

# 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

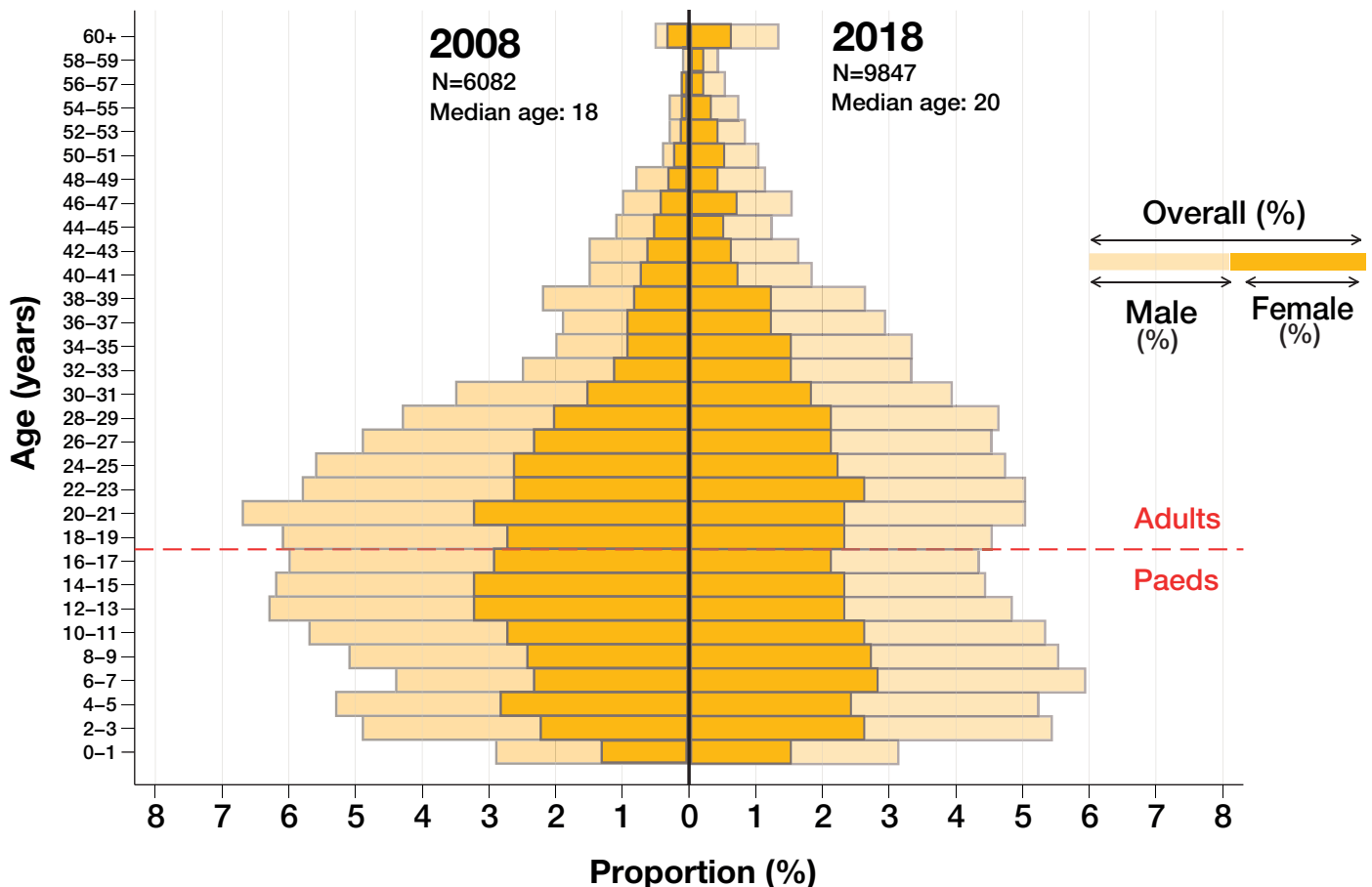
45 men with CF became fathers in 2018

64%

people aged 16 years and over were in work or study

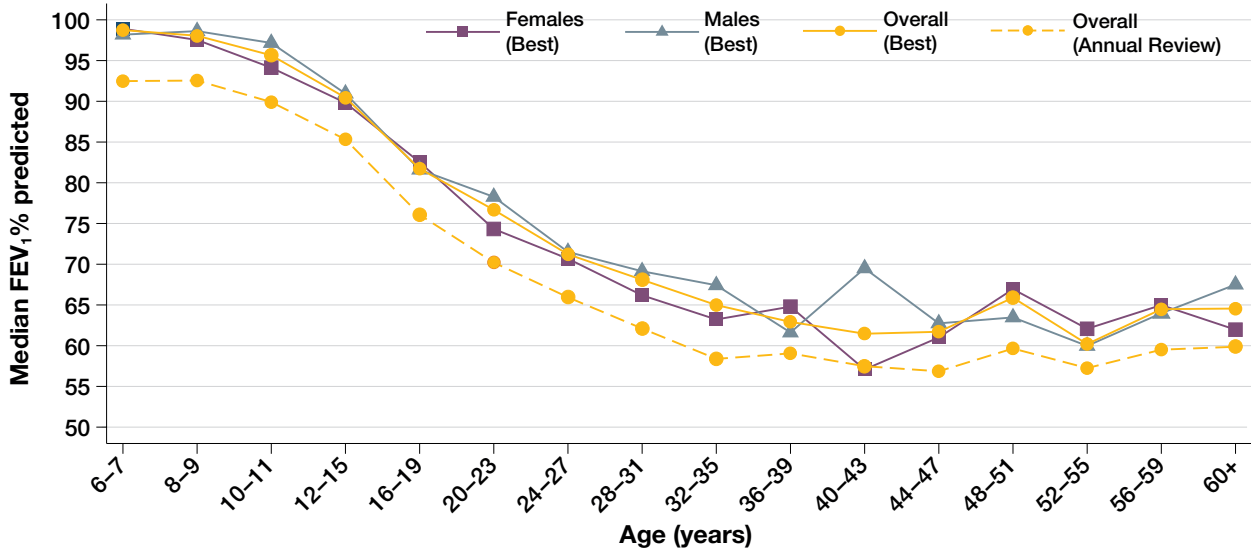


### Age and sex distribution of the UK CF population in 2008 vs 2018



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

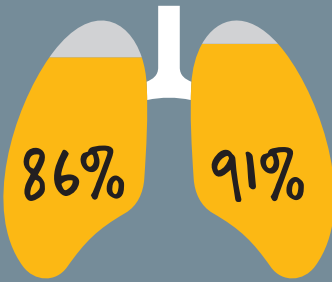
## FEV<sub>1</sub>% predicted



## FEV<sub>1</sub>% predicted in 2008 vs 2018

### 10 year olds

Median FEV<sub>1</sub>% predicted

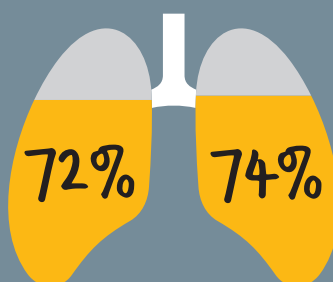


2008

2018

### 18 year olds

Median FEV<sub>1</sub>% predicted

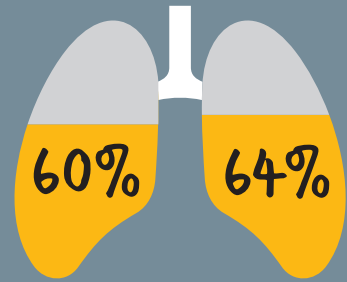


2008

2018

### 30 year olds

Median FEV<sub>1</sub>% predicted



2008

2018

FEV<sub>1</sub>% 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%

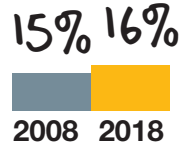


Adults meeting  
BMI target  
2018

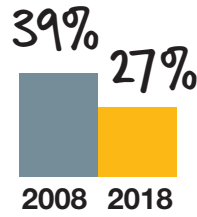
The target BMI is 23kg/m<sup>2</sup> for males and 22kg/m<sup>2</sup> for females.

## Changes in infection rates

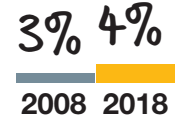
Chronic *Staphylococcus aureus*



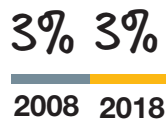
Chronic *Pseudomonas aeruginosa*



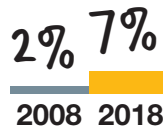
*B. cepacia* complex



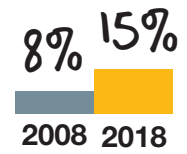
MRSA



NTM

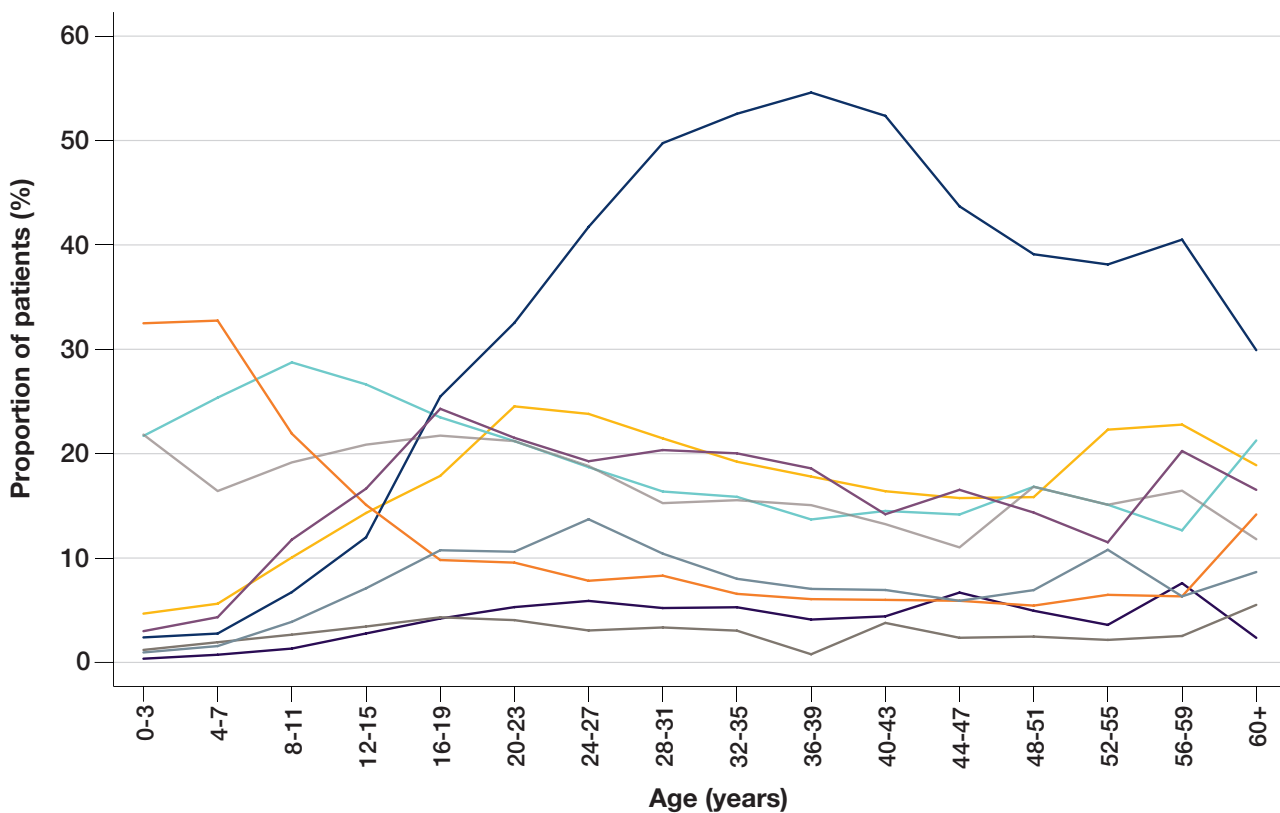


*Aspergillus*

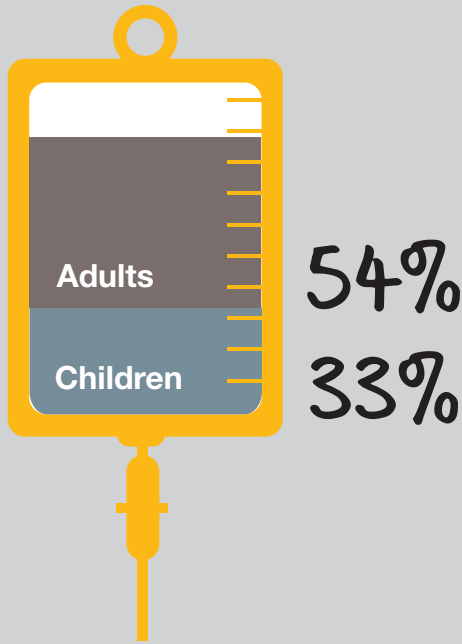


The Registry diagnosis of chronic infection requires three positive sputum samples in the past 12 months.

## Lung infections in 2018

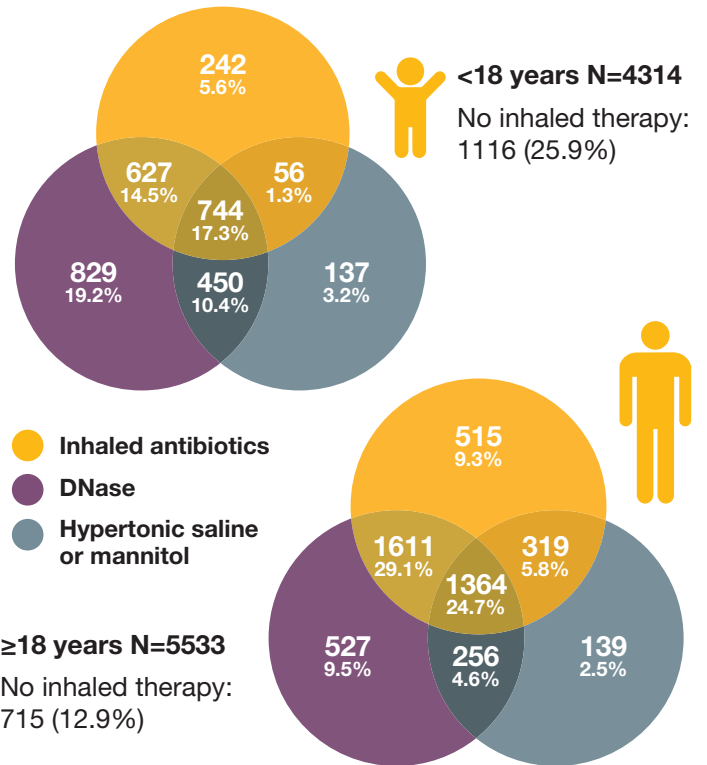


## IV antibiotics

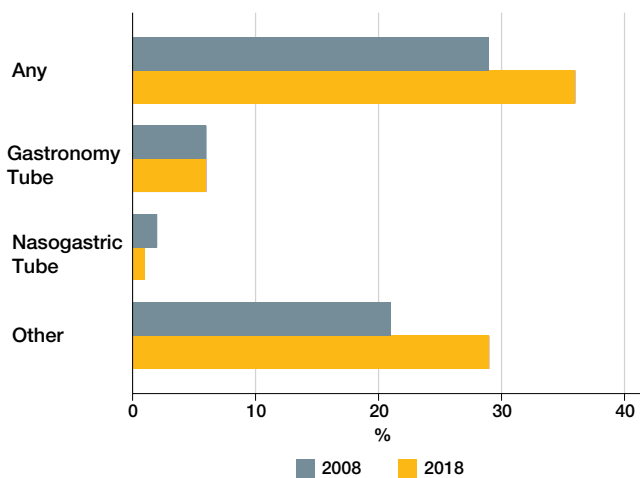


The proportion of patients receiving at least one home or hospital IV course in 2018.

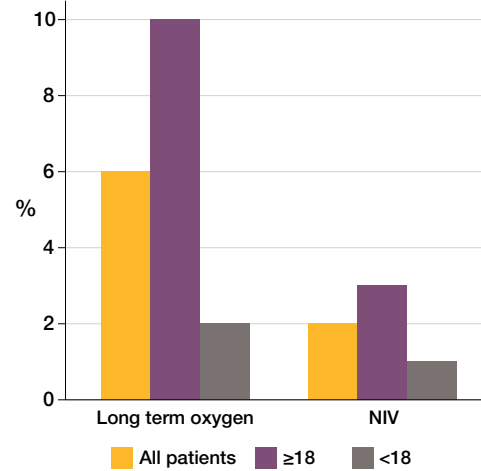
## Inhaled medicines



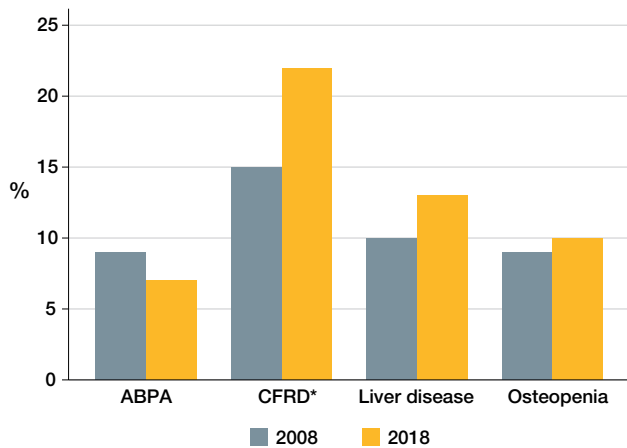
## Supplemental feeding



## Respiratory support in 2018

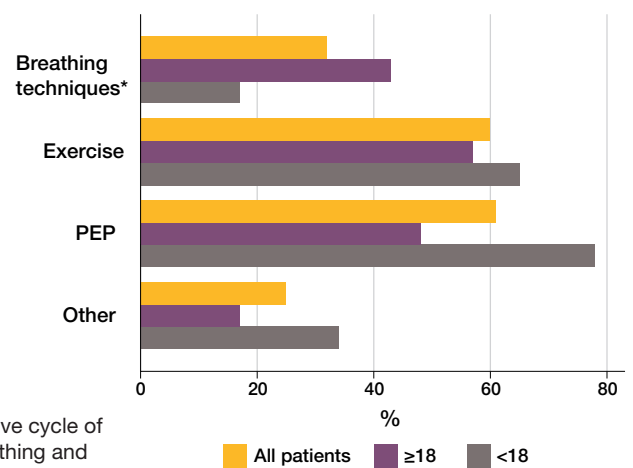


## Complications



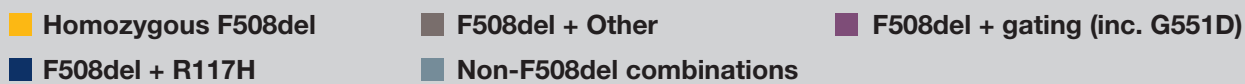
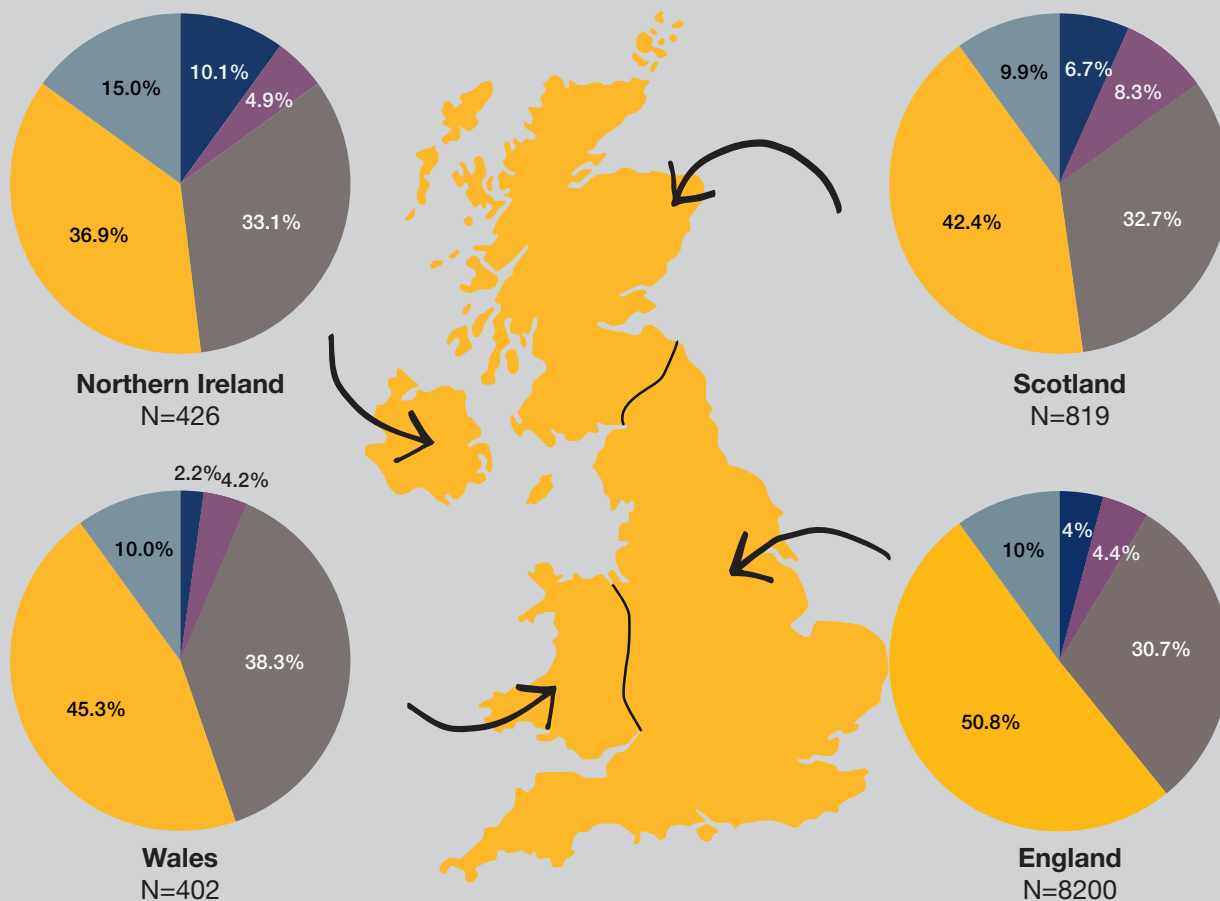
\*On CFRD treatment.

## Physiotherapy in 2018

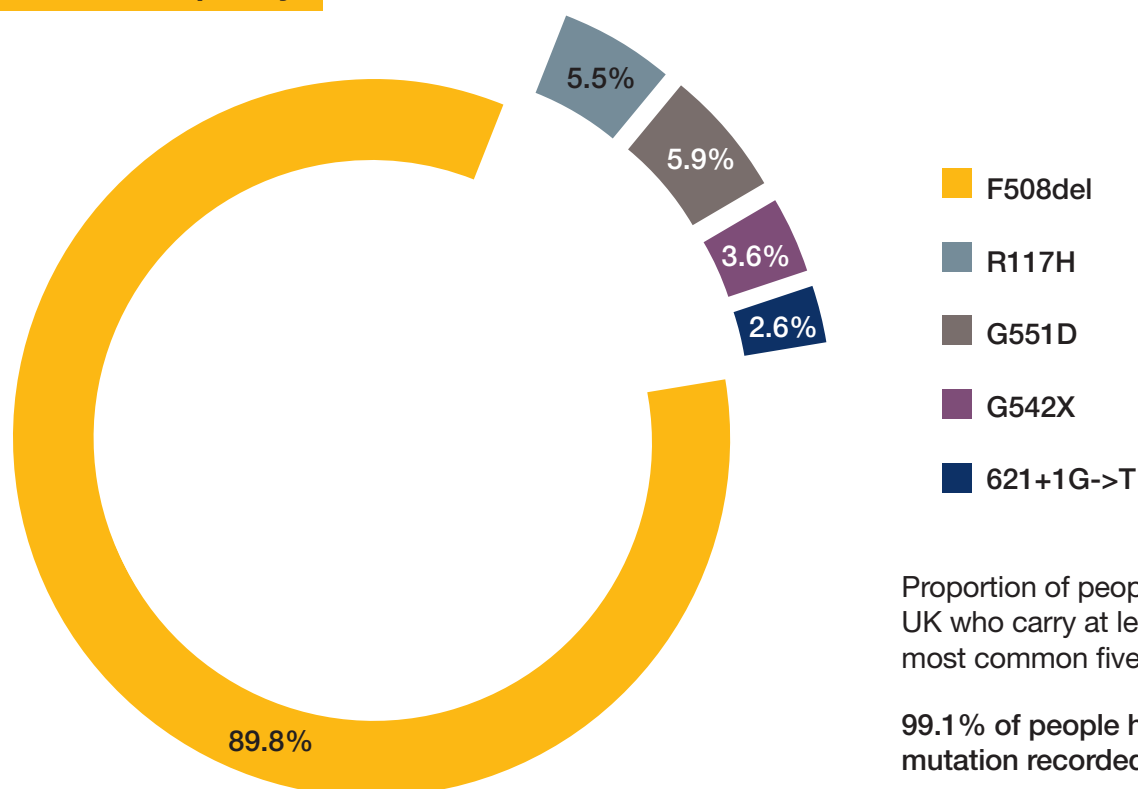


\*Active cycle of breathing and autogenic drainage.

## 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](http://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.