

Cystic Fibrosis strength in numbers

European Cystic Fibrosis Society
Annual Data Report 2013

At-A-Glance: what it means for the UK

Fighting for a
Life Unlimited

About this report

In February 2016, the European Cystic Fibrosis Society Patient Registry (ECFSPR) published its [Annual Data Report 2013](#). The analysis has been carried out using anonymised data entered by 27 participating countries, including the UK. This document is designed to highlight key points that the European data tells us about cystic fibrosis (CF) in the UK in 2013.

About the ECFSPR

The European Cystic Fibrosis Society Patient Registry (ECFSPR) measures data of people with cystic fibrosis living in Europe and neighboring countries who agree to be in the Registry.

The ECFSPR aims to compare cystic fibrosis care across Europe, to better understand CF, encourage new European standards of care, conduct research, and inform public health planning.

Some countries enter information directly into the European Registry. Those, like the UK, which have their own National Registry, import anonymous data into the ECFSPR once a year.

The UK is one of the biggest contributors to the ECFSPR, making up almost 1/3 of its' data overall.

A guide to this report

Some countries have well established data collection processes, enabling them to submit data on a large number of patients that are representative of their overall CF population. Other countries submit data on smaller numbers of patients, which may not represent their overall CF population. It is important to consider the amount and coverage of data submitted by each country when interpreting results, particularly if they appear to show differences in care or health outcomes. For details of the proportion of people with cystic fibrosis represented by each country's data, please see p16 of the [ECFSPR Annual Data Report](#).

Glossary

Chronic – Persistent, or long-lasting. When referring to Pseudomonas infection means this bacterial was present in >2 sputum samples in 12 months or in > than 50% of samples taken.

Homozygous – A person with two CF causing mutations that are the same.

Heterozygous – A person with two different CF causing mutations.

Median – The middle number, when all numbers are arranged from smallest to largest.

Prevalence – The overall number of people currently diagnosed with a condition.

Pseudomonas aeruginosa – A tough bacterial strain, that can cause respiratory infection.

Staphylococcus aureus – a bacteria that can cause respiratory infection.

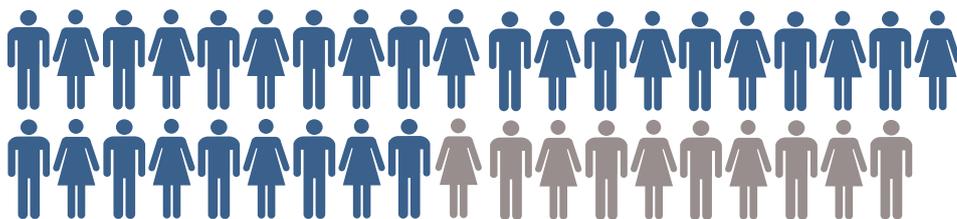
cysticfibrosis.org.uk

38,985

people with cystic fibrosis registered, from 27 countries.

10,336*

are people with cystic fibrosis from the UK.



The UK cystic fibrosis population makes up 26.5% of the ECFS Patient Registry data. In the UK, Registry data covers approximately 99% of people with cystic fibrosis. Elsewhere in Europe, the coverage is lower, meaning that not everyone with cystic fibrosis is represented by the data. Where not all people are included, the results drawn from the data should be interpreted with caution.

Reference: [ECFSPR Annual Data Report 2013: Table 1.1](#)

*The total is less than the UK CF Registry as since the 2013 report was published two people have been removed from the Registry.



3.6 months

median age of diagnosis across Europe.

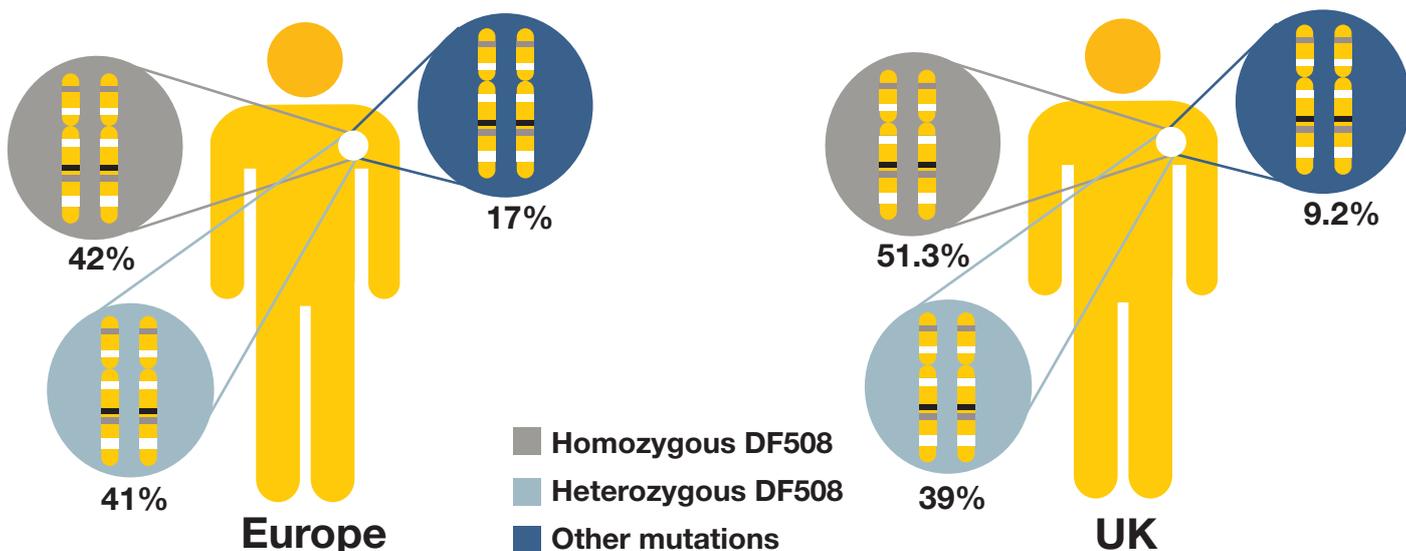


3 months

median age of diagnosis in the UK.

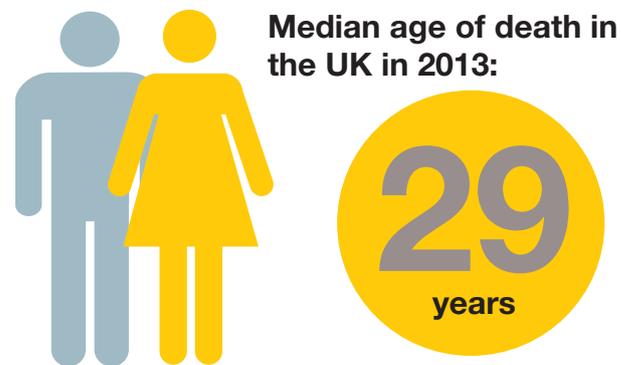
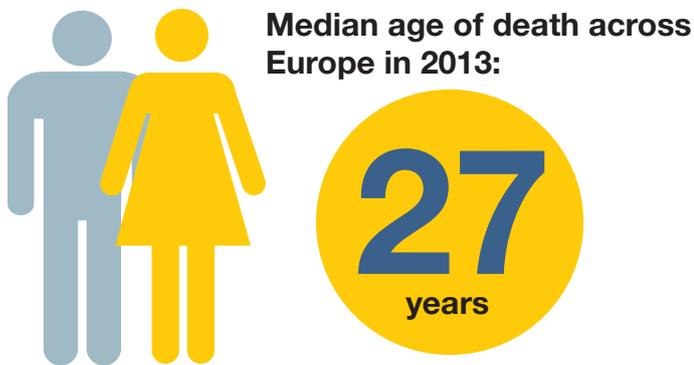
The UK has a national newborn screening programme for cystic fibrosis. Since its introduction the median age of diagnosis has dropped from five months in 2007 to two months in 2014.

Reference: [ECFSPR At-A-Glance report 2013: Page 2](#)



Different CF-causing gene mutations tend to be associated with ethnicity. The F508del mutation (also known as DF508) is the most common mutation in the UK.

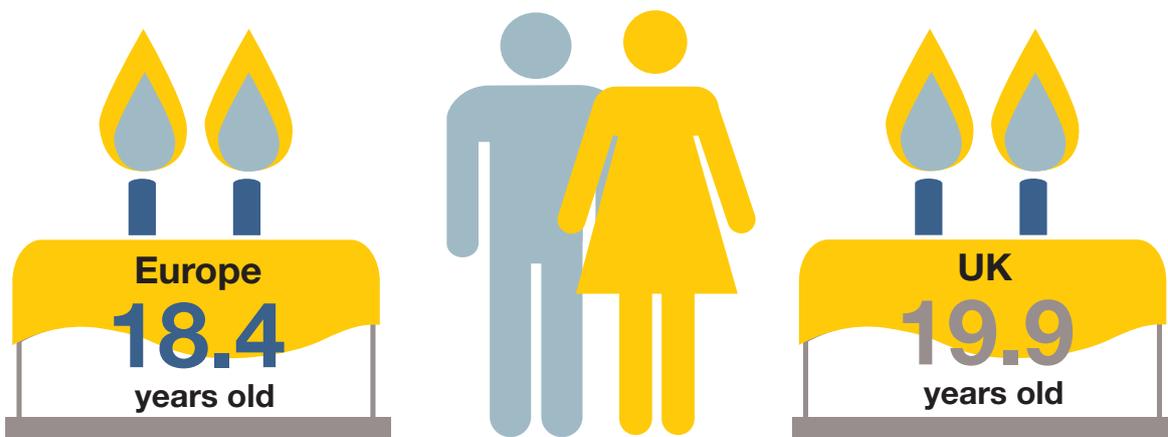
Reference: [ECFSPR Annual Data Report 2013: Figure 3.2](#)



Of the people with cystic fibrosis who died in 2013, in Europe half were aged 27 or younger, in the UK half were 29 or younger. Cystic fibrosis healthcare teams strive for a high median age of death, with fewer people dying very young from the disease.

Reference: [ECFSPR Annual Data Report 2013: Page 14](#)

The median age of people with cystic fibrosis in 2013



An older population suggests that people with cystic fibrosis are living longer. As people with cystic fibrosis live longer, you would expect to see more complications like lower lung function (FEV₁) and CF-related diabetes.

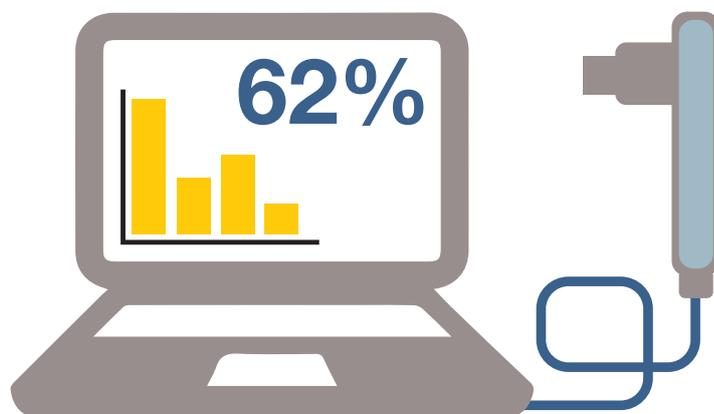
Reference: [ECFSPR Annual Data Report 2013: Page 14](#)

Of the 25 countries for which FEV₁ data is presented in the ECFSPR report, 19 (including the UK) have a median FEV₁% predicted over

60%



Median FEV₁% predicted for patients aged 18 or over for the UK:



FEV₁% predicted, a measure of lung health, is collected differently in different countries, which can affect the results. Some record the best value during the year, others the first taken. The UK reports the lung function recorded at the annual review assessment. Lower data coverage and completeness, as well as the median age of the population, in a country may also shape the results.

Reference: [ECFSPR Annual Data Report 2013: Table 4.2](#)



Chronic *Pseudomonas aeruginosa* prevalence across Europe:

33%

In the UK: **33%**

The risk of developing chronic *Pseudomonas aeruginosa* increases with age, so you would expect a higher prevalence in countries with a higher median age.

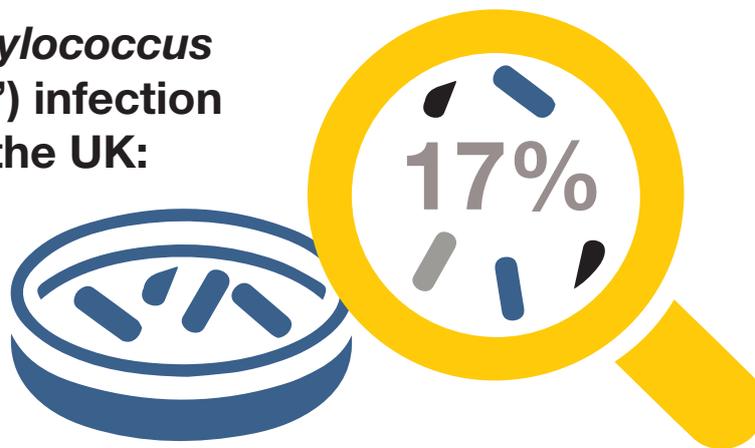
Reference: [ECFSPR Annual Data Report 2013: Table 5.1](#)



Reference: [UK CF Registry 2013 report: Page 25](#)



Chronic *Staphylococcus aureus* ('staph') infection prevalence in the UK:

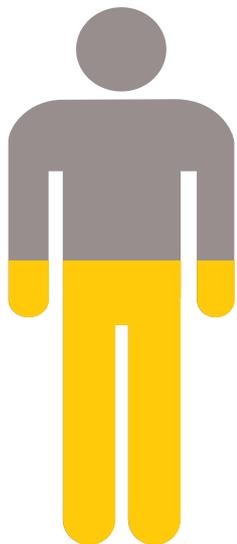


This low rate is likely to reflect the efficiency of the aggressive approach to treating this pathogen in the UK. Of the 22 countries submitting data to the ECFSPR, the percentage of patients with chronic staphylococcus aureus ranges from 10–69%.

Reference: [ECFSPR Annual Data Report 2013: Table 5.1](#)

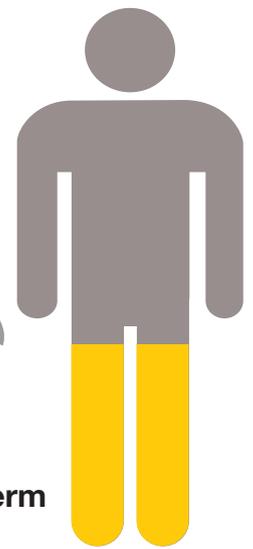
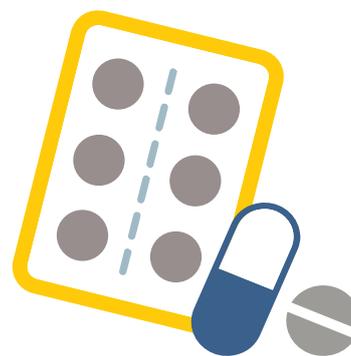
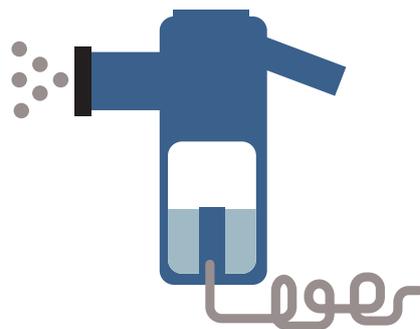


Reference: [UK CF Registry 2013 report: Page 25](#)



55%

of people in the UK with cystic fibrosis were prescribed long-term inhaled antibiotics.



43%

were prescribed long-term macrolides in the UK

High levels of long-term prescribing in the UK show that people with chronic infections are getting the therapies they need.

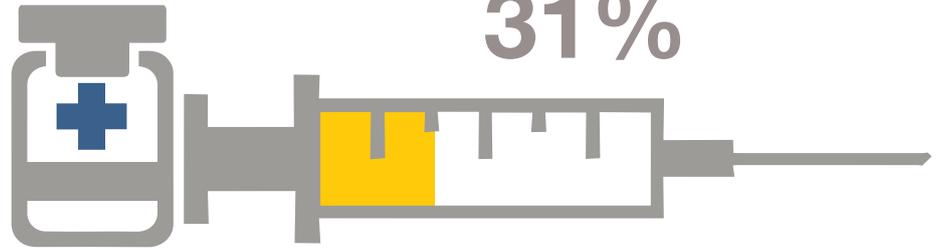
Reference: [ECFSPR Annual Data Report 2013: Table 7.5](#)





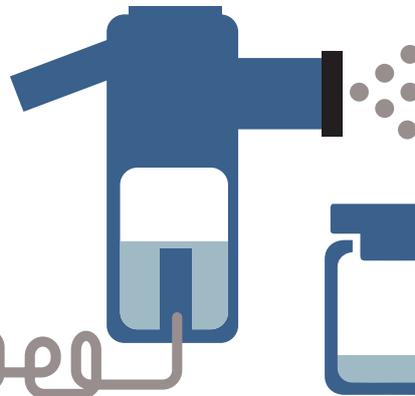
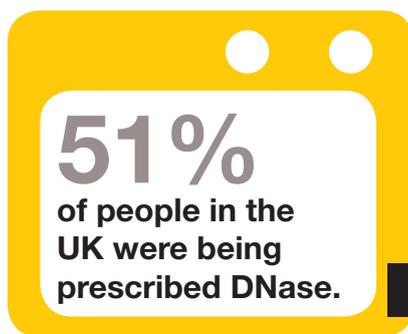
UK patients aged 18 and over with CF-related diabetes prescribed daily insulin:

31%



The high percentage of people with CF-related diabetes receiving insulin may indicate earlier intervention, which should have a positive long-term impact on health outcomes. Of the 22 countries submitting data to the ECFSPR, the percentage of patients with CF-related diabetes prescribed daily insulin ranged from 0–41%.

Reference: [ECFSPR Annual Data Report 2013: Table 7.1](#)



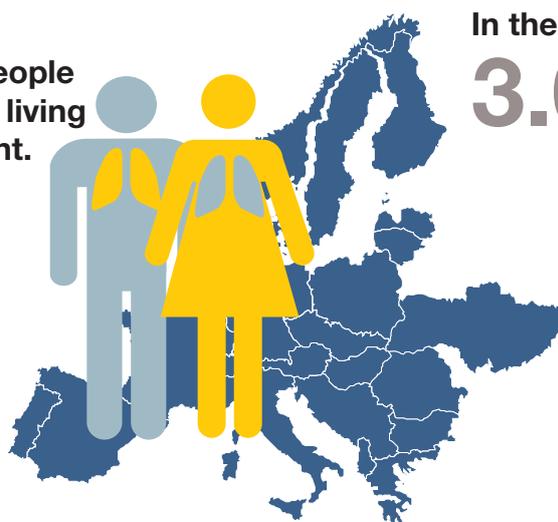
23%
were prescribed hypertonic saline in the UK.

These treatments, known as mucolytics, are used to thin mucus, so it's easier to cough out of the body. While the proportion of patients being prescribed DNase is roughly comparable with the other participating countries, the prescribing rate for hypertonic saline is relatively low.

Reference: [ECFSPR Annual Data Report 2013: Table 7.4](#)

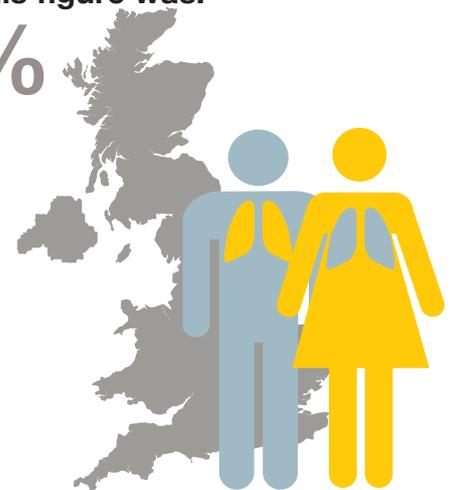


5.3% of people across Europe were living with a lung transplant.



In the UK this figure was:

3.0%



The UK has performed the second highest number of transplants across all participating countries, and 273 people were living with a lung transplant in 2013. However, evidence suggests that, while France utilises around 90% of its 'standard' lungs (donor organs that fit specific safety criteria), the UK only uses around 60%. Work to increase the number of donor organs that are used for transplant is ongoing.

Reference: [ECFSPR Annual Data Report 2013: Table 8.1](#)

