# Cystic Fibrosis strength in numbers

# UK CF Registry highlights 2018

## 2018 Cohort

**9,847**
**Annual Reviews**

**10,509**
**Number of active patients**

Active patients are people who are currently alive and have submitted an annual review in the last three years.

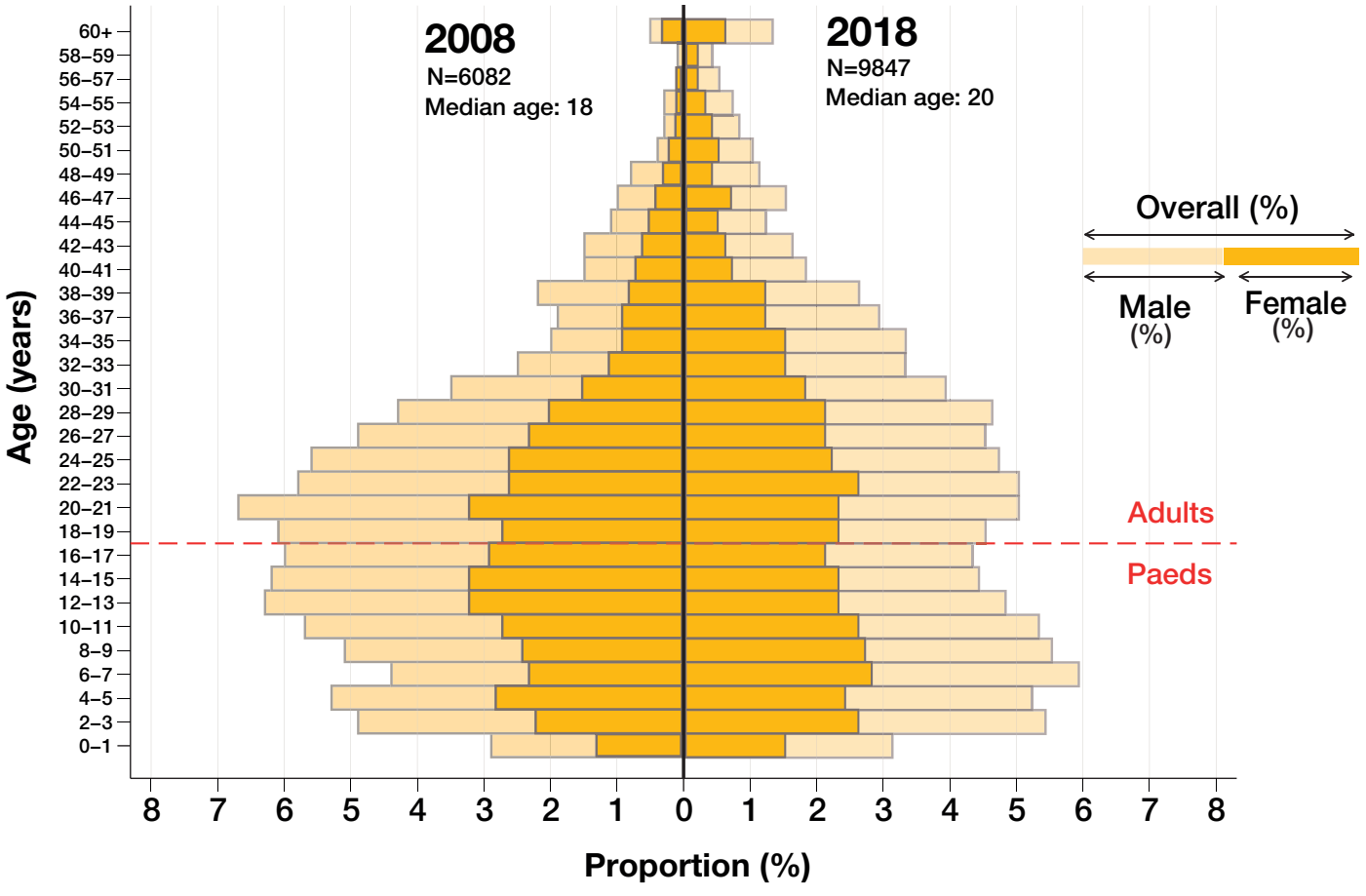
## Life statistics

**65** women with CF had babies in 2018

**45** men with CF became fathers in 2018

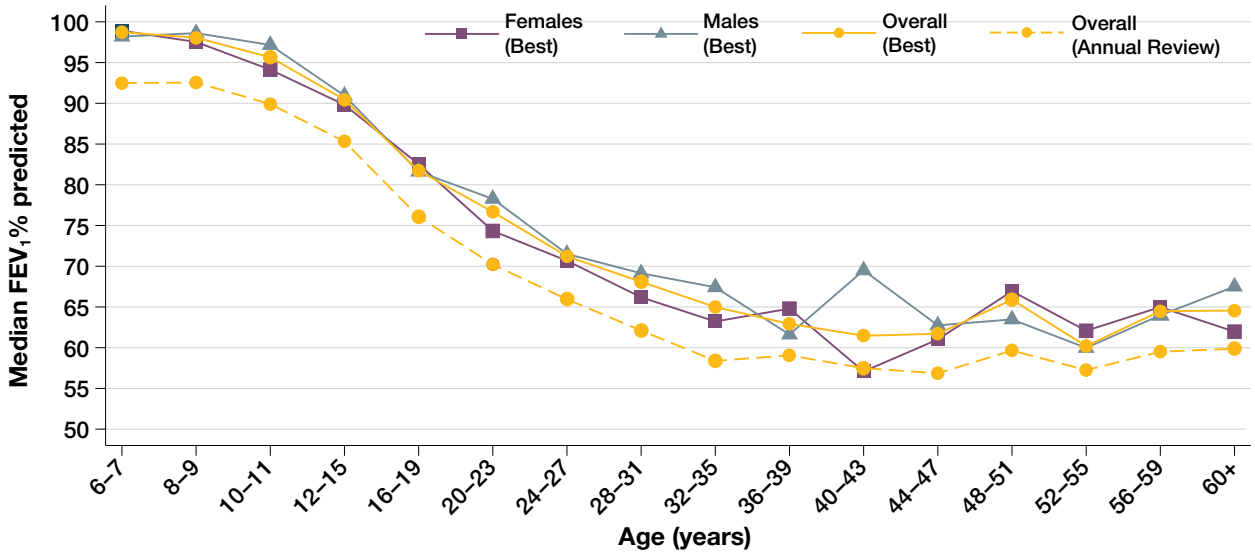
**64%** people aged 16 years and over were in work or study

## Age and sex distribution of the UK CF population in 2008 vs 2018



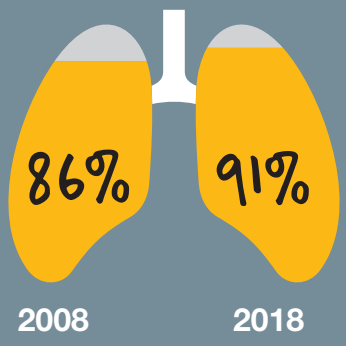

Note the different demographic distribution across the years, namely higher proportion in older age groups in 2018.

## $FEV_1$% predicted

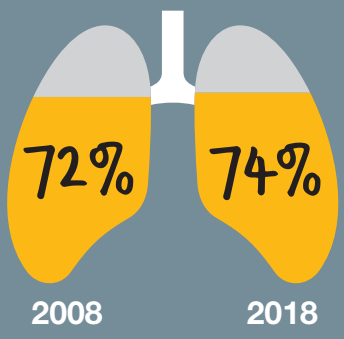



## $FEV_1$% predicted in 2008 vs 2018

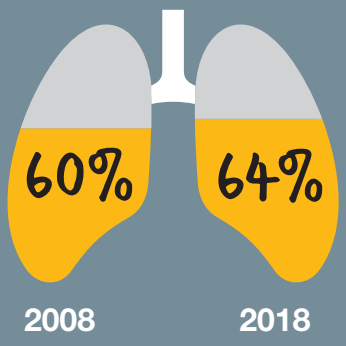
### 10 year olds
Median $FEV_1$% predicted




### 18 year olds
Median $FEV_1$% predicted




### 30 year olds
Median $FEV_1$% predicted




**$FEV_1$% is reported for people who have not had a lung transplant.**

## Transplant

| 2008 | | 2018 |
|---|---|---|
| 126 | Evaluated | 247 |
| 55 | Accepted | 104 |
| 16 | Double lung transplant | 58 |
| 8 | Other transplant | 7 |

## Nutrition

**50**
Median BMI Percentile
Age 2-17
**2008**

**52**
Median BMI Percentile
Age 2-17
**2018**

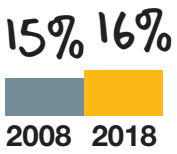
**41%**
Adults meeting
BMI target
**2008**

**48%**
Adults meeting
BMI target
**2018**

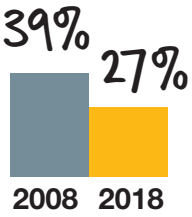
**The target BMI is 23kg/m² for males and 22kg/m² for females.**

## Changes in infection rates

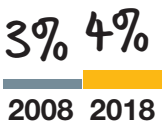
Chronic *Staphylococcus aureus*



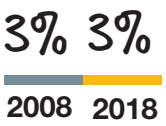

Chronic *Pseudomonas aeruginosa*



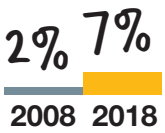

*B. cepacia* complex



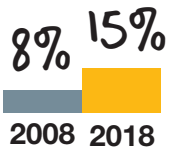

MRSA




NTM

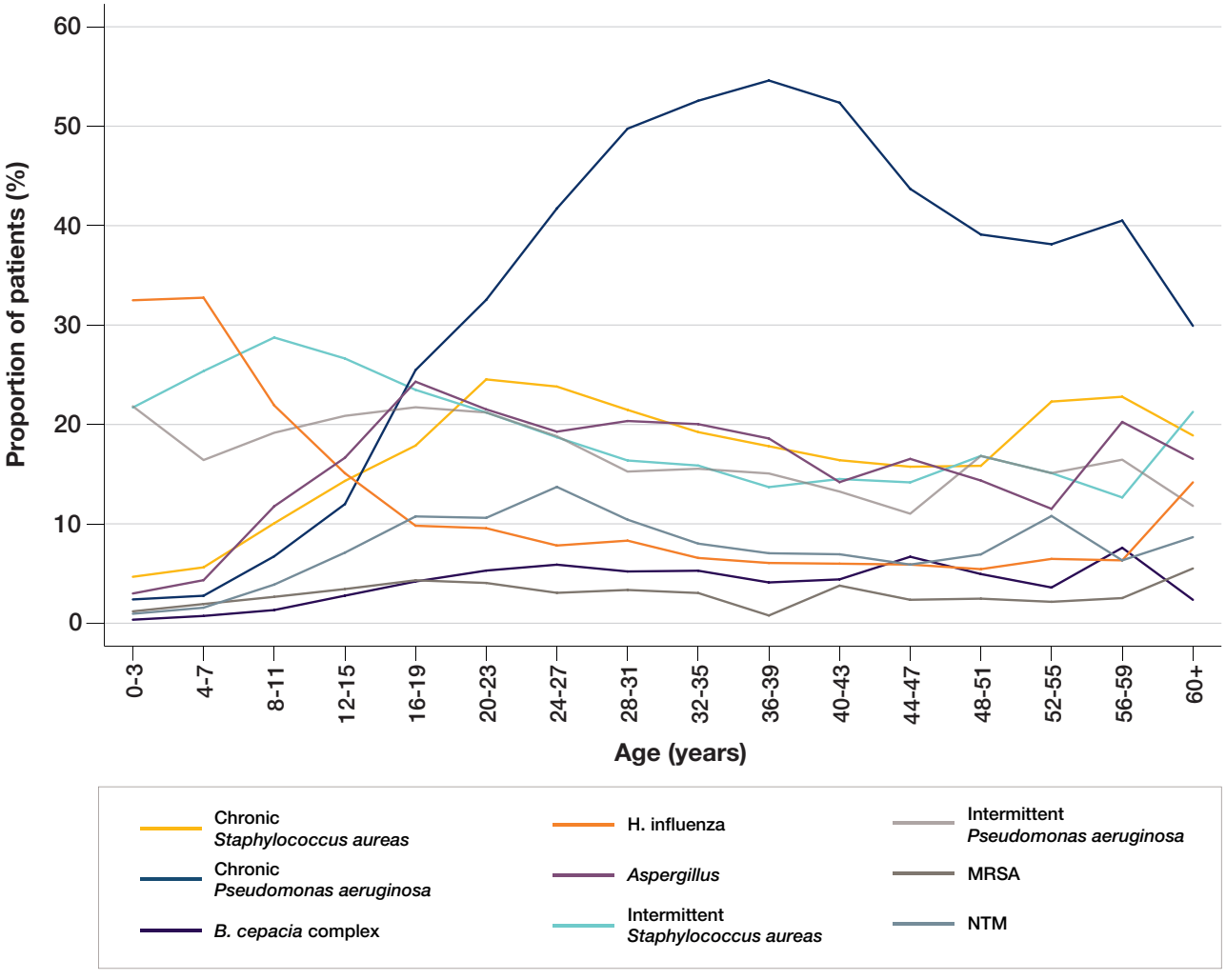



*Aspergillus*

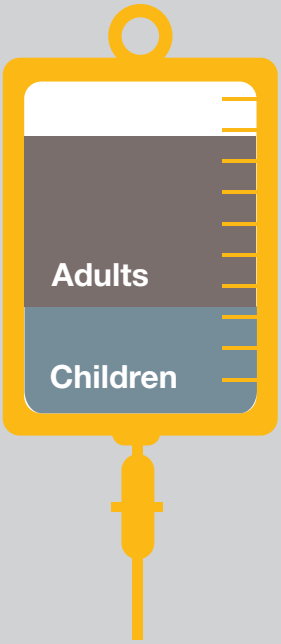



The Registry diagnosis of chronic infection requires three positive sputum samples in the past 12 months.

## Lung infections in 2018



## IV antibiotics

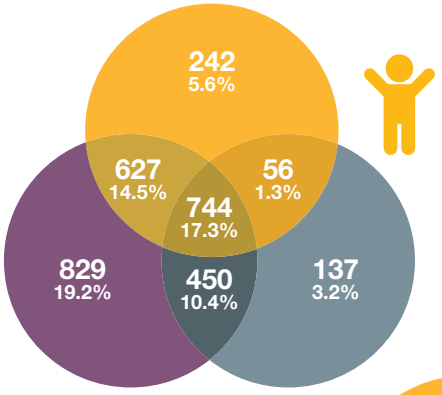



54%

33%

**The proportion of patients receiving at least one home or hospital IV course in 2018.**

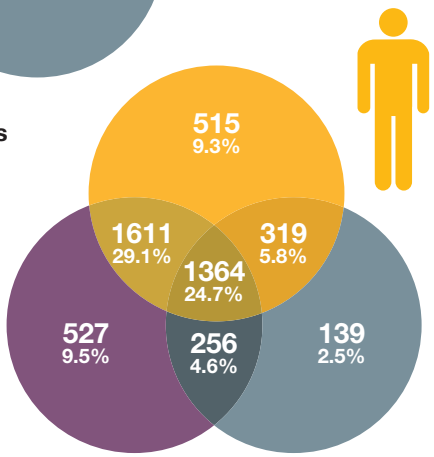
## Inhaled medicines




**<18 years N=4314**

No inhaled therapy: 1116 (25.9%)

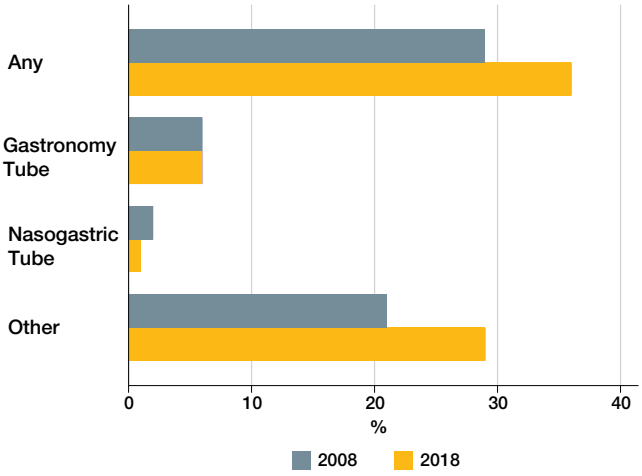
- Inhaled antibiotics
- DNase
- Hypertonic saline or mannitol



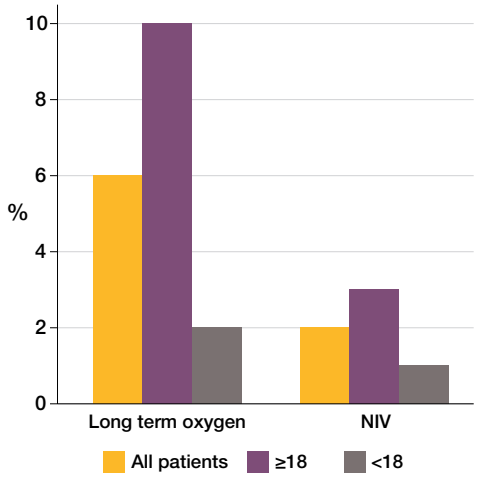

**≥18 years N=5533**

No inhaled therapy: 715 (12.9%)

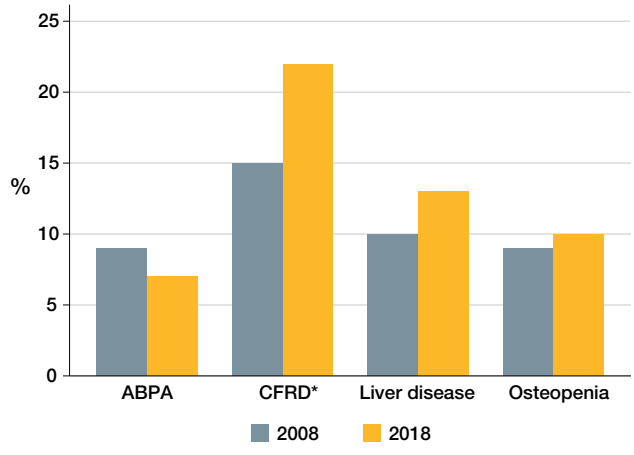
## Supplemental feeding




## Respiratory support in 2018

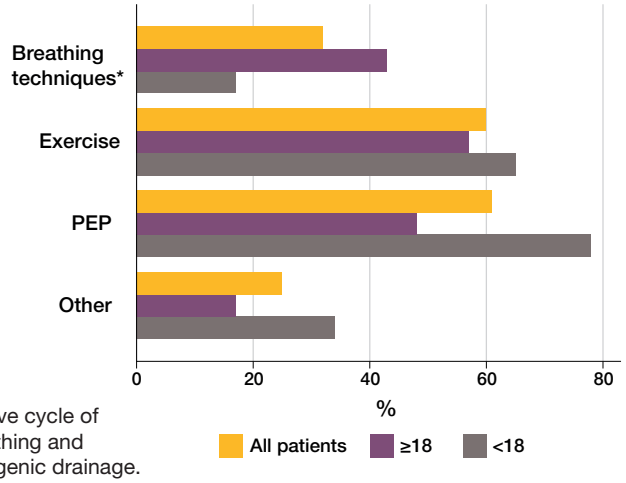



## Complications



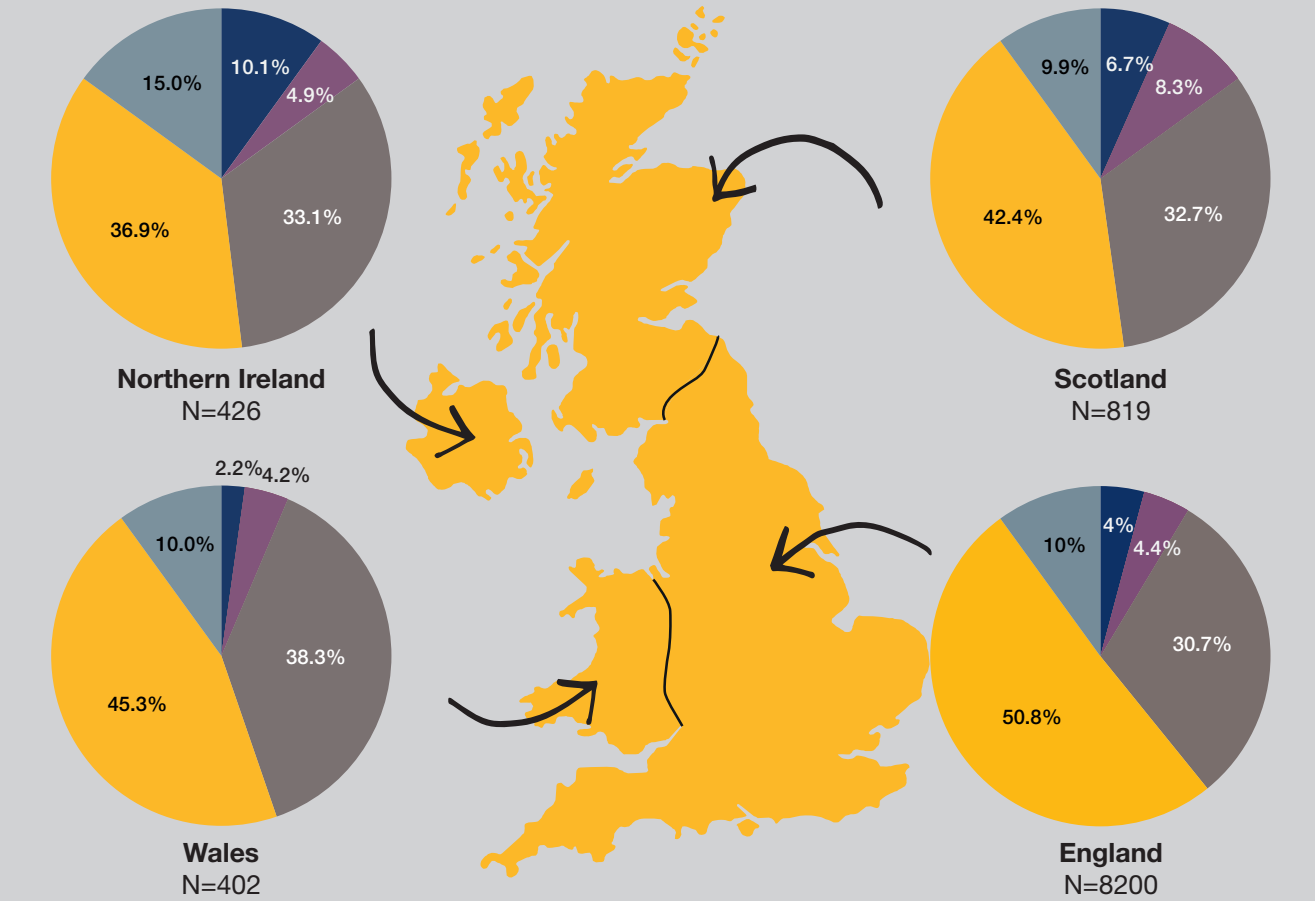

*On CFRD treatment.

## Physiotherapy in 2018

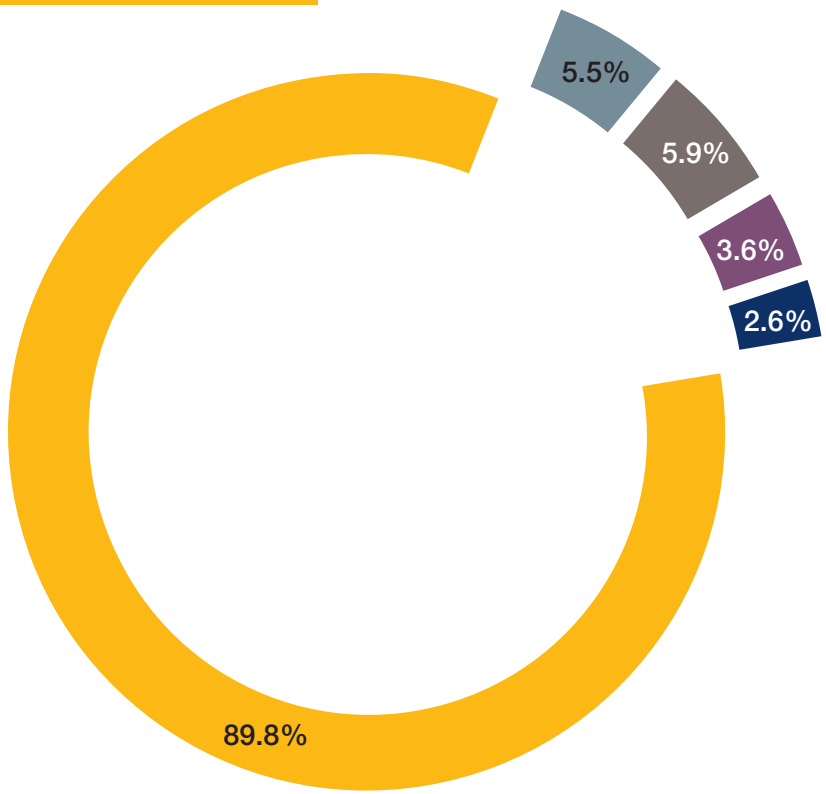



*Active cycle of breathing and autogenic drainage.

## Genotype distribution by devolved nation




## Mutation frequency




Proportion of people with CF in the UK who carry at least one of these most common five mutations.

**99.1% of people have at least one mutation recorded.**

## Quality of data

**100%** UK CF centre participation

**99%** Participation of people with CF

**94%** Active patients with Annual Review

**99%** Key variables complete

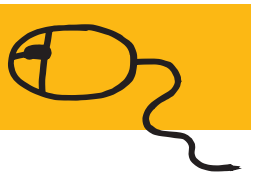
**98%** Key variables accurate

## UK CF Registry data request service

A vital function of the UK CF Registry is to make pseudonymised, anonymised and aggregated data available to researchers from recognised institutions, for the benefit of people with cystic fibrosis.

Requests for access to Registry data are evaluated by the independent UK CF Registry Research Committee, which ensures that the requested data are available and the research team are appropriately configured to receive the data in the format requested. The data request form, along with the Data Sharing Policy, and a list of previous data requests can be accessed via the Cystic Fibrosis Trust website.

**Request UK CF Registry data: www.cysticfibrosis.org.uk/registry**



## Working with industry to make medicines safer

Pharmacovigilance, or 'drug-safety', studies monitor the long-term safety of drugs in everyday use, once they have completed clinical trials and been given a license. There are key advantages to cystic fibrosis pharmacovigilance studies, which usually span a five-year period, being run within the UK CF Registry.

### Quality of the study

The UK CF Registry is a powerful tool for the monitoring of long-term outcomes in a large cohort of people. It allows access to a comparator cohort that are naïve to the study drug, which may not be possible in other models of drug-safety studies. The scope of the information recorded by the UK CF Registry enables detailed comparisons of patient cohorts receiving different medications, with the ability to disentangle drug safety signals from the numerous other aspects of cystic fibrosis.

### Independence

The principal investigator for each study is an NHS CF physician who provides independent clinical expertise. The UK CF Registry statisticians provide input to the study design, conduct the analysis and contribute to reporting and interpretation of the results. Summary data only (no patient-level data relating to individuals) are provided to the pharmaceutical company, and results and their interpretation are agreed by the experts independent of the industry collaborators.

**References**

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.

Taylor-Robinson D, Archangelidi O, Carr SB, Cosgriff R, Gunn E et al. Data Resource Profile: The UK Cystic Fibrosis Registry. Int J Epidemiol. 2018 Feb; 47(1): 9-10e.