aeruginosa* if they fulfil the modified Leeds criteria for chronicity (more than 50% of respiratory samples collected during the year are positive for *P aeruginosa*, with at least four samples collected during that period), with or without bacteria-specific antibodies, as determined by local laboratories. The ECFSPR also defines people with cystic fibrosis as having a chronic infection when the aforementioned criteria were met over the past year and the physician has no reason to believe the status has changed. Underweight is defined as a BMI lower than 18·5 kg/m^2. Cystic fibrosis-related diabetes is considered to be present when the patient is treated with the use of insulin. Liver disease is categorised into two main types, cirrhosis and non-cirrhotic liver disease. Further detailed information on variable definitions can be found on the ECFSPR official website.

Ethical approval

All participating centres and national registries obtained ethical approval and patients' informed consent for data collection and participation in the ECFSPR, including consent for the future use of research data. This study received approval from the ECFSPR Scientific Committee and the ECFSPR Steering Committee.

Statistical analysis

As a preliminary analysis, we computed the number of people with cystic fibrosis across the 20 countries included in the study over the study period (2014–24), and we described the distribution of people with cystic fibrosis by age group. Four age groups were considered: 0–11 years; 12–17 years; 18–29 years; and people aged 30 years or older.

We did a survival analysis considering all-cause mortality as the endpoint. We fitted cox proportional hazard models using age as the timescale, with a 5-year follow-up period, as suggested by Sykes and colleagues.[9] From these models, we extracted the median survival, providing a comprehensive overview of the entire study period.

Our study adopts a repeated cross-sectional design, in which annual estimates are conditional on individuals alive and recorded in each calendar year. The aim was to describe population-level temporal trends rather than within-individual longitudinal changes. All remaining analyses were restricted to adults (aged ≥18 years) with cystic fibrosis, enabling a focused assessment of temporal changes in the adult cystic fibrosis population. Demographic and clinical characteristics, along with complications and treatments, were described annually from 2014 to 2024. We summarised categorical variables as percentages and continuous variables as means. We evaluated changes over time using generalised estimating equations to account for repeated measures within individuals. We applied logistic models to dichotomous outcomes and linear models to continuous variables. Results are reported as average annual percentage changes or mean changes, with corresponding 95% CIs and p values.

We also performed descriptive and regression analyses after stratifying individuals by genotype group and age group (18–29 years and ≥30 years). All estimates are conditional on individuals alive and recorded in each calendar year. No causal interpretation is implied, and observed associations should be interpreted as population-level temporal trends rather than individual-level treatment effects. We carried out all statistical analyses in R version 4.5.0.

Role of the funding source

There was no funding source for this study.

Results

Between 2014 and 2024, the number of people with cystic fibrosis across the 20 countries in this study increased

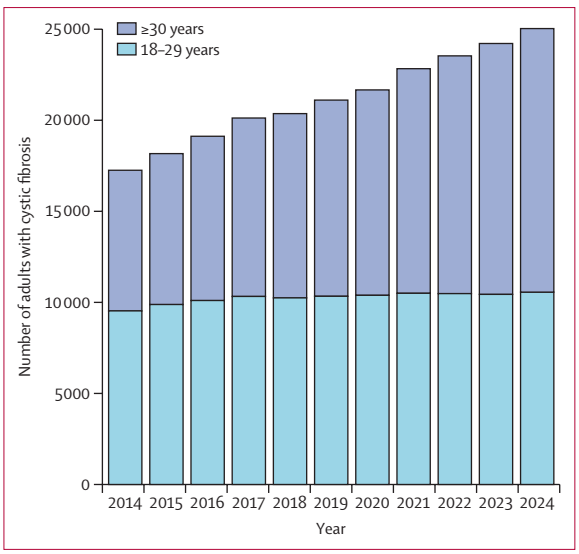

Figure 1: **Number of adults with cystic fibrosis aged 18–29 years and 30 years or older in 20 European countries from 2014 to 2024**

from 33 916 to 41 397. The number of children (aged <18 years) remained relatively stable, with 16 650 individuals in 2014 and to 16 362 in 2024, whereas the adult population increased by 45·0%, from 17 266 to 25 035. As a result, the proportion of adults rose from 17 266 (50·9%) to 25 035 (60·5%) of the total cystic fibrosis population. When examined by age group, the number of people with cystic fibrosis aged younger than 12 years remained stable, with 11 028 individuals in 2014 and 10 525 in 2024, suggesting no increase in the number of children born with cystic fibrosis. The cystic fibrosis population aged 12–17 years showed only a slight increase, with 5622 in 2014 and to 5837 in 2024. By contrast, the number of adults rose substantially; those aged 18–29 years increased from 9551 to 10 584 individuals in 2014–24, whereas the population aged 30 years or older nearly doubled, from 7715 to 14 451 individuals over the same period (figure 1; appendix p 3). The number of new adult diagnoses remained stable. Notably, the number of people with cystic fibrosis who are older than 50 years has tripled in the past 10 years, increasing from 896 to 2930 individuals.

The proportion of adults with cystic fibrosis with two non-responsive variants increased slightly from 2·1 to 2·4 in 2014–24 compared with those with at least one variant. The percentage of pancreatic enzyme use decreased slightly over the study period, from 83·7% to 80·4% (table 1).

The number of adults who received a lung transplant in 2014 was 224 (1·3%) and the number of adults who received a lung transplant in 2024 was only 38 (0·2%), showing a significant decrease in the number and percentage of adults receiving lung transplants in the study period (table 1). However, the number of adults living with a lung transplant has increased from

	2014	2015	2016	2017	2018	2019	2020	2021	2022	2023	2024	2014–19 annual difference (95% CI)	p value	2020–24 annual difference (95% CI)	p value	2014–24 annual difference (95% CI)	p value
Number of adults with cystic fibrosis	17 266	18 179	19 130	20 131	20 372	21 113	21 671	22 835	23 539	24 217	25 035	..	..	..	..	..	..
Demographic and diagnostics characteristics																	
Gender																	
Woman	7953 (46·1%)	8394 (46·2%)	8804 (46·0%)	9253 (46·0%)	9439 (46·3%)	9796 (46·4%)	10 070 (46·5%)	10 653 (46·7%)	10 996 (46·7%)	11 287 (46·6%)	11 658 (46·6%)	0·1 (0 to 0·2)	0·26	0 (−0·1 to 0·1)	0·79	0·1 (0 to 0·1)	0·03
Man	9313 (53·9%)	9785 (53·8%)	10 326 (54%)	10 878 (54%)	10 933 (53·7%)	11 317 (53·6%)	11 601 (53·5%)	12 182 (53·3%)	12 543 (53·3%)	12 930 (53·4%)	13 377 (53·4%)	..	..	..	..	..	..
Current age	30·9 (10·2)	31·2 (10·4)	31·6 (10·6)	32·0 (10·7)	32·2 (10·8)	32·5 (11·0)	32·9 (11·1)	33·3 (11·4)	33·8 (11·6)	34·2 (11·7)	34·5 (12·0)	0·3 (0·3 to 0·3)	<0·0001	0·4 (0·4 to 0·4)	<0·0001	0·4 (0·3 to 0·4)	<0·0001
Genotype, two non-responsive variants	365 (2·1%)	390 (2·1%)	408 (2·1%)	446 (2·2%)	469 (2·3%)	499 (2·4%)	517 (2·4%)	555 (2·4%)	559 (2·4%)	564 (2·3%)	592 (2·4%)	0·1 (0 to 0·1)	<0·01	0 (0 to 0)	0·36	0 (0 to 0)	0·01
Use of pancreatic enzymes	13 640 (83·7%)	14 558 (82·7%)	15 330 (82·0%)	16 125 (81·2%)	16 592 (82·1%)	17 161 (81·8%)	17 650 (82·1%)	18 511 (81·4%)	18 941 (80·9%)	19 322 (80·1%)	20 081 (80·4%)	−0·3 (−0·4 to −0·2)	<0·0001	−0·4 (−0·6 to −0·3)	<0·0001	−0·3 (−0·3 to −0·2)	<0·0001

(Table 1 continues on next page)

	2014	2015	2016	2017	2018	2019	2020	2021	2022	2023	2024	2014–19 annual difference (95% CI)	p value	2020–24 annual difference (95% CI)	p value	2014–24 annual difference (95% CI)	p value
(Continued from previous page)																	
Age at diagnosis <3 months	4908 (30·3%)	5232 (30·1%)	5637 (30·4%)	5907 (30·3%)	6178 (31·2%)	6422 (31·3%)	6626 (31·4%)	7031 (31·6%)	7298 (31·8%)	7660 (32·5%)	8018 (32·8%)	0·2 (0·1 to 0·3)	<0·0001	0·4 (0·3 to 0·5)	<0·0001	0·3 (0·2 to 0·3)	<0·0001
Age at diagnosis >18 years	2264 (14·0%)	2478 (14·3%)	2649 (14·3%)	2837 (14·5%)	2714 (13·7%)	2805 (13·7%)	2894 (13·7%)	3117 (14·0%)	3285 (14·3%)	3381 (14·3%)	3540 (14·5%)	−0·1 (−0·2 to 0)	0·02	0·2 (0·1 to 0·3)	<0·0001	0 (0 to 0·1)	0·33
Neonatal screening performed	1106 (10·2%)	1246 (10·8%)	1332 (10·3%)	1718 (11·0%)	2022 (12·4%)	2139 (12·5%)	2341 (13·3%)	2662 (13·0%)	2827 (13·5%)	3105 (13·4%)	3438 (14·3%)	0·5 (0·4 to 0·6)	<0·0001	0·3 (0·2 to 0·4)	<0·0001	0·4 (0·3 to 0·5)	<0·0001
Transplant history																	
Adults living with lung transplant	1719 (10·2%)	1859 (10·3%)	2059 (10·8%)	2241 (11·3%)	2239 (11·0%)	2432 (11·6%)	2388 (11·1%)	2453 (10·8%)	2350 (10%)	2217 (9·2%)	2133 (8·5%)	0·3 (0·2 to 0·4)	<0·0001	−0·7 (−0·7 to −0·6)	<0·0001	−0·2 (−0·2 to −0·1)	<0·0001
Adults' lung transplanted in the year of follow-up	224 (1·3%)	204 (1·1%)	247 (1·3%)	249 (1·2%)	216 (1·1%)	233 (1·1%)	78 (0·4%)	55 (0·2%)	43 (0·2%)	43 (0·2%)	38 (0·2%)	0 (−0·1 to 0)	0·06	0 (−0·1 to 0)	<0·0001	−0·1 (−0·2 to −0·1)	<0·0001
Adults living with liver transplant	147 (0·9%)	169 (0·9%)	186 (1·0%)	187 (1·0%)	198 (1·0%)	201 (1·0%)	218 (1·0%)	239 (1·0%)	245 (1·0%)	251 (1·0%)	265 (1·1%)	0 (0 to 0)	0·25	0 (0 to 0)	0·47	0 (0 to 0)	0·04
Adults' liver transplanted in the year of follow-up	5 (<0·1%)	9 (<0·1%)	11 (0·1%)	8 (<0·1%)	9 (<0·1%)	10 (<0·1%)	7 (<0·1%)	2 (<0·1%)	5 (<0·1%)	8 (<0·1%)	13 (0·1%)	0 (0 to 0)	0·67	0 (0 to 0)	0·10	0 (0 to 0)	0·38
Lung function and nutrition																	
ppFEV$_1$	66·1 (24·3)	66·8 (24·3)	67·4 (24·2)	67·9 (24·1)	68·4 (24·0)	68·7 (24·0)	69·5 (23·4)	74·6 (23·8)	77·1 (24·0)	77·5 (24·0)	78·8 (24·0)	0·5 (0·4 to 0·6)	<0·0001	2·1 (2·0 to 2·2)	<0·0001	1·4 (1·4 to 1·5)	<0·0001
ppFEV$_1$<40	2256 (17·1%)	2398 (16·6%)	2533 (16%)	2522 (15·1%)	2526 (14·6%)	2574 (14·3%)	2134 (11·9%)	1694 (8·8%)	1543 (7·6%)	1585 (7·6%)	1503 (6·9%)	..	..	..	..	..	..
ppFEV$_1$ 40–59	3250 (24·7%)	3487 (24·1%)	3757 (23·7%)	3889 (23·4%)	4000 (23·1%)	4221 (23·4%)	4387 (24·5%)	3923 (20·3%)	3760 (18·5%)	3786 (18·1%)	3747 (17·1%)	..	..	..	..	..	..
ppFEV$_1$ 60–79	3570 (27·1%)	3912 (27·1%)	4271 (26·9%)	4554 (27·4%)	4706 (27·2%)	4867 (26·9%)	4953 (27·7%)	5123 (26·5%)	5122 (25·1%)	5128 (24·5%)	5179 (23·6%)	..	..	..	..	..	..
ppFEV$_1$ ≥80	4101 (31·1%)	4659 (32·2%)	5301 (33·4%)	5682 (34·1%)	6049 (35%)	6398 (35·4%)	6406 (35·8%)	8590 (44·4%)	9948 (48·8%)	10457 (49·9%)	11497 (52·4%)	..	..	..	..	..	..
BMI (kg/m^2)	21·8 (3·5)	21·9 (3·6)	22·0 (3·6)	22·1 (3·6)	22·1 (3·6)	22·2 (3·7)	22·4 (3·8)	22·9 (3·8)	23·1 (4·0)	23·2 (4·1)	23·3 (4·1)	0·1 (0·1 to 0·1)	<0·0001	0·2 (0·2 to 0·2)	<0·0001	0·2 (0·2 to 0·2)	<0·0001
<18·5	2371 (14·5%)	2416 (14%)	2460 (13·3%)	2554 (13·2%)	2521 (12·8%)	2519 (12·3%)	2141 (10·4%)	1791 (8·3%)	1825 (8·1%)	1758 (7·5%)	1797 (7·4%)	..	..	..	..	..	..
18·5–25	11 485 (70·1%)	12 067 (69·9%)	12 944 (69·9%)	13 471 (69·6%)	13 700 (69·6%)	14 237 (69·5%)	14 429 (70·3%)	14 831 (68·7%)	15 113 (66·9%)	15 449 (66·2%)	15 741 (65·2%)	..	..	..	..	..	..
>25	2535 (15·5%)	2790 (16·2%)	3119 (16·8%)	3330 (17·2%)	3475 (17·6%)	3740 (18·2%)	3962 (19·3%)	4970 (23%)	5637 (25%)	6114 (26·2%)	6602 (27·3%)	..	..	..	..	..	..

(Table 1 continues on next page)

	2014	2015	2016	2017	2018	2019	2020	2021	2022	2023	2024	2014–19 annual difference (95% CI)	p value	2020–24 annual difference (95% CI)	p value	2014–24 annual difference (95% CI)	p value
(Continued from previous page)																	
Microbiology infections																	
Chronic *Pseudomonas aeruginosa*	6688 (48·3%)	7046 (47·2%)	8955 (48·2%)	9187 (47·2%)	8821 (44·7%)	8980 (44·0%)	8444 (40·1%)	7376 (33·6%)	6893 (30·4%)	6470 (27·5%)	6369 (26·1%)	−0·9 (−1·1 to −0·7)	<0·0001	−3·4 (−3·5 to −3·2)	<0·0001	−2·6 (−2·7 to −2·5)	<0·0001
Chronic *Staphylococcus Aureus*	3458 (34·5%)	3777 (34·6%)	5526 (37·6%)	7089 (37·1%)	7111 (35·9%)	7190 (35·3%)	7150 (34·0%)	6388 (29·1%)	6167 (27·2%)	5885 (25·0%)	6270 (25·7%)	0·1 (−0·1 to 0·3)	0·38	−2·0 (−2·2 to −1·8)	<0·0001	−1·4 (−1·5 to −1·3)	<0·0001
Stenotrophomonas Maltophilia	999 (7·3%)	1088 (7·4%)	1532 (8·3%)	1731 (8·9%)	1758 (9·0%)	1972 (9·6%)	1817 (8·6%)	1622 (7·3%)	1295 (5·7%)	1200 (5·1%)	1256 (5·2%)	0·5 (0·4 to 0·6)	<0·0001	−0·9 (−1·0 to −0·8)	<0·0001	−0·3 (−0·4 to −0·3)	<0·0001
Chronic *Burkholderia Cepacia Complex*	611 (4·4%)	666 (4·4%)	766 (4·1%)	766 (3·9%)	756 (3·8%)	726 (3·6%)	728 (3·5%)	441 (2·0%)	403 (1·8%)	368 (1·6%)	340 (1·4%)	−0·2 (−0·2 to −0·1)	<0·0001	−0·4 (−0·5 to −0·4)	<0·0001	−0·4 (−0·4 to −0·3)	<0·0001
Methicillin-resistant *Staphylococcus aureus*	NA	NA	NA	NA	934 (5·3%)	1088 (5·3%)	1100 (5·2%)	1058 (4·8%)	943 (4·2%)	901 (3·8%)	866 (3·6%)	−0·1 (−0·4 to 0·2)	0·70	−0·4 (−0·5 to −0·3)	<0·0001	−0·3 (−0·4 to −0·3)	<0·0001
Achromobacter species	NA	NA	NA	NA	1117 (6·5%)	1403 (6·9%)	1320 (6·3%)	1318 (6·0%)	1057 (4·7%)	997 (4·3%)	988 (4·1%)	0·4 (0·1 to 0·7)	0·01	−0·6 (−0·7 to −0·5)	<0·0001	−0·5 (−0·6 to −0·5)	<0·0001

N (%) by year is reported for categorical variables and mean (SD) by year for numerical variables (ppFEV1 and BMI). NA=not applicable. $ppFEV_1$=percent predicted FEV_1.

Table 1: **Description of demographic and clinical characteristics in adults with cystic fibrosis, from 2014 to 2024**

1719 to 2133 people, consistent with improved survival of adults after lung transplant and better coverage of people with cystic fibrosis after transplantation in the registry. A different trend is observed for liver transplant, with the number and percentage of transplants increasing, although the increase is not statistically significant (p=0·38). Only five (<0·1%) adults received liver transplants in 2014, and 13 (<0·1%) adults received liver transplants in 2024. Moreover, the number of people living with liver transplants almost doubled from 147 in 2014 to 265 in 2024.

Percent predicted FEV_1 in adults with cystic fibrosis without a lung transplant increased from 66·1 to 78·8 (p<0·0001), with 75% of the increase in the past 4 years. With regards to race and ethnicity, the registry only collected this information from 2022 onwards. In 2024, 24 359 (97·3%) of adults with cystic fibrosis were Caucasian. Mean BMI increased from 21·8 kg/m² to 23·3 kg/m² (p<0·0001), and correspondingly, the percentage of people who were underweight (BMI <18·5 kg/m²) decreased from 14·5% to 7·4% (p<0·0001), whereas the percentage of people who were overweight (BMI >25 kg/m²) increased from 15·5% to 27·3% and the percentage of people with obesity (BMI >30 kg/m²) more than doubled, from 2·7% to 6·1% (table 1; figure 2; appendix p 2). The prevalence of chronic *P aeruginosa* infection decreased from 6688 (48·3%) to 6369 (26·1%) (p<0·0001). All other cystic fibrosis-related lung infections (chronic *Staphylococcus aureus, Stenotrophomonas maltophilia*, chronic *Burkholderia cepacia complex*, methicillin-resistant *Staphylococcus aureus,* and *Achromobacter species*) showed significant decreases of prevalence over time. Complications of cystic fibrosis (allergic bronchopulmonary aspergillosis, major haemoptysis, and pneumothorax), liver cirrhosis, and insulin-treated diabetes decreased, whereas the prevalence of liver disease without cirrhosis and malignancy increased (table 2; appendix pp 4–6). The median predicted survival age of the overall population has considerably increased, from 50·6 years (95% CI 49·1–52·2) in 2010–14 to 69·6 years (67·1–72·4) in 2020–24, with more than a 10-year increase from 2016–20 (55·8 years [95% CI 54·7–57·2]) to 2020–24 (69·6 years [67·1–72·4]; figure 3).

From the analysis stratified by genotype group (ETI responsiveness), both groups showed significant changes from 2014 to 2024; however, the main improvements occurred among adults with cystic fibrosis with at least one ETI-responsive variant. In adults with cystic fibrosis without ETI responsive variants, percent predicted FEV_1 ($ppFEV_1$) increased by 6% from 62 to 66, whereas in adults with cystic fibrosis with at least one ETI responsive variant, $ppFEV_1$ had a steeper increase from 66·1 to 78·8 (by 20%), with the majority of this increase occurring after 2020. The same optimistic trend is observed in both groups for chronic *P aeruginosa* infection, with a greater decline in the ETI-responsive group (from 49·1% to 24·9%). The ETI non-responsive group still

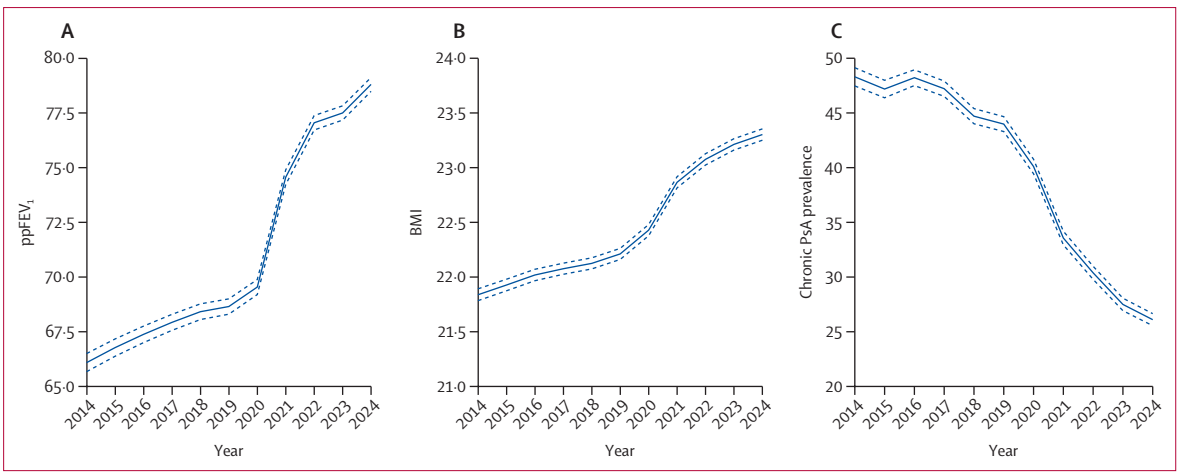

Figure 2: **Mean ppFEV1 (A), mean BMI (B), prevalence of chronic *Pseudomonas aeruginosa* (C) from 2014 to 2024**
Dashed lines represent 95% CIs. ppFEV1=percent predicted FEV_1.

showed a decrease, albeit to a lesser extent, and maintained a very high colonisation rate in 2024 (from 56·8% to 51·1%). BMI increased significantly only in people with at least one ETI-responsive variant (from 21·8 kg/m² to 23·4 kg/m²), whereas it remained stable in adults with cystic fibrosis without ETI-responsive variants (from 21·1 kg/m² to 21·2 kg/m²; figure 4; appendix pp 4–6).

From the analysis stratified by age group (18–29 years *vs* ≥30 years), the results indicated that improvements in $ppFEV_1$, BMI, and chronic *Pseudomonas aeruginosa* prevalence were similar and significant in both groups (appendix p 2).

Discussion

This multinational analysis of more than 25 000 adults with cystic fibrosis across 20 European countries documents a decade of remarkable improvement in both survival and health outcomes of people with cystic fibrosis, coinciding with a profound demographic transformation of the cystic fibrosis population. Between 2014 and 2024, the number of adults with cystic fibrosis increased by 45%, and the number of those aged 30 years or older nearly doubled. The number of new adult diagnoses remained stable, suggesting limited contribution to adult population growth. The increase in older age groups was consistent with improved survival. These shifts reflect a steady increase in survival[2,10,11–14] and signal a redefinition of cystic fibrosis from a predominantly paediatric illness to a chronic adult disease requiring long-term, age-appropriate care.[1,7,15,16]

During the study period, adults with cystic fibrosis showed striking health gains across several domains. Among individuals without a transplant, mean $ppFEV_1$ increased significantly, with 75% of this gain occurring after 2020. Nutritional status also improved markedly, and the proportion of adults who were underweight halved, whereas the proportion of those who were overweight increased. These improvements occurred in parallel with reductions in the prevalence of chronic *P aeruginosa* infections and other respiratory pathogens, including the *Burkholderia cepacia* complex and methicillin-resistant *Staphylococcus aureus*.[17]

Notably, these population-level health improvements were not confined to younger adults but were equally evident among individuals aged 30 years or older, an age group historically at higher risk of disease progression, complications, and mortality.[18,19] This finding suggests that therapeutic advances might have contributed to improvements, even in those with long-standing disease, reversing trajectories once considered irreversible. Survival estimates reflect the overall population and might differ by genotype.

A major factor in this transformation appears to be the swift and widespread adoption of CFTR modulator therapies. Data from 2018 to 2024 indicate a remarkable increase in the use of ETI, rising from less than 1% in 2018 to more than 70% in 2024 among adults in the registry. Stratified analyses demonstrate that individuals using ETI showed notably greater improvements in lung function and BMI, and more pronounced reductions in chronic infection rates, compared with non-users. Cystic fibrosis-specific complications and insulin-treated diabetes declined. Possible explanations include improved metabolic control with CFTR modulators and earlier disease modification. These observations are consistent with findings from phase 3 clinical trials,[4,5] real-world studies, and previous registry studies from Europe and North America.[10,19–21] Although the design of our observational study limits our ability to draw definitive causal conclusions, the temporal correlation between the escalation in ETI use and improvements in health indicators presents a compelling narrative that aligns with previous registry-based studies.[19–21] These findings reflect ecological associations at the population level and

	2014	2015	2016	2017	2018	2019	2020	2021	2022	2023	2024	2014–19 annual difference (95% CI)	p value	2020–24 annual difference (95% CI)	p value	2014–24 annual difference (95% CI)	p value
Number of adults with cystic fibrosis	17 266	18 179	19 130	20 131	20 372	21 113	21 671	22 835	23 539	24 217	25 035	..	..	..	..	..	..
Complications																	
Diabetes treated with insulin	4975 (33·2%)	5309 (30·9%)	5228 (28·1%)	5601 (28·4%)	5774 (28·7%)	6024 (29·0%)	6158 (28·9%)	6436 (29·0%)	6428 (27·7%)	6350 (26·5%)	6390 (25·9%)	−0·7 (−0·9 to −0·6)	<0·0001	−0·9 (−1·0 to −0·7)	<0·0001	−0·5 (−0·5 to −0·4)	<0·0001
Liver cirrhosis	889 (5·1%)	972 (5·3%)	1082 (5·7%)	1010 (5·0%)	933 (4·6%)	1017 (4·8%)	1076 (5·0%)	1094 (4·8%)	1154 (4·9%)	1179 (4·9%)	1239 (4·9%)	−0·1 (−0·2 to −0·1)	<0·01	0 (−0·1 to 0·1)	0·86	0 (−0·1 to 0)	0·02
Liver disease without cirrhosis	2045 (14·5%)	2655 (15·9%)	3155 (17·2%)	3790 (19·5%)	3704 (18·6%)	4308 (20·8%)	4716 (22·3%)	5250 (23·4%)	5791 (25·1%)	6211 (25·9%)	6609 (27·0%)	1·2 (1·1 to 1·3)	<0·0001	1·2 (1·0 to 1·3)	<0·0001	1·2 (1·2 to 1·3)	<0·0001
Allergic bronchopulmonary aspergillosis	1511 (11·0%)	1577 (9·8%)	1385 (7·5%)	1441 (7·4%)	1419 (7·1%)	1509 (7·3%)	1347 (6·3%)	1288 (5·7%)	1075 (4·7%)	924 (4·0%)	976 (3·9%)	−0·7 (−0·8 to −0·6)	<0·0001	−0·6 (−0·7 to −0·6)	<0·0001	−0·6 (−0·7 to −0·6)	<0·0001
Pneumothorax	168 (1·2%)	169 (1·0%)	134 (0·7%)	123 (0·6%)	150 (0·7%)	106 (0·5%)	104 (0·5%)	57 (0·3%)	58 (0·3%)	40 (0·2%)	31 (0·1%)	−0·1 (−0·2 to −0·1)	<0·0001	−0·1 (−0·1 to −0·1)	<0·0001	−0·1 (−0·1 to −0·1)	<0·0001
Major haemoptysis	485 (3·5%)	532 (3·2%)	622 (3·5%)	383 (2·0%)	420 (2·2%)	530 (2·6%)	472 (2·2%)	328 (1·5%)	243 (1·1%)	204 (0·9%)	253 (1·0%)	−0·2 (−0·3 to −0·2)	<0·0001	−0·3 (−0·3 to −0·2)	<0·0001	−0·3 (−0·3 to −0·2)	<0·0001
Malignancy	127 (0·9%)	159 (0·9%)	156 (0·8%)	206 (1·0%)	206 (1·1%)	213 (1·0%)	227 (1·1%)	231 (1·0%)	236 (1·0%)	260 (1·1%)	308 (1·3%)	0 (0 to 0·1)	0·07	0 (0 to 0·1)	0·04	0 (0 to 0)	<0·01
At least 1 day in hospital	NA	NA	NA	NA	7388 (41·7%)	8126 (41·6%)	6744 (33·9%)	5461 (25·5%)	5118 (22·7%)	5090 (21·7%)	5220 (21·2%)	−0·1 (−0·9 to 0·6)	0·73	−2·8 (−3·0 to −2·6)	<0·0001	−3·9 (−4·1 to −3·8)	<0·0001
Treatments																	
Hypertonic saline	3575 (26·8%)	3926 (27·2%)	6802 (36·6%)	7512 (38·4%)	7882 (39·7%)	8195 (39·8%)	8891 (41·5%)	9457 (41·8%)	9104 (39·2%)	8771 (36·5%)	8883 (35·7%)	2·8 (2·7 to 3·0)	<0·0001	−1·7 (−1·9 to −1·6)	<0·0001	0·7 (0·6 to 0·7)	<0·0001
Inhaled antibiotic	8207 (56·4%)	9316 (54·9%)	10 822 (58·3%)	11 584 (59·2%)	11 600 (57·8%)	11 729 (56·2%)	12 120 (56·7%)	11 972 (53·1%)	11 082 (47·8%)	10 094 (42·1%)	9511 (38·2%)	0·2 (0 to 0·4)	0·02	−4·8 (−5·0 to −4·6)	<0·0001	−1·9 (−2·0 to −1·8)	<0·0001
Bronchodilators	8557 (64·2%)	9174 (63·7%)	12 976 (69·9%)	14 139 (72·3%)	14 424 (72·6%)	15 054 (73·1%)	15 098 (71·5%)	14 718 (66·2%)	16 155 (70·8%)	16 492 (68·8%)	16 334 (65·8%)	2·0 (1·9 to 2·2)	<0·0001	−0·9 (−1·0 to −0·7)	<0·0001	0 (−0·1 to 0·1)	0·85
Oxygen therapy	1470 (10·5%)	1533 (10·1%)	1710 (9·2%)	1768 (9·0%)	1809 (9·3%)	1839 (8·9%)	1648 (7·7%)	1329 (5·9%)	1101 (4·7%)	1074 (4·5%)	1023 (4·1%)	−0·3 (−0·4 to −0·2)	<0·0001	−0·8 (−0·9 to −0·8)	<0·0001	−0·7 (−0·8 to −0·7)	<0·0001
RhDNase	8544 (55·3%)	8957 (52·6%)	9861 (52·4%)	10 408 (52·4%)	10 613 (52·6%)	11 431 (54·7%)	11 946 (55·8%)	11 974 (53·0%)	11 250 (48·4%)	10 280 (42·9%)	9919 (39·9%)	0 (−0·2 to 0·1)	0·75	−4·2 (−4·4 to −4·0)	<0·0001	−1·2 (−1·3 to −1·1)	<0·0001
Azithromycin	7286 (52·9%)	7580 (50·9%)	8691 (46·9%)	8748 (44·7%)	8654 (42·9%)	8927 (42·7%)	9143 (42·7%)	8958 (39·7%)	8292 (35·8%)	7306 (30·5%)	6959 (28·0%)	−2·1 (−2·3 to −2·0)	<0·0001	−3·9 (−4·0 to −3·7)	<0·0001	−2·3 (−2·4 to −2·2)	<0·0001
Ursodeoxycholic acid	5595 (36·1%)	5988 (34·8%)	5439 (28·9%)	6107 (30·8%)	6542 (32·4%)	6934 (33·1%)	7183 (33·5%)	7277 (32·1%)	7771 (33·3%)	7848 (32·5%)	8002 (32·1%)	−0·5 (−0·6 to −0·3)	<0·0001	−0·2 (−0·4 to −0·1)	<0·01	−0·1 (−0·1 to 0)	0·04

(Table 2 continues on next page)

www.thelancet.com/respiratory Published online June 3, 2026 https://doi.org/10.1016/S2213-2600(26)00149-9

	2014	2015	2016	2017	2018	2019	2020	2021	2022	2023	2024	2014–19 annual difference (95% CI)	p value	2020–24 annual difference (95% CI)	p value	2014–24 annual difference (95% CI)	p value
(Continued from previous page)																	
CFTR modulator	NA	NA	NA	NA	4133 (20·3%)	5991 (28·4%)	10542 (48·6%)	14412 (63·1%)	16506 (70·1%)	17866 (73·8%)	18754 (74·9%)	8·1 (7·7 to 8·5)	<0·0001	6·2 (6·0 to 6·4)	<0·0001	9·7 (9·6 to 9·8)	<0·0001
Ivacaftor	NA	NA	NA	NA	936 (4·6%)	1040 (4·9%)	1220 (5·6%)	1191 (5·2%)	767 (3·3%)	597 (2·5%)	536 (2·1%)	0·3 (0·2 to 0·5)	<0·0001	−1·0 (−1·0 to −0·9)	<0·0001	−0·5 (−0·6 to −0·5)	<0·0001
Lumacaftor–ivacaftor	NA	NA	NA	NA	2776 (13·6%)	2968 (14·1%)	2441 (11·3%)	1906 (8·3%)	682 (2·9%)	194 (0·8%)	76 (0·3%)	0·4 (0·2 to 0·7)	<0·01	−2·9 (−3·0 to −2·8)	<0·0001	−2·7 (−2·8 to −2·6)	<0·0001
Tezacaftor–ivacaftor	NA	NA	NA	NA	437 (2·1%)	2298 (10·9%)	4045 (18·7%)	1850 (8·1%)	977 (4·2%)	422 (1·7%)	337 (1·3%)	8·7 (8·4 to 9·1)	<0·0001	−4·0 (−4·1 to −3·9)	<0·0001	−1·4 (−1·4 to −1·3)	<0·0001
Elexacaftor–tezacaftor–ivacaftor	NA	NA	NA	NA	32 (0·2%)	312 (1·5%)	6219 (28·7%)	12159 (53·2%)	15349 (65·2%)	16928 (69·9%)	17866 (71·4%)	1·3 (1·2 to 1·5)	<0·0001	10 (9·8 to 10·2)	<0·0001	13·7 (13·5 to 13·8)	<0·0001
Intravenous antibiotics	NA	NA	NA	NA	7123 (46·5%)	7992 (43·2%)	6964 (33·9%)	5196 (24·2%)	4158 (18·5%)	4067 (17·6%)	3966 (16·5%)	−3·3 (−4·1 to −2·5)	<0·0001	−4·1 (−4·2 to −3·9)	<0·0001	−5·5 (−5·6 to −5·3)	<0·0001

N (%) by year. CFTR=cystic fibrosis transmembrane conductance regulator. NA=not applicable.

Table 2: **Description of complications and treatment in adults with cystic fibrosis, from 2014 to 2024**

Figure 3: **Median predicted survival age of the overall registry population**
Shadow represents 95% CIs.

should not be interpreted as causal effects at the individual level. Additionally, heterogeneity between countries in access to CFTR modulators, health-care delivery, and registry practices might influence observed trends. The aggregated nature of the analysis might therefore mask important between-country variability, and the findings should be interpreted as overall European patterns rather than uniform effects across all settings.

These clinical advances have also been translated into changing transplant dynamics. Despite the overall growth of the adult cystic fibrosis population, the annual number of lung transplants declined over the study period, whereas the number of individuals living with transplants increased. Increasingly, optimisation of medical therapy, including CFTR modulators, has become standard practice before transplantation and is associated with reduced transplant waiting lists, probably reflecting improvements in lung function, nutritional status, and exacerbation burden before transplant.[22] The incorporation of new disease-modifying therapies appears to improve overall clinical status and reduce the need for lung transplantation.[23] In particular, the introduction of ETI has been associated with substantial gains in lung function and survival, further supporting improved long-term outcomes in this population.[24] Overall, these trends underscore sustained progress in cystic fibrosis care, reflected in greater clinical stability and improved survival following transplantation.[25] This pattern probably reflects improved registry capture of patients after transplant alongside greater disease stability, fewer acute deteriorations requiring transplantation, and enhanced survival after the procedure—findings consistent with transplant registry analyses.[11–14]

Sex-related differences in cystic fibrosis outcomes have been consistently reported, with female individuals historically experiencing poorer survival and more rapid disease progression. Although our study did not specifically examine sex-stratified trends, it remains unclear whether the introduction of CFTR modulators

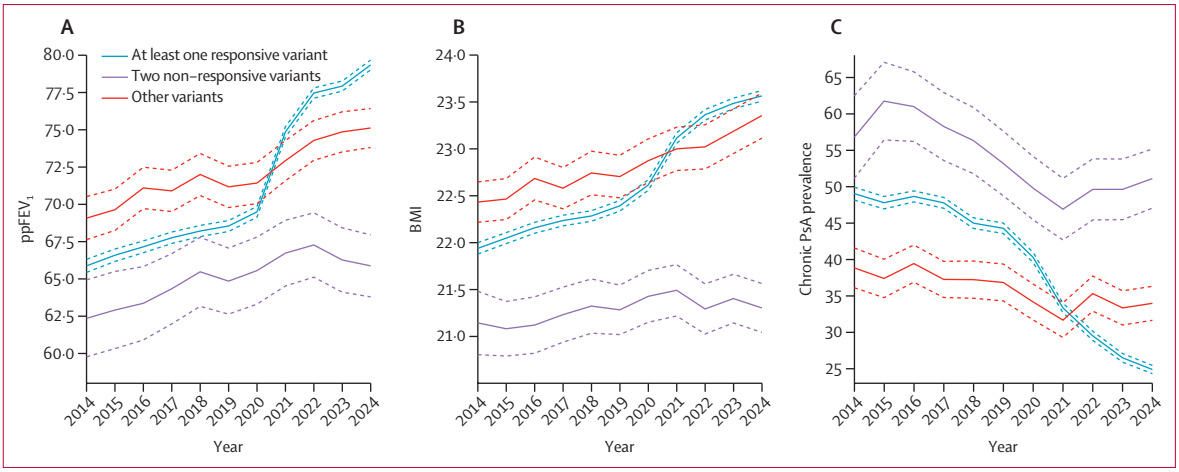

Figure 4: **ppFEV$_1$, BMI, and chronic *Pseudomonas Aeruginosa* infection from 2014 to 2024, in people with cystic fibrosis using or not using CFTR modulators**
Dashed lines represent 95% CIs. ppFEV$_1$=percent predicted FEV$_1$.

has mitigated these disparities. Future analyses using registry data should investigate whether sex-based differences persist in the modulator era.

Despite the notable advancements in the care of individuals with cystic fibrosis, these improvements present new challenges. As people with cystic fibrosis live longer, age-associated complications are becoming more prevalent. Our analysis documented a marked increase in impaired liver function and a modest but statistically significant rise in reported malignancies. These findings align with previous studies showing elevated cancer risk in older adults with cystic fibrosis, particularly gastrointestinal cancers.[26,27] These trends underscore the need to shift cystic fibrosis care beyond its historical paediatric model towards one that incorporates geriatric and chronic disease management frameworks. Cancer screening, bone health surveillance, cardiovascular risk assessment, and psychosocial support for older adults should be integrated into future care models.[1,2,7,9,28,29]

From a systems perspective, the growing adult cystic fibrosis population places increasing demands on health-care infrastructure. Multidisciplinary adult cystic fibrosis centres must expand their capacity and expertise, not only in pulmonary medicine but also in hepatology, endocrinology, oncology, reproductive health, and mental health.[30] The need for cross-specialty collaboration will only intensify as the median survival age continues to rise—a trend our data clearly document, with the median predicted survival age increasing from 50·6 years in 2010–14 to 69·6 years in 2020–24.[9]

Notably, although this study focused on 20 high-coverage countries, ensuring equitable access to CFTR modulators and high-quality care across all regions remains a pressing priority. Disparities in access to ETI—whether due to licensing, cost, or health system limitations—will probably widen outcome gaps in the coming years if not addressed proactively.[16] Policy makers must consider the long-term value of modulator therapies not only in terms of health improvements but also in reductions in hospital admissions, transplant needs, and broader societal costs.

The principal strength of our study lies in its large-scale, longitudinal design. Drawing on standardised, registry-based data spanning more than a decade and encompassing more than 80% of the European cystic fibrosis population, the analysis provides a robust picture of population-level changes in adult cystic fibrosis care. The use of generalised estimating equations models and survival analysis provides statistically rigorous insights into long-term trends.

Nonetheless, several limitations warrant mention. Limitations include the observational, cross-sectional design, which precludes individual-level causal inference, potential heterogeneity in reporting across centres, loss to follow-up, and possible sampling bias. Additional constraints include the absence of cardiovascular outcome data and insufficient detail on treatment adherence and socioeconomic or ethnicity determinants of health. The ECFSPR, however, applies rigorous data-quality controls and standardised definitions, which help mitigate inter-country heterogeneity.[7]

Interpretation of the observed decline in infection prevalence should take into account potential changes in sampling practices, particularly after 2020, and reduced health-care use during the COVID-19 pandemic, both of which could have influenced detection rates. Furthermore, residual confounding arising from concurrent improvements in infection control, nutritional status, or health-care access remains possible. Nevertheless, the magnitude and consistency of the observed trends support the interpretation of substantial improvements in outcomes occurring alongside the adoption of ETI. A key limitation of this study is the repeated cross-sectional design, which means annual estimates are conditional on individuals

being alive and recorded each year. As survival improves, the composition of the population changes over time, with an increasing representation of older, potentially healthier individuals. This aspect of the study might have contributed to observed improvements in clinical indicators independently of within-individual changes. Accordingly, the findings should be interpreted as reflecting dynamic changes in both population composition and clinical status over time, rather than true longitudinal trajectories at the individual level.

The decade from 2014 to 2024 has witnessed a paradigm shift in the health and demographics of adults with cystic fibrosis in Europe. Improvements in lung function, nutritional status, infection burden, and survival reflect both the maturation of comprehensive cystic fibrosis care and the transformative effect of CFTR modulators, particularly ETI. As the adult cystic fibrosis population continues to expand and age, care delivery models must evolve accordingly, and the need for cross-specialty collaboration will intensify as median survival increases. Ensuring equitable access to life-extending therapies, integrating age-appropriate chronic disease management, and investing in adult cystic fibrosis services will be essential to sustain the progress achieved to date. Our study provides important insights into the evolving landscape of cystic fibrosis care in the modulator era. At the same time, preserving expertise in the management of advanced and complex cystic fibrosis remains crucial, particularly for individuals who are not eligible for CFTR modulators and might continue to experience a more traditional disease trajectory. Health-care systems must therefore balance innovation with continuity, maintaining capacity for intensive, multidisciplinary cystic fibrosis care while adapting to the emerging needs of an ageing population.

Contributors

AO and EH contributed equally to the manuscript. AO, KDB, and EH were responsible for the study's conceptualisation. AO, KDB, EK, P-RB, AZ, EB, AO, and EH were responsible for data curation. AO and AZ were responsible for formal analysis. AO, AZ, and EH were responsible for the investigation. AO, KDB, EK, P-RB, EB, LN, and EH were responsible for the methods. AO and AZ were responsible for the software. AO and EH were responsible for visualisation. EH was responsible for supervision. AO was responsible for project administration. According to European Cystic Fibrosis Society Patient Registry (ECFSPR) rules and standard operating procedures, ECFSPR raw data are not directly available to all people working on a project, but only to the statisticians conducting the analysis. Two authors of this Article (AO and AZ) had direct access to the raw data and verified the underlying data used in the current research article. All other authors have access to aggregated data in the form of tables and figures, reporting completely anonymised information. AO, AZ, and EH were responsible for validation. All authors were responsible for writing the original draft. All authors critically reviewed and provided feedback on all manuscript versions. All authors had final responsibility for the decision to submit for publication.

Declaration of interests

KDB received consulting fees from Boehringer Ingelheim and Splisence and participated on Splisence's Data Safety Monitoring Board or Advisory Board. P-RB received institutional grants from Vertex and Vaincre la Mucoviscidose, and consulting fees from AstraZeneca, Chiesi, GSK, Insmed, MSD, Vertex, and Viatris, he received institutional support for attending meetings and travel from Chiesi and Viatris, and he is the President of the French Cystic Fibrosis Society. EB is the Director of the European Cystic Fibrosis Society Patient Registry, ECFSPR and has received payment or honoraria from Vertex Pharmaceuticals, scientific committee Nordic CF Masterclass. LN received Institutional fees for study participation from Vertex Pharmaceuticals and the German Centre for Lung Research. He is the Pharmacoepidemiology study director of the ECFSPR and the Medical lead of the German cystic fibrosis registry. He has also completed medical writing for Articulate Science. EH received Institutional fees for study participation from Vertex Pharmaceuticals; she received institutional support for attending meetings and travel from Chiesi, and honoraria for lectures from Vertex Pharmaceuticals. All other authors declare no competing interests.

Data sharing

Data are provided to ECFSPR under existing ethical approvals and data governance structures, and all participants or guardians signed informed consent before data collection (details are available at www.ecfs.eu/ecfspr). ECFSPR data are collected in accordance with agreed inclusion criteria, definitions, and coding standards, and are rigorously checked through several standardised measures. Additionally, the Data Quality Onsite Validation Programme validates data directly against the source at the centres, in accordance with SOP 131, Validation Visit at Source. This rich source of high-quality harmonised data provides information on many aspects of cystic fibrosis, is used to produce both annual reports, and is also available to support and foster scientific research. Data from the ECFSPR are available to researchers upon fulfilling specific criteria (a formal request outlining the intended use of the data and adherence to ethical and data protection regulations) and a rigorous application review process. Further information is available at coordination@ecfregistry.eu.

Acknowledgments

We thank the people with cystic fibrosis and their families for consenting to their data being included in the ECFSPR. We thank the centres and individual country representatives for allowing the use of the data, and the ECFSPR for providing access to anonymised patient data.

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