Cystic Fibrosis Trust

Since 1964 | We won't stop until CF does

UK Cystic Fibrosis Registry
2023 data highlights

Number of people with CF in 2023

11,319

registered* people

* People who have had at least one annual review recorded in the past three years

Births

116 women with CF had babies in 2023

31 men with CF became fathers in 2023

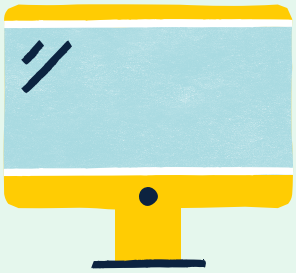
173 new diagnoses in 2023

124 of these were identified by new born screening

6,588

people with CF are 16 years or older.

Of these, **4,459 (68%)** were in work or study in 2023

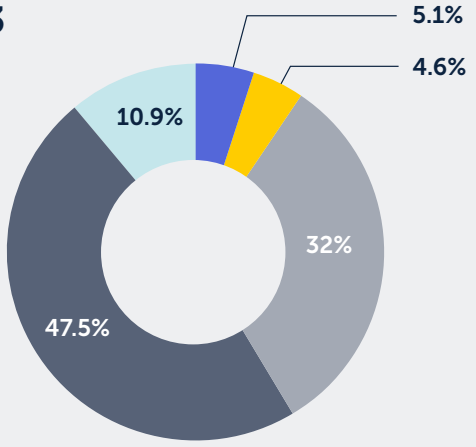


Ethnic group	%*
White	94.5
Asian	3.2
Black	0.3
Mixed	1.1
Other	0.9

* % of those with known ethnic group recorded

Genotype distribution in UK in 2023

- Homozygous F508del
- F508del + other*
- F508del + gating (incl. G551D)
- F508del + R117H
- Non-F508del combinations



* not gating or R117H

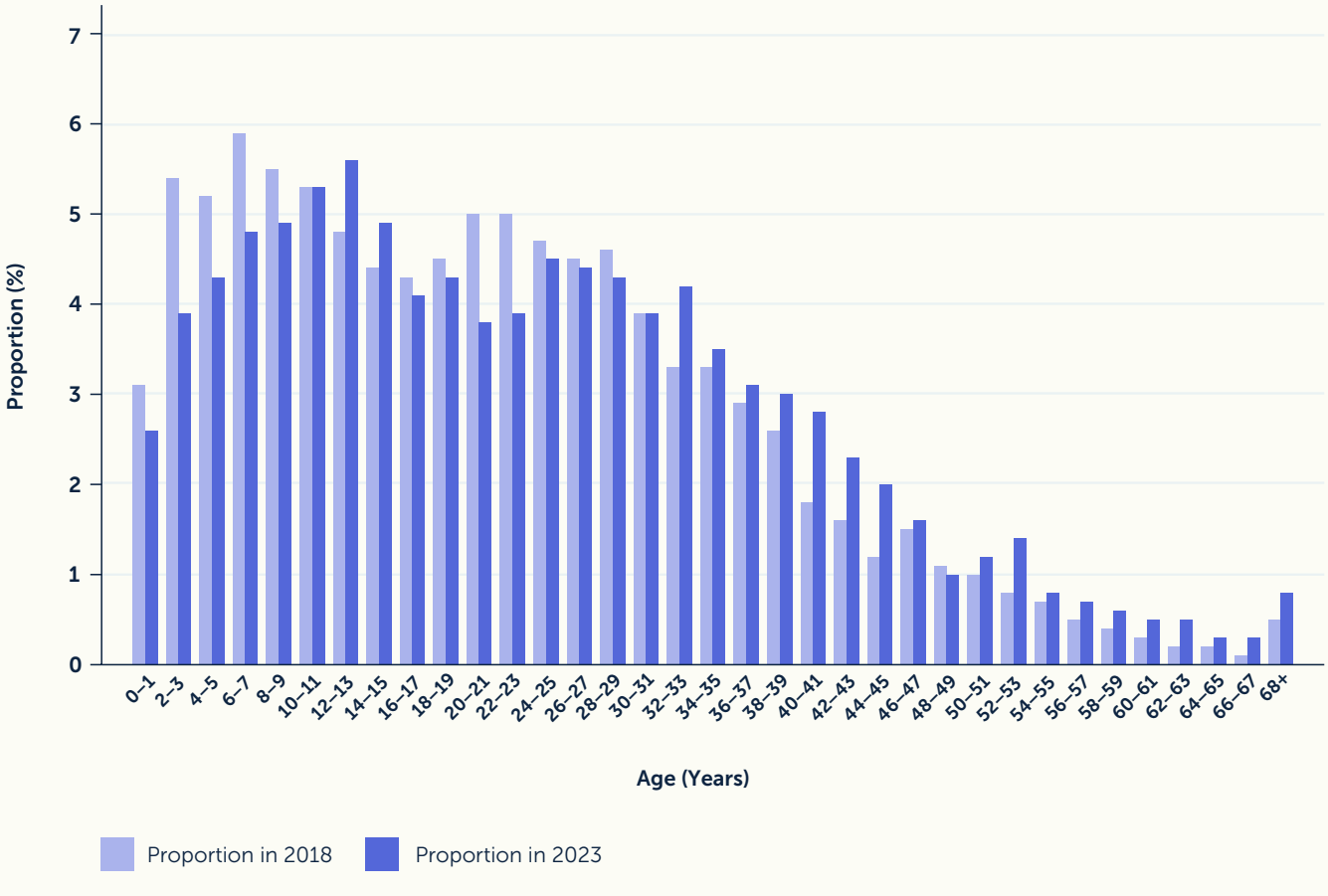
CFTR variant combinations in the UK population in 2023

This table shows the proportion (%) of people with the most common CFTR variant combinations in their genotype.

For example, **5.1%** of the UK population have one copy of F508del and one copy of R117H.

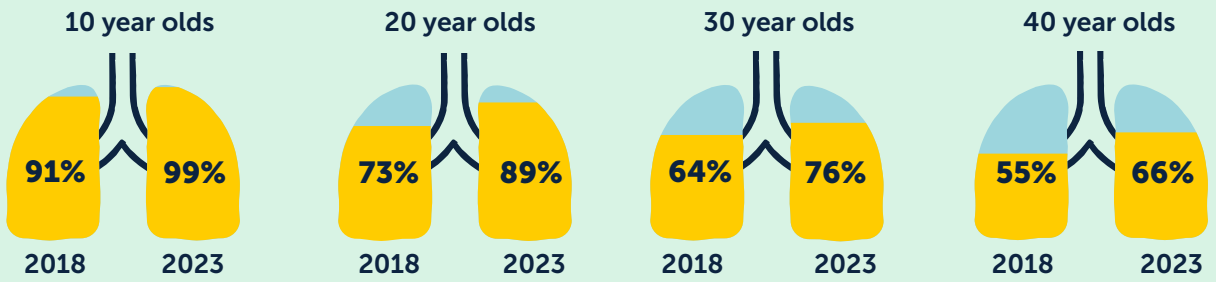
Mutation	F508del	R117H	G551D	G542X	621+1G->T	Other	Unknown	Total
F508del	47.5							47.5
R117H	**5.1**	0.1						5.2
G551D	4.0	0.2	0.2					4.4
G542X	2.5	0.1	0.1	0.1				2.8
621+1G->T	1.7	0.1	0.1	0.1	0.1			2.0
Other	26.8	0.6	1.0	0.8	0.5	5.5		35.2
Unknown	1.5	0.1	0.1	0.1	<0.1	0.5	0.7	2.9
Total	**89.1**	**1.2**	**1.4**	**1.0**	**0.6**	**6.0**	**0.7**	**100.0**

Age distribution of people with CF in 2018 and 2023



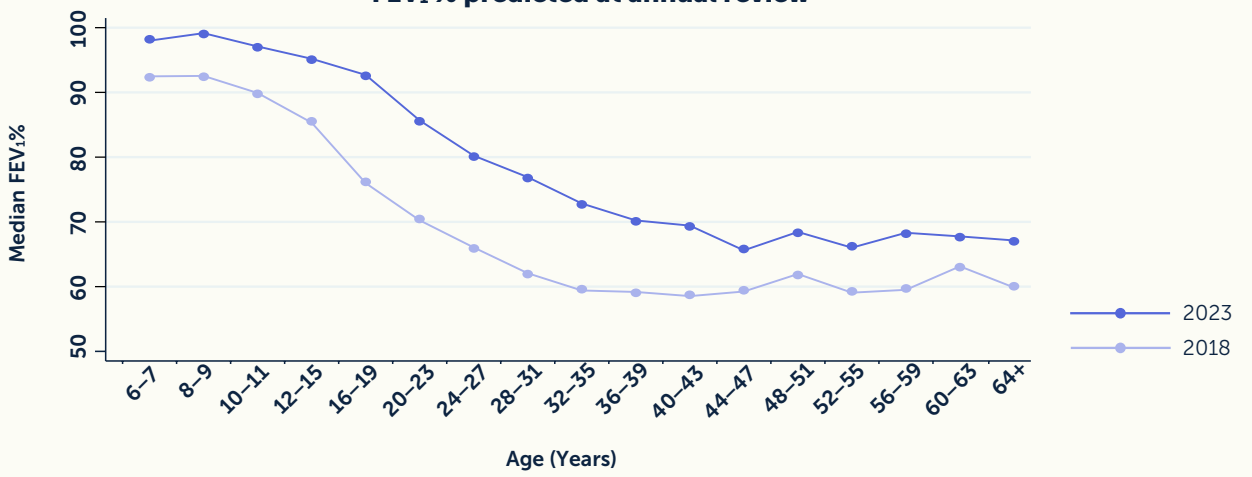
Lung health outcomes in 2018 and 2023

Median FEV₁% predicted at annual review



FEV₁% is reported for people who have not had a lung transplant

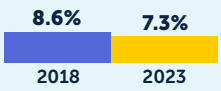
FEV₁ % predicted at annual review



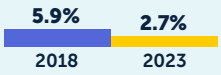
Lung infections and respiratory culture samples 2018 and 2023

Age under 16 years

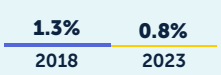
Chronic *Staphylococcus aureus*



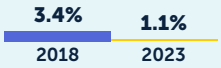
Chronic *Pseudomonas aeruginosa*



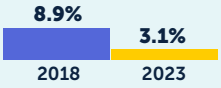
B. cepacia complex



Non-tuberculous mycobacteria (NTM)



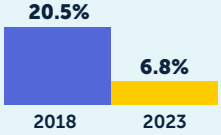
Aspergillus



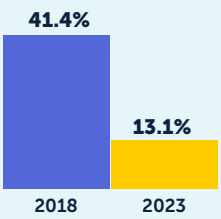
Age under 16 years	2018	2023
at least 3 samples of any type	97%	93%
at least 1 sputum sample	40%	30%
at least 1 cough sample	96%	98%

Age 16+ years

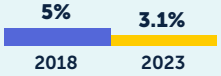
Chronic *Staphylococcus aureus*



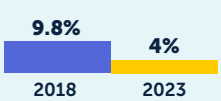
Chronic *Pseudomonas aeruginosa*



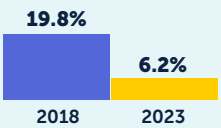
B. cepacia complex



Non-tuberculous mycobacteria (NTM)



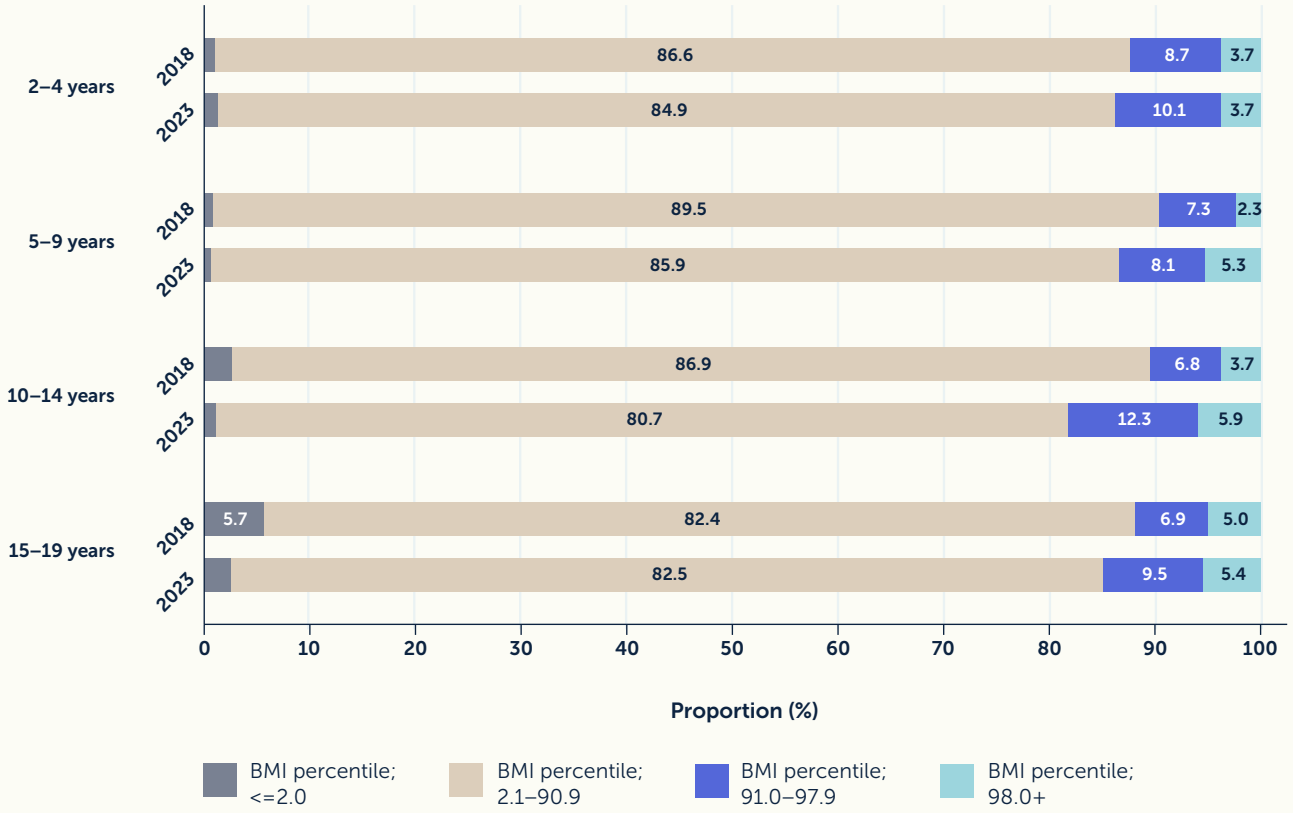
Aspergillus



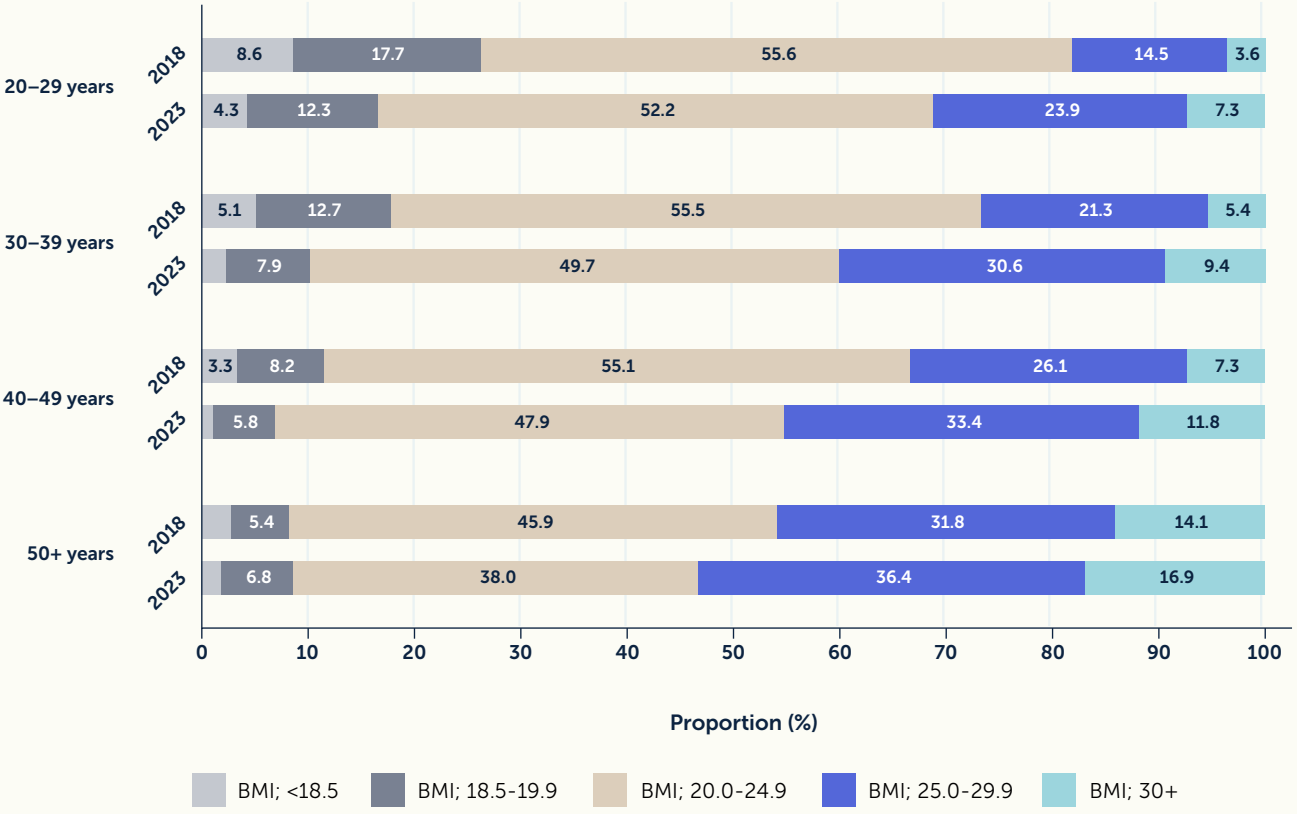
Age 16+ years	2018	2023
at least 3 samples of any type	80%	48%
at least 1 sputum sample	88%	73%
at least 1 cough sample	42%	53%

Body Mass Index: 2018 and 2023

BMI percentiles for children and young people



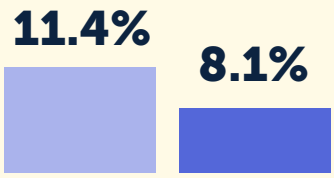
BMI in adults



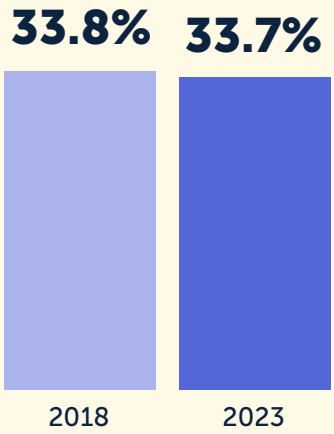
Complications in 2018 and 2023

CF Diabetes*

Age 10** to 15 years

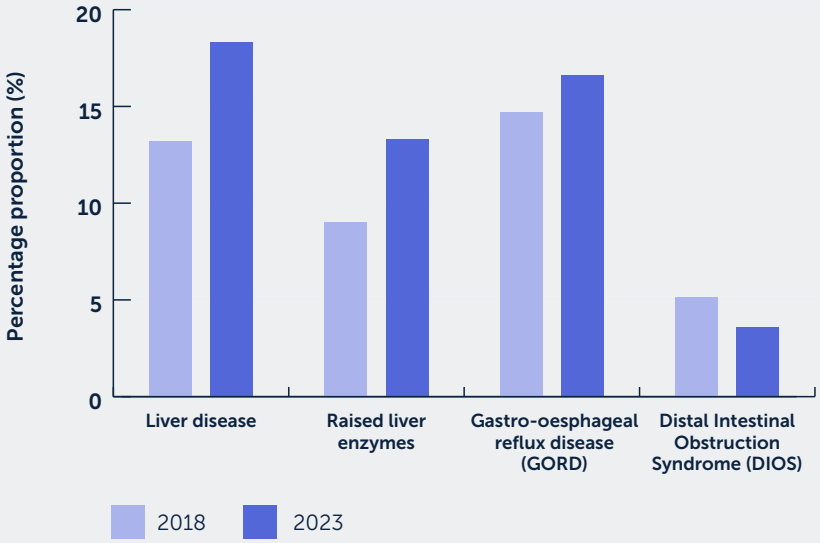


Age 16+ years

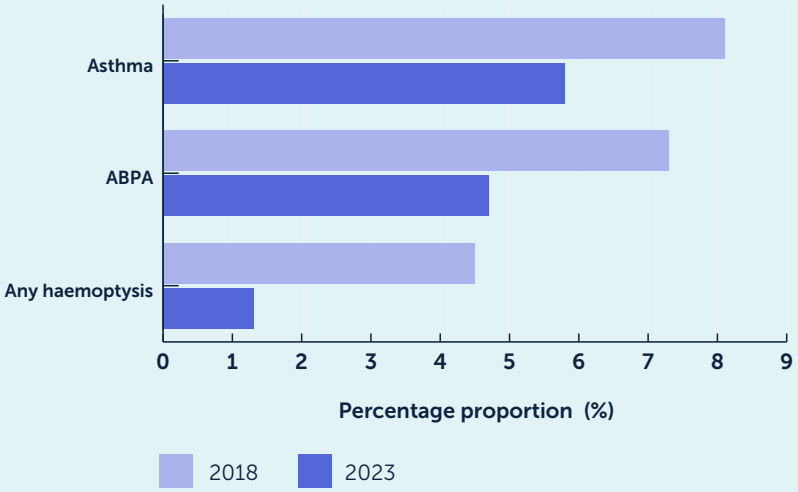


* on treatment for CF diabetes.
** in both 2018 and 2023, 1% of people under 10 years had CF diabetes

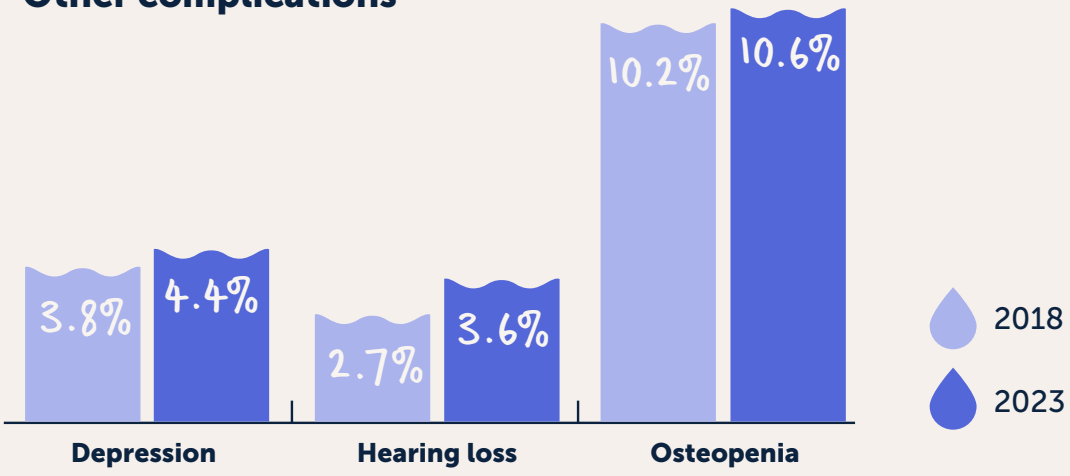
Liver and Gastrointestinal complications



Respiratory related complications



Other complications

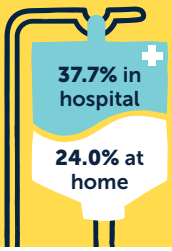


Medications and other treatments

Intravenous antibiotics (IV) in 2018 and 2023

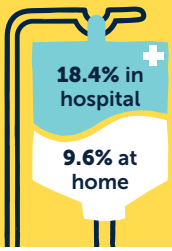
44.7%

of people had at least one course of IV antibiotics (at home or in hospital) in **2018**

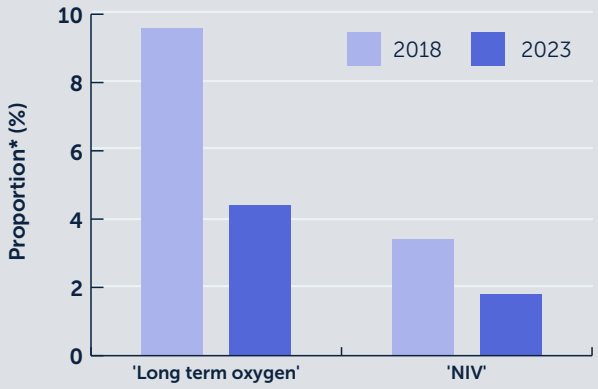


22.0%

of people had at least one course of IV antibiotics (at home or in hospital) in **2023**

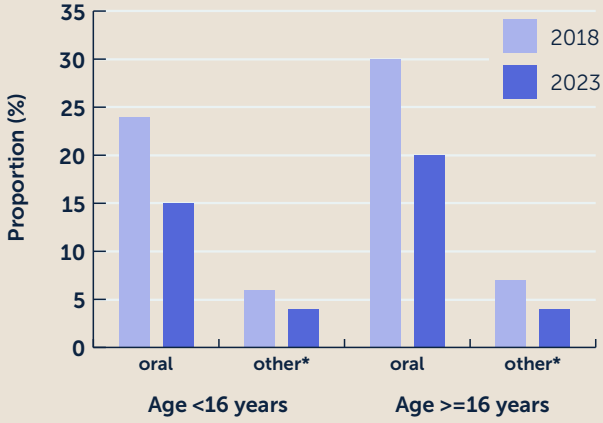


Respiratory support



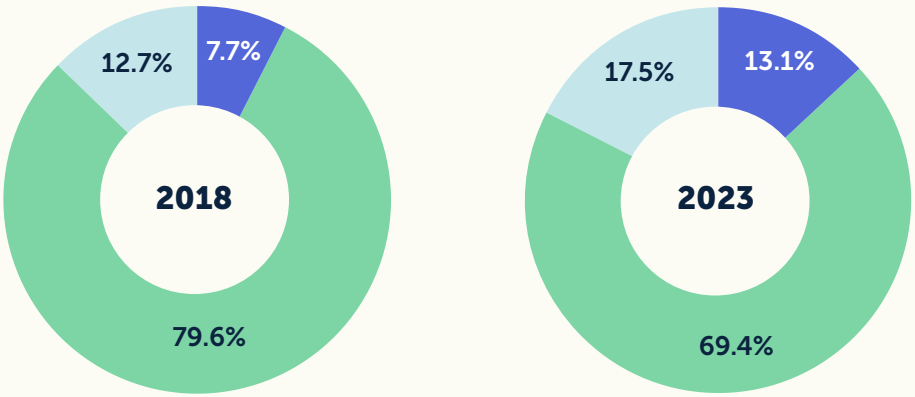
* age 18+ years

Supplemental feeding



* enteral and parenteral feeding

Primary airway clearance



* includes all varieties of ACT inc. breathing ex, manual techniques and devices

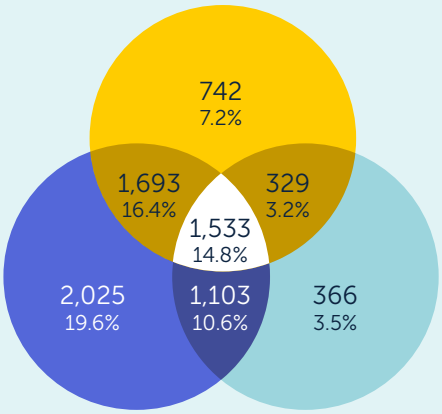
Inhaled medications therapy in 2023

The Venn diagram shows how many people with CF are on one or more of some inhaled therapies and the combinations they take.

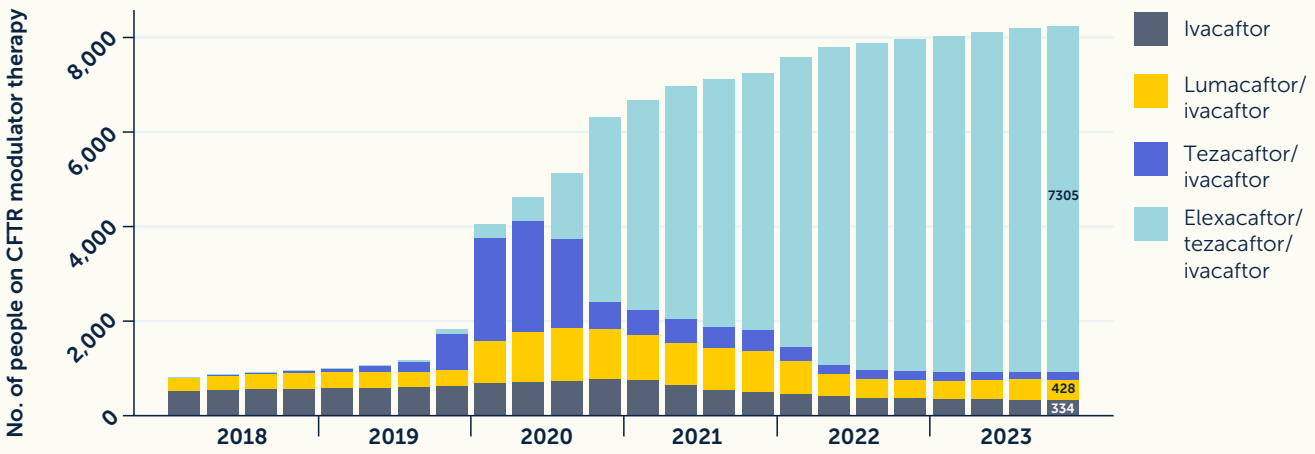
The proportion of people taking all 3 types of inhaled medication has fallen from 21.4% in 2018 to 14.8% in 2023.

None of these inhaled medications: 2,553 (24.7%)

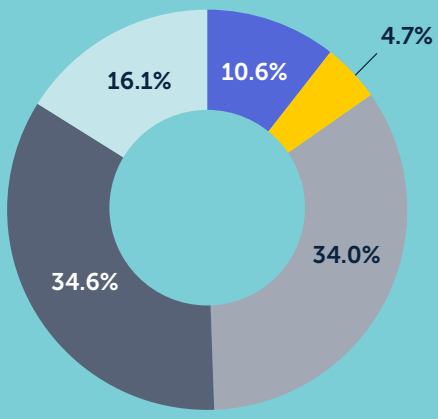
- Inhaled antibiotics
- DNase
- Hypertonic saline or mannitol



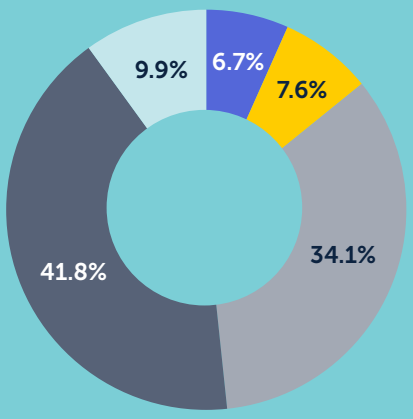
Quarterly CFTR modulator use since 2018



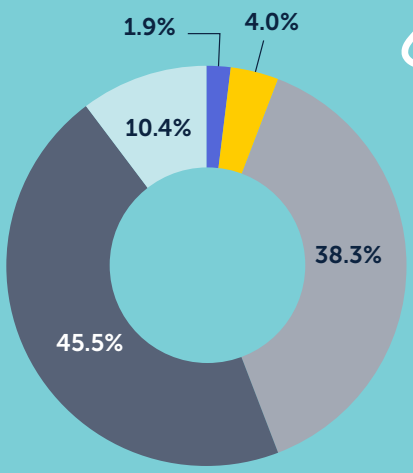
Genotype distribution by devolved nation



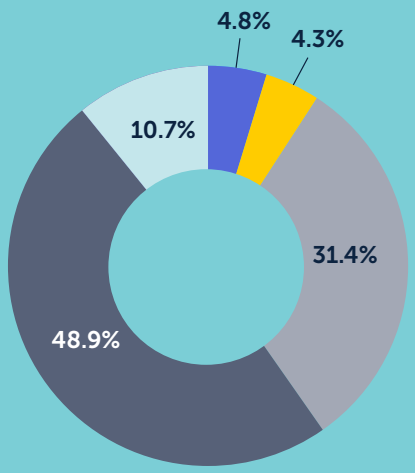
Northern Ireland
N=509



Scotland
N=972



Wales
N=473



England
N=9365

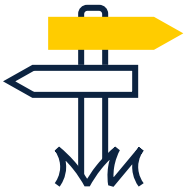
Homozygous F508del | **F508del + other*** | **F508del + gating (incl. G551D)** | **F508del + R117H** | **Non-F508del combinations**

*not gating or R117H

The UK Cystic Fibrosis Registry

The UK CF Registry has been sponsored and hosted by Cystic Fibrosis Trust since 2007. Data collected as part of the routine clinical care of a person with CF is recorded on the UK CF Registry, with consent of the person (or their parent or guardian if they are a child).

The purpose of the UK CF Registry is to improve the health of people with cystic fibrosis in the UK by:



helping people with CF and their families understand CF, and make informed decisions

giving clinical teams the evidence they need to improve the quality of care

monitoring the safety and effectiveness of new treatments for cystic fibrosis

providing data to researchers

helping commissioners provide funding to NHS CF centres that is proportionate to the severity of their patients' condition

supporting clinical trials through feasibility studies and pragmatic data collection

About the UK CF Registry

- A key strength of the UK CF Registry is the population level coverage, providing insights into the natural history of CF and the demographic profile of the CF population of the UK.
- The longitudinal nature of the Registry data allows researchers to study disease progression, long-term outcomes and treatment patterns over time[1].
- The data reflects real-world clinical practice and patient experiences, providing crucial insights into managing a rare disease in routine settings.
- The UK CF Registry is recognised in the NICE real-world evidence framework[2] as an exemplar source of real-world data. UK CF Registry data has been collected and analysed to support technology appraisal submission.
- The Registry has a track record[3] of delivering post-marketing pharmacovigilance studies such as Post-Authorisation Safety Studies (PASS)[4,5], reporting with long-term safety outcomes to support regulatory submissions.
- UK CF Registry team can support with study protocol design, including defining inclusion and exclusion criteria to appropriately select study cohorts. Statistical analysis support can also be provided.
- Registry data can inform the design and planning of clinical trials through our feasibility service and/or data request process.

To find out more, apply for data, or seek support for your project, please visit our website: **cysticfibrosis.org.uk/registry**

Contact us: **registry@cysticfibrosis.org.uk**

References:

1. Granger E, Davies G, Keogh RH. Treatment patterns in people with cystic fibrosis: have they changed since the introduction of ivacaftor? J Cyst Fibros. 2022 Mar;21(2):316-322. doi: 10.1016/j.jcf.2021.08.014. Epub 2021 Sep 6. PMID: 34497037; PMCID: PMC9097695.
2. NICE real-world evidence framework. 2022. https://www.nice.org.uk/corporate/ecd9/chapter/overview
3. Bilton D, Caine N, Cunningham S, Simmonds NJ, Cosgriff R & Carr SB. Use of a rare disease patient registry in long-term post-authorisation drug studies: a model for collaboration with industry. Lancet Respir Med. 2018 Jul; 6(7): 495-496.
4. Kaplan S, Lee A, Caine N, Charman SC, Bilton D. Long-term safety study of colistimethate sodium (Colobreathe®): Findings from the UK Cystic Fibrosis Registry. J Cyst Fibros. 2021 Mar;20(2):324-329.
5. Volkova N, Moy K, Evans J, Campbell D, Tian S, Simard C, Higgins M, Konstan MW, Sawicki GS, Elbert A, Charman SC, Marshall BC, Bilton D. Disease progression in patients with cystic fibrosis treated with ivacaftor: Data from national US and UK registries. J Cyst Fibros. 2020 Jan;19(1):68-79

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