ecfs pr Patient Registry

2023 HIGHLIGHTS REPORT

Cystic Fibrosis in Europe Facts and Figures

The European Cystic Fibrosis Society Patient Registry (European CF Registry) is proud to present this report with key information about how cystic fibrosis (CF) affects people with CF and their families throughout Europe.

The European CF Registry collects, measures and compares data of people with CF living in Europe and neighbouring countries who agree to be in the Registry. The information is important to better understand CF, encourage new European standards of care and treatment, conduct research and inform public health planning.

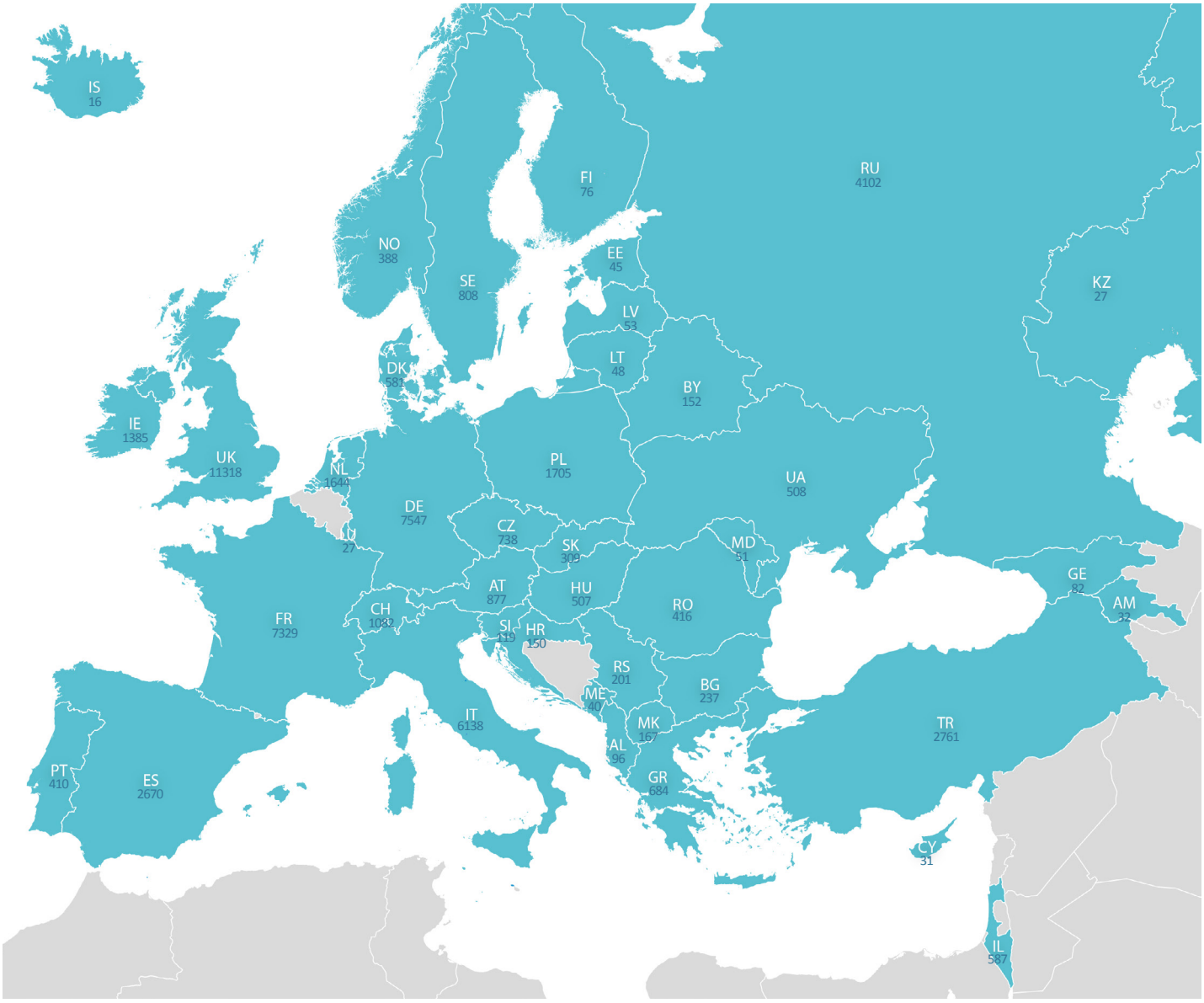
If you want to know more about the European CF Registry visit our website.

January 2025

www.ecfs.eu/ecfspr

WHO & FROM WHERE?

Total number of people and countries that contributed 2023 data to the ECFSPR



56,144

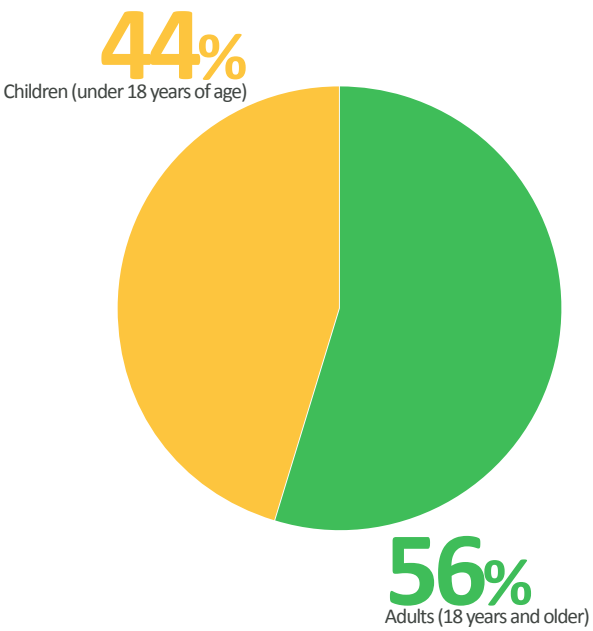
Registered people with CF in 42 countries

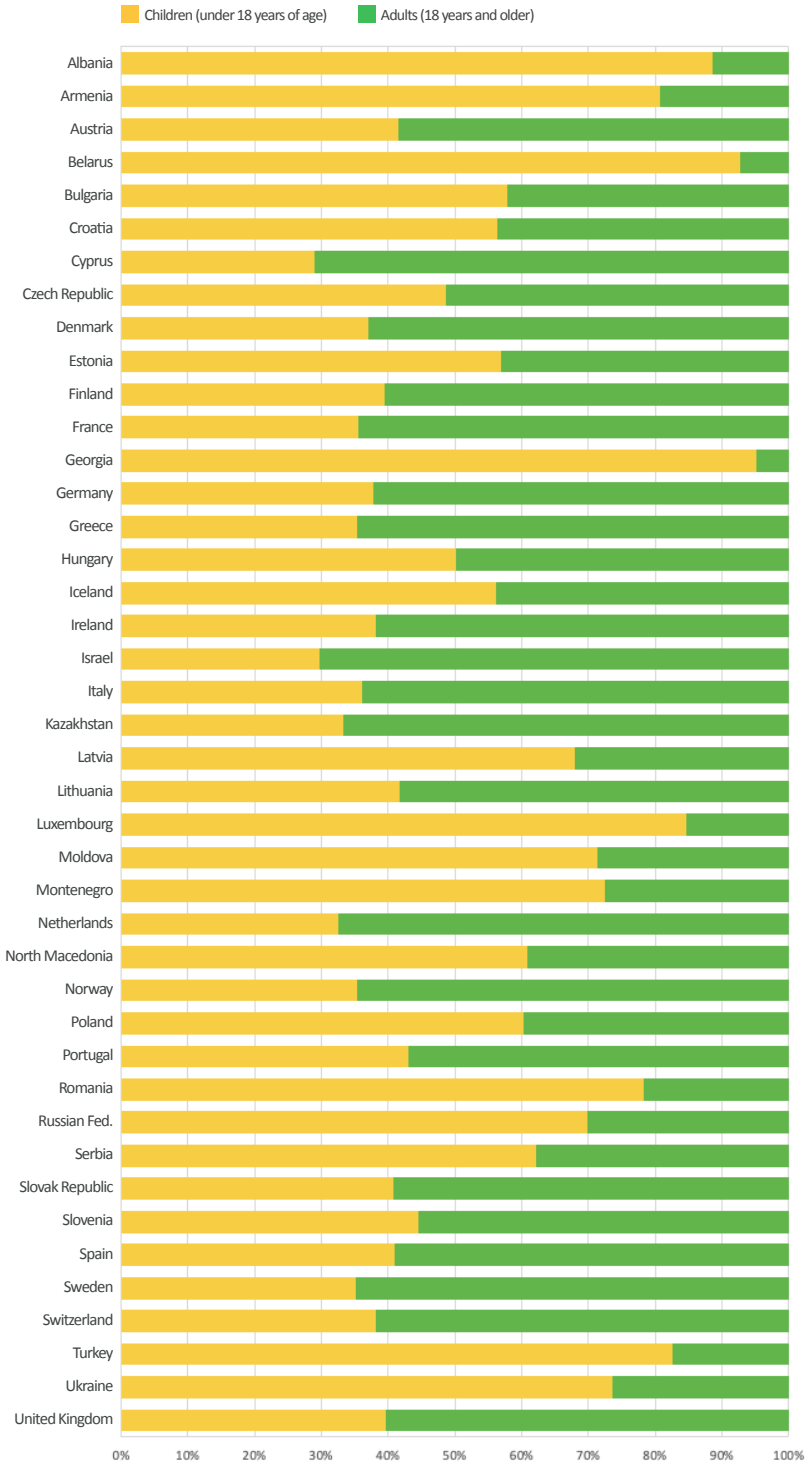
For some countries only a few individual centres sent data to the European CF Registry.

People with CF

% CHILDREN & ADULTS

The proportion children-adults varies between the countries.

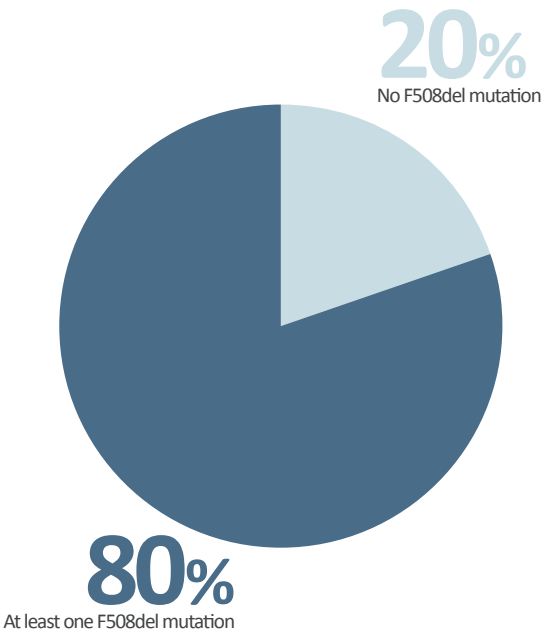


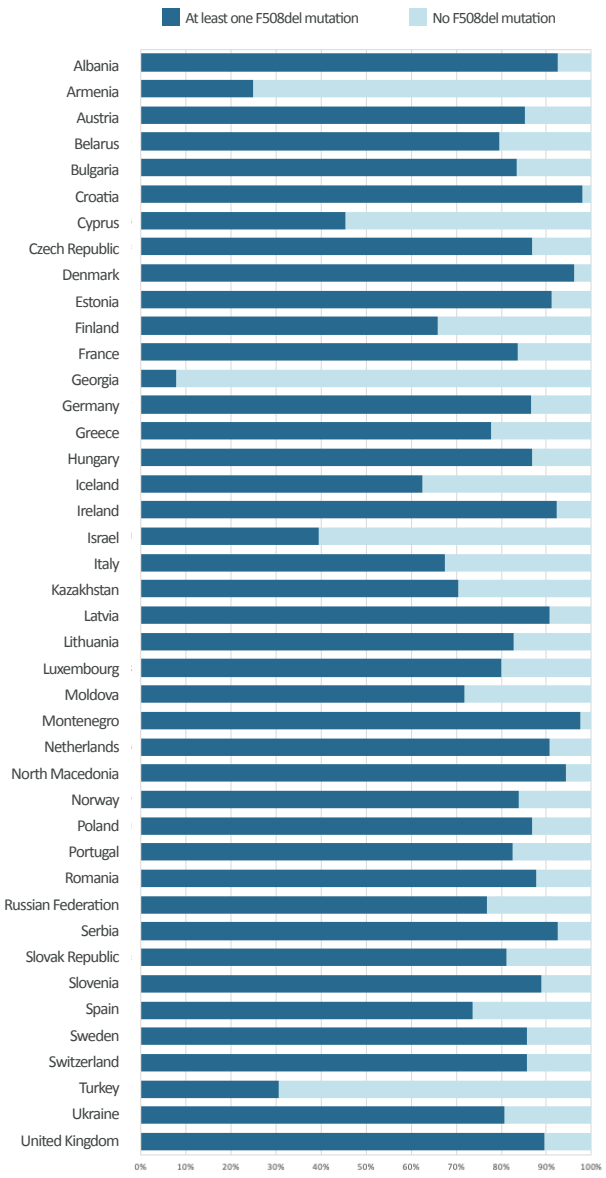


MUTATIONS AND DIAGNOSIS

Distribution of mutations

People with CF have two CF-causing mutations in their DNA.
One mutation is inherited from the mother and one from the father.
F508del is the most common CF-causing mutation in Europe.





Diagnosis of CF

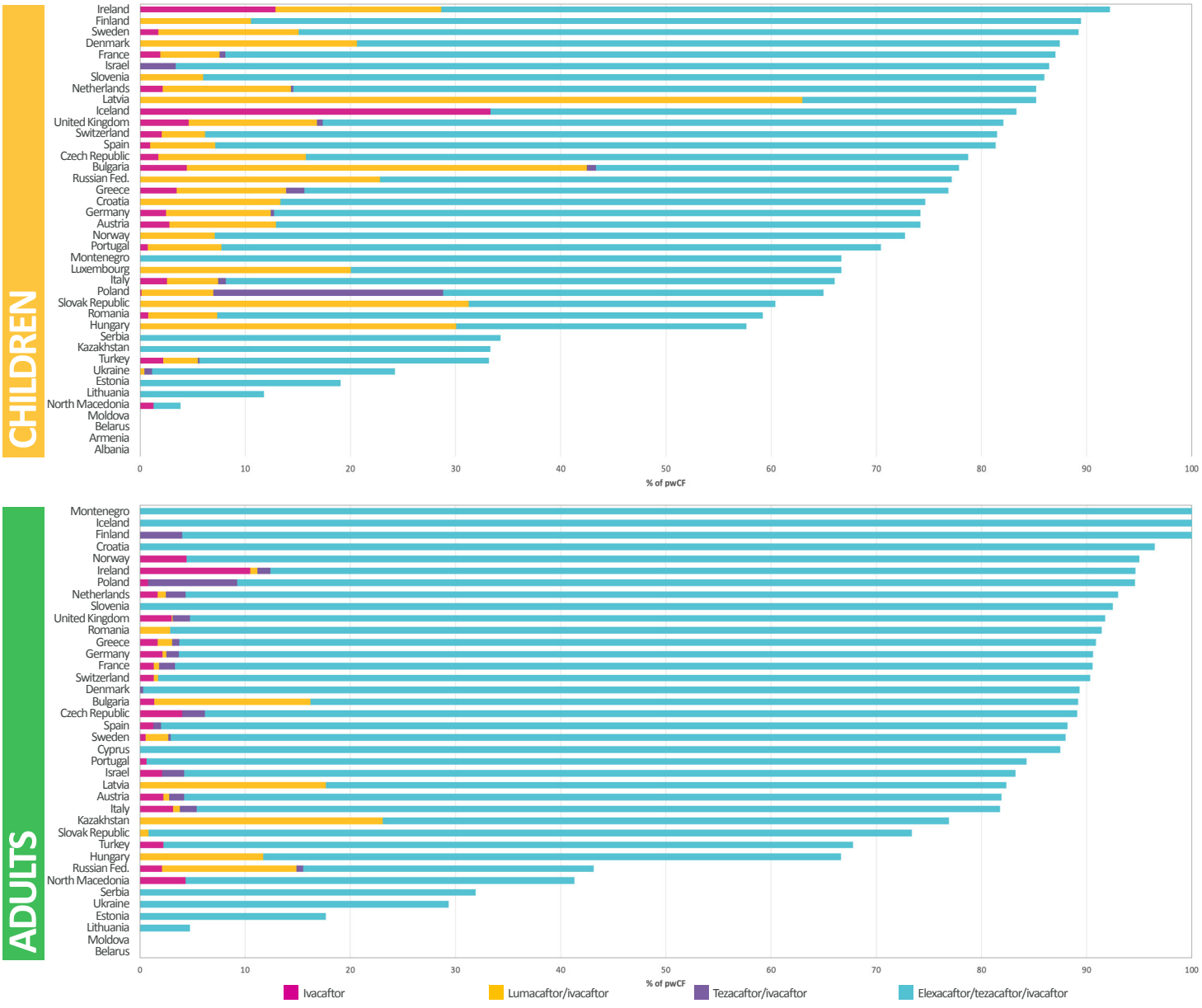
Most people are diagnosed with CF in their childhood. The age at diagnosis, especially in children and adolescents, is strongly influenced by the presence or absence of a national CF newborn screening programme.
The median age at CF diagnosis is 3.6 months, meaning half of all people with CF are diagnosed before they are 3.6 months old and the other half are diagnosed at a higher age. The majority of people with CF are diagnosed before they are a year old.

MEDIAN AGE DIAGNOSIS
3.6 MONTHS

CFTR MODULATORS

Modulator information

CHILDREN & ADULTS with CF eligible for and treated with at least one modulator - by country and last CFTR modulator prescribed, seen in 2023 and who have never had a transplant.



Note: Not included in this graph are countries with a high number of missing values or when <5 people with CF.

CFTR modulators are medicines that act by improving the production, functioning or processing of the malfunctioning CFTR protein which is the cause of CF. Not everyone with CF can benefit from a CFTR modulator, either because the medicines do not work for the mutation(s) they have or because the medicines are not reimbursed by their national health service. The combination of elexacaftor/tezacaftor/ivacaftor is currently prescribed for most children and adults with CF. In the majority of countries in Europe more than 50% of all adults with CF are now eligible and treated with one of the CFTR modulators.

INFECTION

People with CF who have a lung infection

Pseudomonas aeruginosa

CHILDREN 19%

ADULTS 38%

Methicillin-sensitive Staphylococcus aureus

CHILDREN 53%

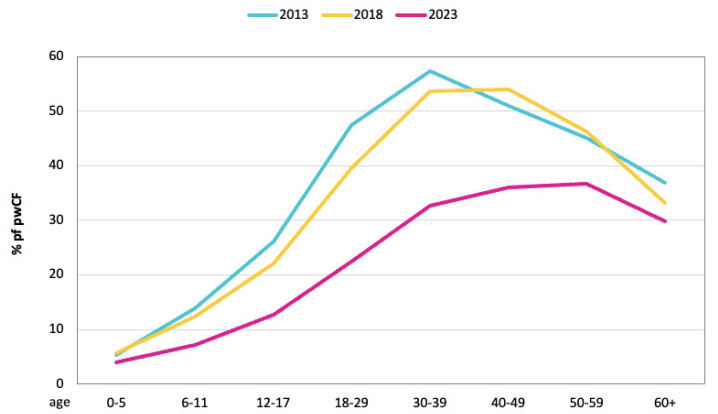
ADULTS 45%

These figures show how many people with CF[1] were infected at least once with *Pseudomonas aeruginosa* and methicillin-sensitive *Staphylococcus aureus* in 2023. These bacteria are the most common causes of lung infection in people with CF.

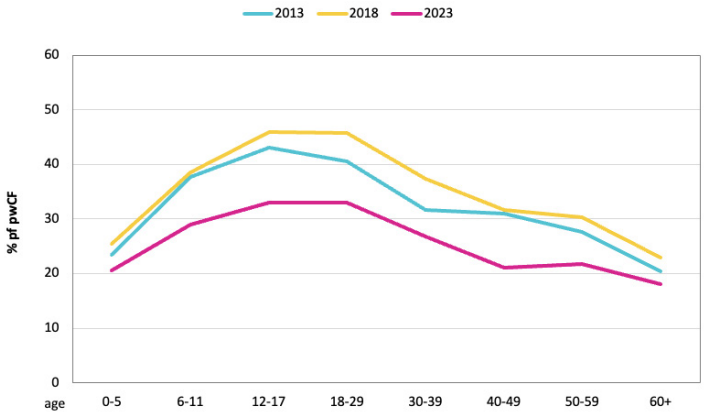
[1] People seen in a CF centre in 2023 who have never had a transplant.

Over the years

Chronic Pseudomonas aeruginosa



Chronic Methicillin-sensitive Staphylococcus aureus

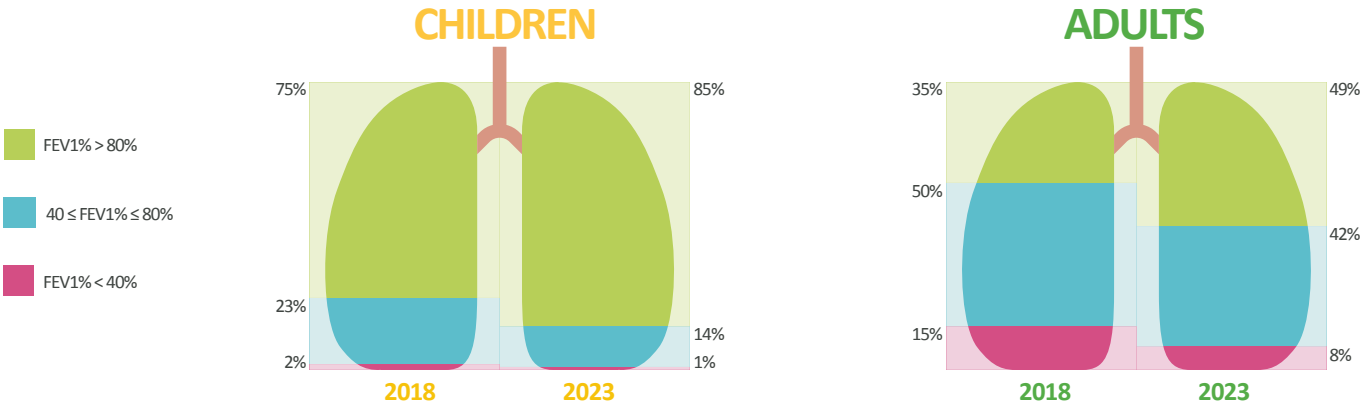


The prevalence of chronic *Pseudomonas aeruginosa* infection increases with age, up to around 30 years old, though over the last 10 years detection of *Pseudomonas aeruginosa* in all age groups has fallen. With chronic methicillin-sensitive *Staphylococcus aureus*, the infection rate decreases in adulthood (18 years and older), with no significant change in the last 10 years.

HEALTH INFORMATION

Lung function

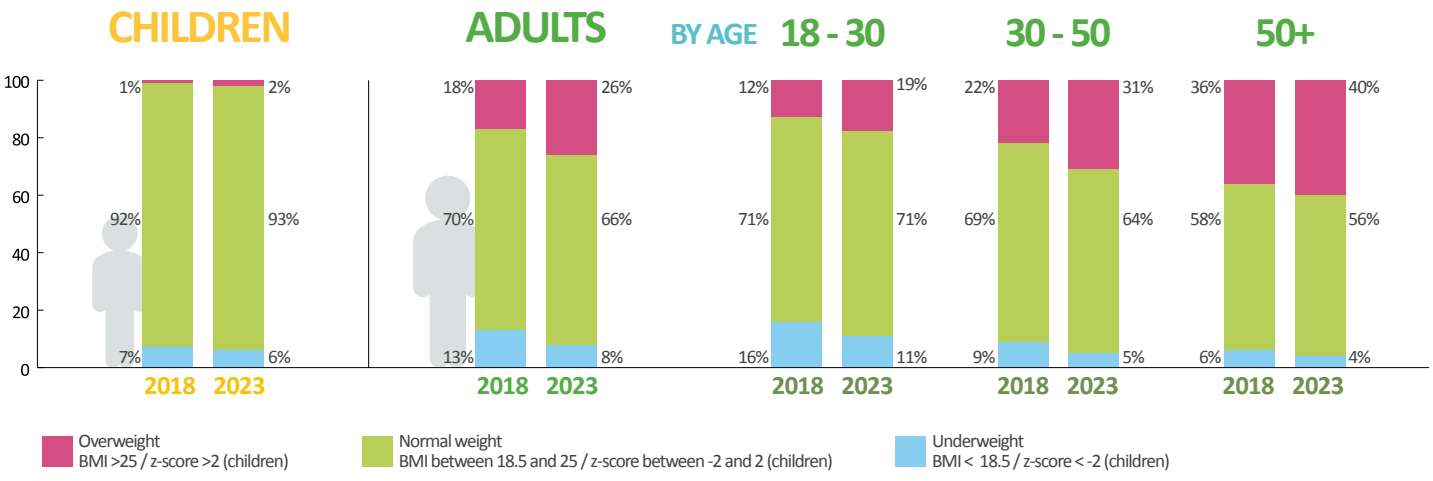
Overall, the number of children and adults with a healthy lung function (FEV1% of 80% or higher) is higher in 2023 than in 2018.



FEV1, or Forced Expiratory Volume 1, is a measure of lung function. It is the maximum amount of air that can be forcefully exhaled in the first second after taking a deep breath.
For people with CF, FEV1 is measured then expressed as a percentage of the average value (FEV1%) for healthy people of the same age, gender and height which is set at 100%. An FEV1% of 80% or higher is considered a healthy lung function.

Height and weight

The percentage of people with CF who are overweight increased in all age groups between 2018 and 2023.
Fewer people are underweight in 2023 than in 2018.



BMI (body mass index) is a measure that determines whether or not someone has a healthy weight compared to their height.
In children with CF, their BMI is compared to that of healthy children and is expressed by a z-score (how different something is from the reference value). A z-score of 0 means the BMI is the same as that of a healthy child of the same age and gender.
As people with CF get older, BMI increases.

ecfs pr Patient Registry

2023 HIGHLIGHTS REPORT

Cystic Fibrosis in Europe
Facts and Figures

version 1.7

Contact us

Follow us

www.ecfs.eu/ecfspr